Chapter 125 – Glomus Tumors

Barry E. Hirsch

araganglioma or glomus tumor is a tumor of the neuroendocrine system derived from neural crest cells and chemoreceptor cells. The tumor was termed a *chemodectoma* by Mulligan in 1950.[1] The ability to pick up chromium salts led to the designation of the histologic staining characteristic as chromaffin or nonchromaffin positive. The largest concentration of paraganglionic cells is with the adrenal medulla. The other predominant locations of paraganglionic tissue are found in the branchiomeric chemoreceptor system of the aortic arch, carotid bifurcation, and temporal bone. Guild described the location of other glomus bodies within the temporal bone, identifying an average of 2.82 foci, half of which were in the jugular bulb and one fourth on the promontory of the cochlea.[2] The term commonly accepted to describe neoplastic tumors of the paraganglionic receptors is *glomus tumor.* Glomus tumors of the temporal bone are neoplasms arising from normally occurring paraganglionic bodies or formations that are located on the dome of the jugular bulb and along the course of Jacobson's nerve (ninth cranial nerve) onto the cochlear promontory. On rare occasions, these tumors take origin from the facial nerve.[3] The tumors of the temporal bone may be grouped into two categories: those arising from the middle ear (promontory) and those arising from the jugular bulb. These are termed *glomus tympanicum tumors* and *glomus jugulare tumors.* As tumor expansion occurs, it may be difficult to distinguish the precise origin of the lesion. Thus larger tumors involving both the middle ear and the jugular bulb are termed *jugulotympanic glomus tumors.*

Although most tumors are sporadic, familial glomus tumors may occur in 20% of patients with glomus tumors. Many of the tumors are multicentric and bilateral at the time of presentation, which typically occur at an earlier age in successive generations in families harboring genetic mutations. Investigation into the hereditary patterns of familial paraganglioma has resulted in the identification of four loci termed *PGL1, PGL2, PGL3,* and *PGL4.* The first two loci were found on chromosome 11 at bands 11q23.1 and 11q13.1. Both regions are associated with an autosomal dominant pattern and maternal imprinting. Inheriting the gene from the mother portends a lower incidence of transmission compared with passing the gene onto offspring from the father. Mutation in genes responsible for production of proteins that are critical components of a cellular oxygen-sensing system may permit proliferation of chief cells in paraganglionic cells. The loci for PGL3 and PGL4 are on chromosome 1. Heth wrote an excellent review of the basic science of glomus jugulare tumors.[4]

Glomus tumors are slow-growing, invasive, highly vascular neoplasms that are in critical proximity to the lower cranial nerves. Although locally destructive, glomus tumors are rarely considered to be malignant. It may be difficult to determine whether a glomus tumor is malignant. Some of the histologic features suggestive of malignancy include increase in mitotic activity, necrosis in the center of the cell nests, and vascular invasion. The hallmark of aggressive malignant behavior is the presence of tumor in the regional lymph nodes or distant organs confirmed with histologic examination.

Metastatic tumors have been found in the vertebrae, ribs, spleen, and lungs. In a series of 175 temporal bone glomus tumors, 5.1% were classified as malignant. Conventional external beam radiation therapy (XRT) is not advocated as the primary modality of treatment because viable tumor cells can remain following radiation treatment. Five-year survival for malignant glomus tumors of the temporal bone is 71.2%.[5]

Two classification systems have been offered for nomenclature and staging. Fisch described and modified a classification system based on the size and extent of the tumor (Table 125-1).[6,7] There are four categories (A, B, C, and D) of staging tumors, from limited to the middle ear cleft (type A) to large lesions demonstrating intracranial extension (type D). The Glasscock and Jackson classification system divides the tumors into tympanic or jugulare tumors (Table 125-2).[8] Each group is further subdivided into four types of progressively larger tumors, from small lesions limited to the site of origin to more extensive tumors involving regional areas of the temporal bone. Although the latter system attempts to define the anatomic origin of the tumor, both classifications address the pertinent issues of tumor size, petrous apex or carotid artery involvement, and intracranial extension. This provides critical information necessary to design an operative approach and plan.

Table 125-1 -- GLOMUS TUMOR CLASSIFICATION: FISCH

Type	Description
A	Tumor limited to middle ear cleft (promontory)
B	Tumor confined to middle ear, hypotympanum, and mastoid
C_1	Tumor eroding jugular bulb and carotid foramen; not invading carotid artery

Type	Description
C_2	Tumor involving infralabyrinthine and apical temporal bone; erosion of vertical carotid artery
C_3	Tumor involving infralabyrinthine and apical temporal bone; erosion of horizontal carotid artery
C_4	Tumor involving infralabyrinthine and apical temporal bone; tumor grows to foramen lacerum and cavernous sinus
D_1	Intracranial tumor <2 cm
D_2	Intracranial tumor >2 cm; *e,* extradural; *i,* intradural
Di_3	Unresectable intracranial extension

Data from Fisch U: Infratemporal fossa approach for glomus tumors of the temporal bone. Ann Otol Rhinol Laryngol 91:474-479, 1982; and Fisch U, Mattox D: Classification of glomus temporal tumors. In Fisch U, Mattox D (eds): Microsurgery of the Skull Base. Stuttgart and New York, Georg Thieme, 1988, pp 149-153.

Table 125-2 -- GLOMUS TUMOR CLASSIFICATION: GLASSCOCK-JACKSON

Type	Description
	Giomus Tympanicum
I	Small mass limited to promontory
II	Tumor completely filling middle ear space
III	Tumor filling middle ear and extending into mastoid
IV	Tumor filling middle ear, extending into mastoid or through tympanic membrane to fill external auditory canal; may extend anterior to carotid
	Giomus Jugulare
I	Small tumor involving jugular bulb, middle ear, and mastoid
II	Tumor extending under internal auditory canal; may have intracranial extension
III	Tumor extending into petrous apex; may have intracranial extension
IV	Tumor extending beyond petrous apex into clivus or infratemporal fossa; may have intracranial extension

Data from Jackson CG, Glasscock ME, Harris PF: Glomus tumors: Diagnosis, classification, and management of large lesions. Arch Otolaryngol 108:401-410, 1982.

PATIENT SELECTION

Patients with glomus tumors frequently report a history of symptoms of pulsatile tinnitus, hearing loss, aural fullness, and not uncommonly, cranial nerve dysfunction such as facial paresis, dysphasia, or hoarseness. Patients should be questioned regarding symptoms including tachycardia, palpitations, headaches, pallor, excessive perspiration, nausea, and problems of control of blood pressure, all of which are related to excess catecholamine secretion by the tumor. A complete otologic and head and neck examination is required. The tympanic membrane and middle ear space are inspected for tumor. When tumor is in the middle ear, a red mass is noted behind the tympanic membrane, and pulsation can be observed under high magnification (Fig. 125-1). The patient should be evaluated for evidence of weakness or dysfunction of cranial nerves VII, VIII, IX, X, XI, and XII. Both sides of the neck are palpated for mass lesions in the jugulodigastric and carotid bifurcation areas. Audiometry should be performed to determine the presence of a conductive, mixed, or sensorineural hearing loss.

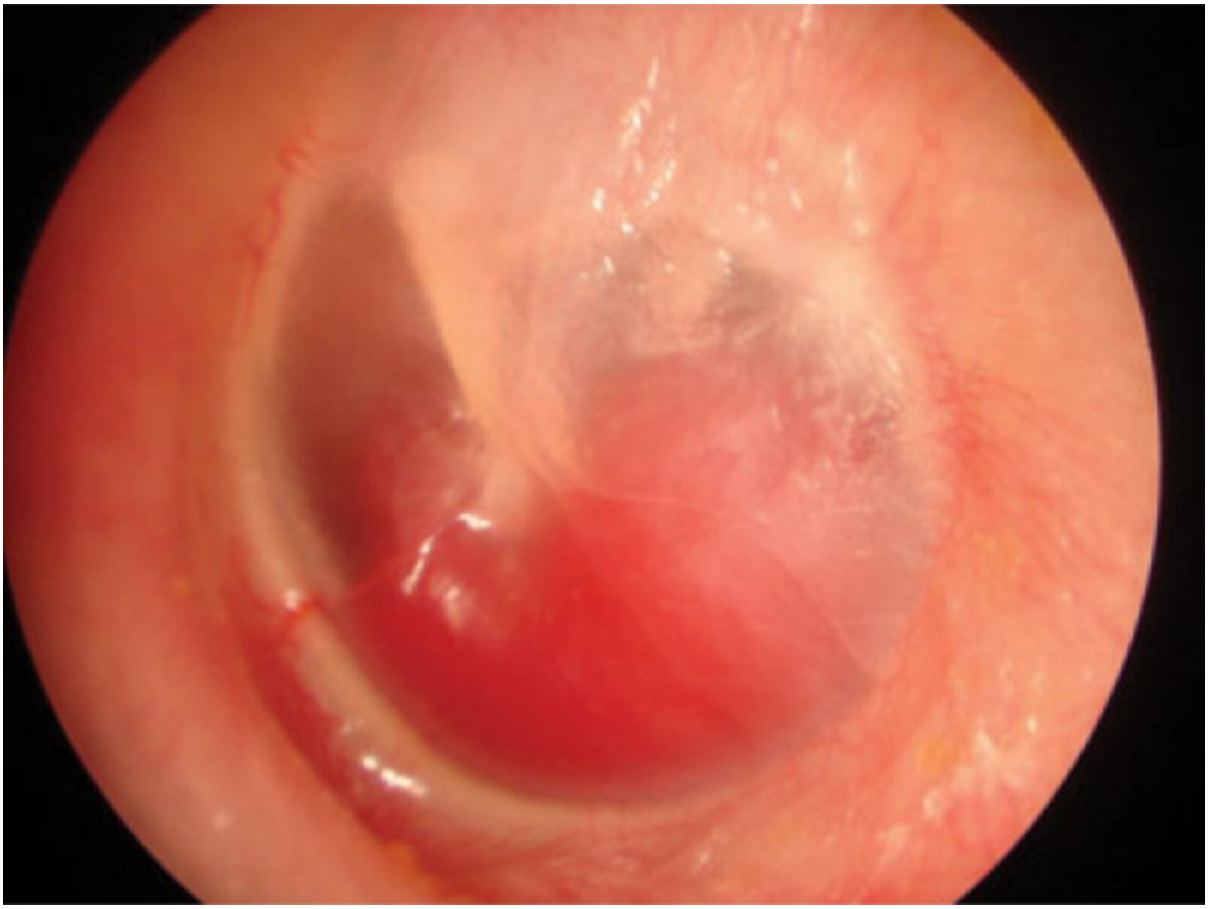

Figure 125-1 Left tympanic membrane with a pulsatile circumscribed vascular tumor filling the inferior portion of the middle ear.

Once a glomus tumor has been presumptively diagnosed by the history and physical examination, the diagnosis and evaluation of the type and extent of the tumor are required. Along with a history of pulsatile tinnitus or the presence of a vascular mass in the middle ear, radiologic imaging can demonstrate characteristic features strongly suggestive of glomus tumor. On computed tomography (CT) scanning, these features include the location of the lesion, bone erosion around the jugular bulb and carotid artery, and enhancement following administration of contrast. Large glomus tympanicum tumors can extend inferiorly into the hypotympanum and erode trabeculated bone toward the jugular bulb. Similarly, glomus jugulare tumors expand superiorly toward the hypotympanum and middle ear. It can occasionally be difficult to definitively characterize the site of origin. The differential diagnosis of an expanding, bone-eroding enhancing tumor in the jugular fossa region include neurofibroma, schwannoma, lymphoma, meningioma, and metastatic disease. Biopsy is to be avoided owing to the vascular nature of the tumor. Only after the extent of tumor is defined (middle ear, jugular bulb, carotid artery, clivus, intracranial, or intradural extension) can decisions regarding optimal management be made. Glomus tumors are rarely malignant. However, because of their propensity for progressive and invasive growth and involvement of major vessels and lower cranial nerves, treatment is routinely required. It is only the occasional asymptomatic small tumor in an older person that can be followed up radiographically for evidence of growth.

Paragangliomas can also occur with other endocrine tumors associated with familial syndromes. Such tumors include carcinoma of the thyroid gland, medullary carcinoma of the thyroid, and those found with multiple endocrine neoplasia (MEN) type I, consisting of pituitary adenoma, parathyroid adenoma, and pancreatic islet cell adenoma.

Treatment options for glomus tumors of the temporal bone include observation with serial scanning to monitor for growth, microsurgical excision, and radiation therapy. Complete surgical resection minimizes the chance for recurrence but may entail significant morbidity. Radiation therapy does not eliminate the tumor but is intended to halt tumor progression. Proponents for surgery suggest radiation is indicated for palliation, following incomplete tumor excision or for patients who are medically infirm or elderly. However, there is strong evidence indicating that radiotherapy could also be considered the definitive treatment strategy for skull base glomus tumors. Glomus jugulare tumors have traditionally been treated by fractionated XRT. Hinerman and colleagues summarized their experience of 53 patients with 55 temporal bone tumors (46 were glomus jugulare and 9 were glomus tympanicum tumors). Patients received megavoltage radiation over a continuous course. With almost half of the patients followed up for 15 years, local tumor control was achieved in 93% of the previously untreated tumors and 92% of those previously treated.[9] The long-term results using stereotactic radiosurgery remain to be seen, but some

early experience suggests there may be insufficient control of tumor if the planning treatment volume is kept too tight.[10] In a series of 42 patients treated with gamma knife surgery, 19 received radiation as their primary therapy and 23 patients for recurrent glomus jugulare tumor. The mean follow-up was 44 months (range 6 to 149 months). Although this is considered a relatively short period of time, tumor control was accomplished, in that 31% decreased in size, 67% were unchanged, and the only one that grew was in a patient who failed previous radiation therapy.[11]

We propose that healthy, young (younger than 65 years of age) patients should consider surgical resection. Large glomus tumors usually encroach on the ninth and tenth cranial nerves, putting them at risk during resection. Asymptomatic patients must be assessed as to whether they could tolerate the consequences of pharyngeal and laryngeal dysfunction. If sacrifice of the ninth and tenth cranial nerves is anticipated, patients with limited pulmonary reserve would be unlikely surgical candidates. In contrast, patients with a history of dysphagia or hoarseness who demonstrate vocal cord dysfunction can better tolerate sacrifice of the ninth and tenth cranial nerves. These people have typically developed compensatory mechanisms to minimize problems with swallowing and aspiration.

Large glomus jugulare tumors in relatively asymptomatic patients pose a significant dilemma in their optimal management. Specifically, patients having large tumors where surgical resection will likely incur new postoperative lower cranial nerve deficits often results in significant morbidity. The possible need for a tracheotomy, nasogastric tube placement, or percutaneous gastrostomy feeding tube, along with complications of aspiration, pneumonia, inanition, and weakness of the shoulder and tongue, must be considered by the patient and their family as well as the physicians and surgeons involved with the patient's care. Although radiation treatment in a relatively young person is not without short- or long-term risk, the aforementioned complications may have immense impact on the patient's future quality of life. Long-term results following total resection of class C and D tumors revealed that full rehabilitation may take 1 to 2 years. In a series by Briner and colleagues, 97% of patients finally resumed improved function, deeming their postoperative dysphasia tolerable and allowing a more normal social life.[12]

The optimal management program for patients 65 years of age and older should be individualized. Older patients with compromised pulmonary function or other chronic unstable medical conditions are not good surgical candidates. Radiation therapy has been shown to be an effective method for tumor palliation and thus is a reasonable alternative treatment.

PREOPERATIVE PLANNING

The granules contained within the chief cells of these tumors contain the precursors for catecholamine synthesis. Patients with intermittent or labile hypertension or those with signs and symptoms of a hypermetabolic state should be evaluated for a secreting tumor. Unlike the adrenal gland, glomus tumors lack the enzyme phenylethanolanine N-methyltransferase, which converts norepinephrine to epinephrine. This is why norepinephrine is the mostly commonly secreted chemotransmitter. Urine collected for 24 hours is screened for metanephrine, vanillylmandelic acid, epinephrine, and norepinephrine. Because the incidence of secreting glomus tumors is low (1% to 3%), elevated levels of these vasoactive peptides should also prompt investigation for pheochromocytoma. A CT scan of the abdomen is used to rule out a retroperitoneal adrenal or extra-adrenal tumor. Magnetic resonance imaging (MRI) can also be used, but it may be plagued by movement artifact. Further diagnostic evaluation is necessary when elevated levels of serum or urine catecholamine precursors and metabolites are identified. The workup for potential metastatic disease may also demand other radiographic techniques to locate sites of distant spread. Imaging with ^{123}I MIBG (metaiodobenzylguanidine) scintigraphy is picked up by active endocrine tissue such as paraganglioma, pheochromocytoma, and neuroblastoma. Anesthesia consultation for coordinating pharmacologic blockade is warranted for patients with elevated catecholamine levels. Both α- and β-adrenergic blockers such as phentolamine and propranolol, respectively, may be necessary.

Patients considered to be surgical candidates must have the extent of tumor involvement delineated. This entails imaging techniques that define the relationship of the tumor to critical anatomic structures. Currently, the diagnosis is usually made with contrast-enhanced CT or MRI scans. Bone-windowed, thin-cut (1.5-mm) CT images define the relationship of the tumor to the carotid artery, sigmoid sinus, jugular bulb, pars nervosa, middle ear, facial nerve, otic capsule, and posterior fossa (Fig. 125-2). MRI enhances the evaluation of glomus tumors of the temporal bone and skull base. Contrast-enhanced MRI identifies the tumor often with a speculated pattern described as “salt and pepper” vascularity with greater sensitivity in demonstrating tumors of the skull base. In addition to delineating the tumor, MRI demonstrates the relationship of the surrounding soft tissues and major vessels.

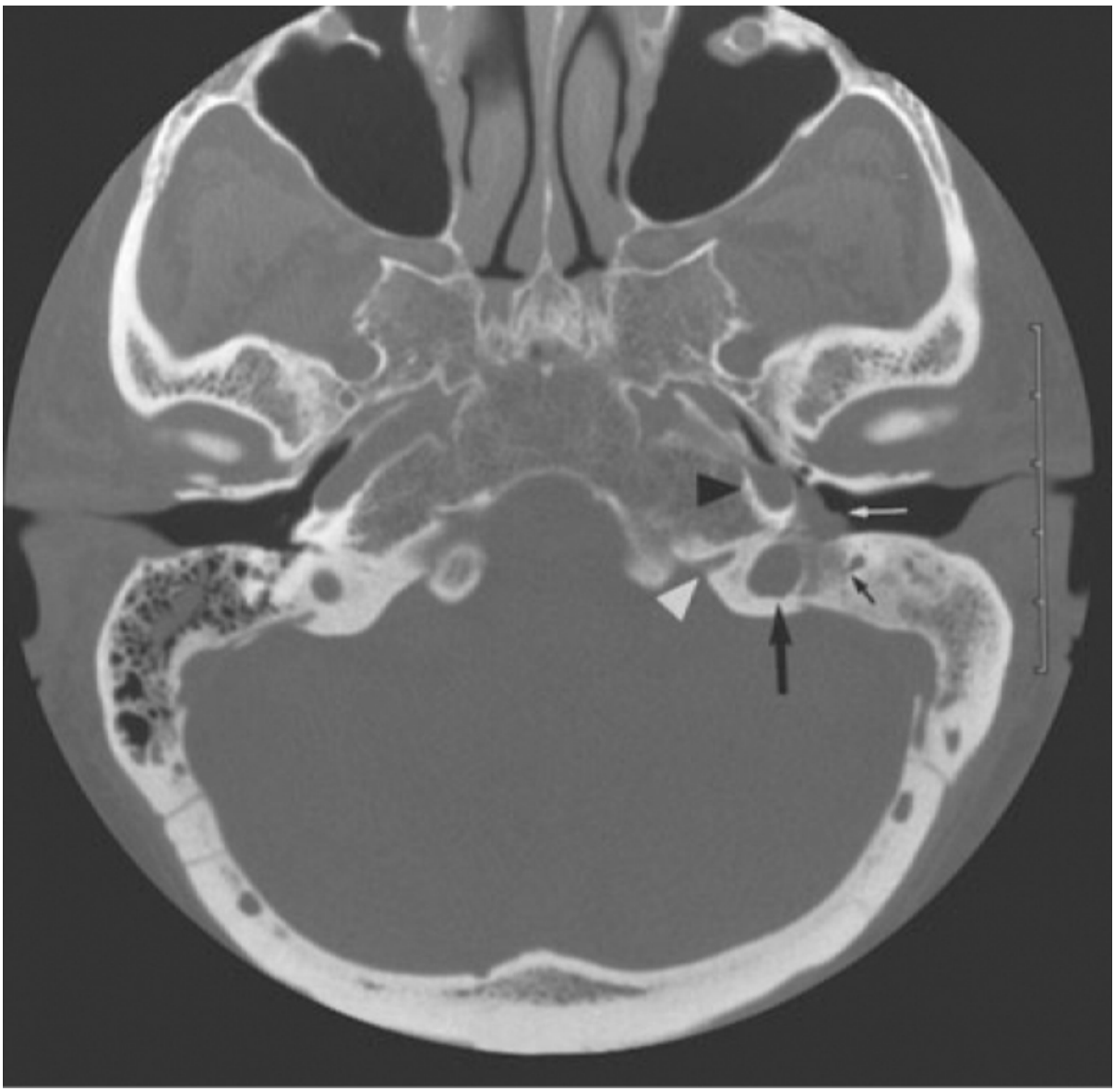

Figure 125-2 Bone-windowed computed tomography (CT) scan demonstrating the left middle ear and a hypotympanic mass *(white arrow)* infiltrating and eroding bone between the carotid artery *(black arrowhead)*, the jugular vein *(large black arrow)*, and the medial aspect of the facial nerve *(small black arrow)*. The pars nervosa is not affected *(white arrowhead)*.

The soft tissue resolution and information obtained from MRI scans help differentiate tumor from surrounding soft tissue, brain parenchyma, and mucosal changes within the temporal bone (Fig. 125-3). The relatively new technique of magnetic resonance angiography (MRA) also shows the relationship of the tumor to carotid artery, sigmoid sinus, and jugular bulb (Fig. 125-4). MRA shows the proximity of the tumor or displacement of the major vessels along with the predominant blood supply. MR venography highlights the anatomy of the transverse, sigmoid, and petrosal sinuses; the jugular bulb; and the descending jugular vein. It is most helpful if the patency of these venous structures can be verified or if thrombosis is present. Although occasionally fraught with difficulty in interpretation, information regarding flow characteristics in these vessels may be helpful in determining the degree of tumor involvement and obstruction to flow.

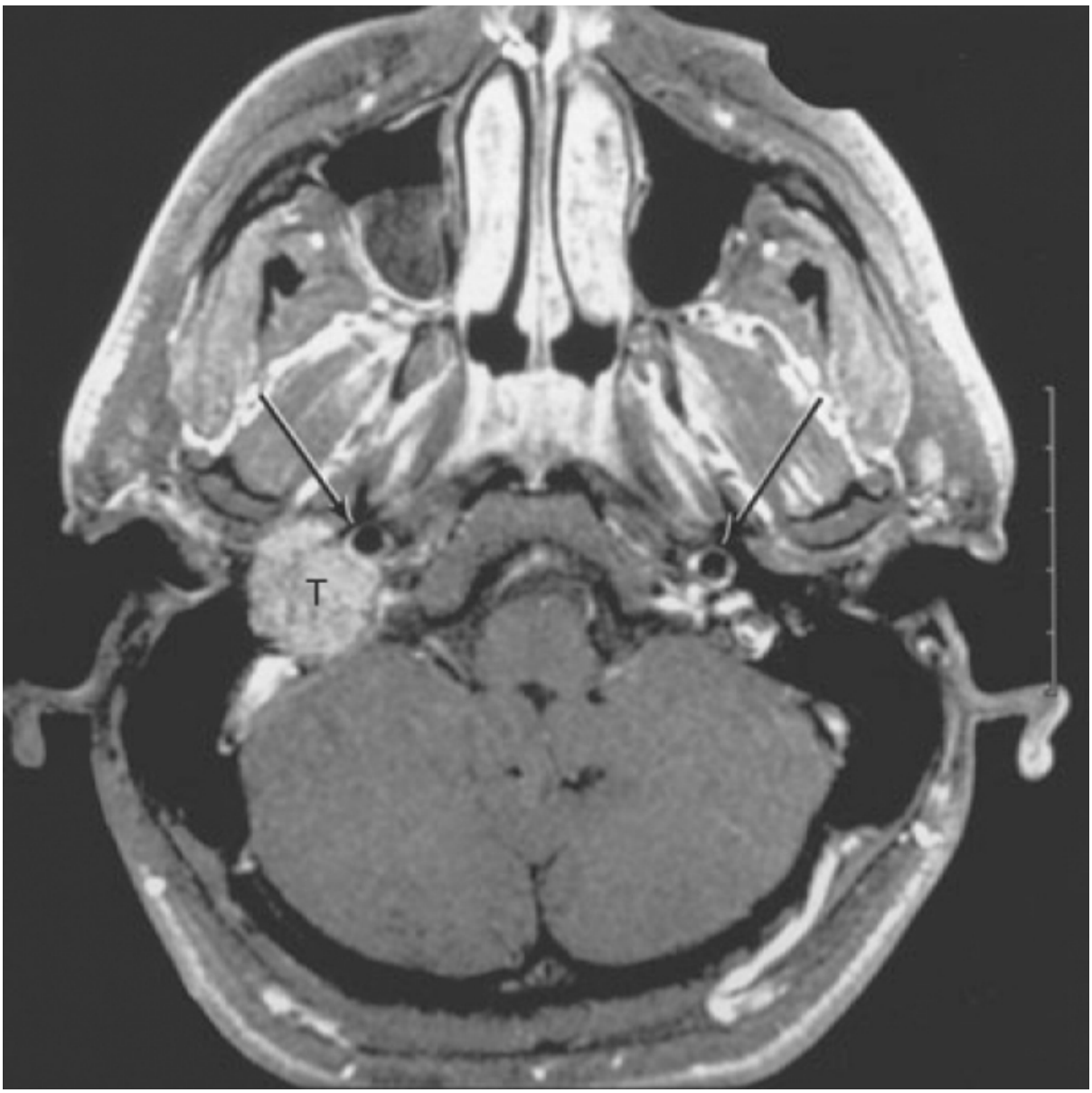

Figure 125-3 T1-weighted magnetic resonance imaging (MRI) scan with contrast showing a glomus tumor (T) occupying the right jugular fossa extending to the skull base. Note the internal carotid arteries *(arrows)*.

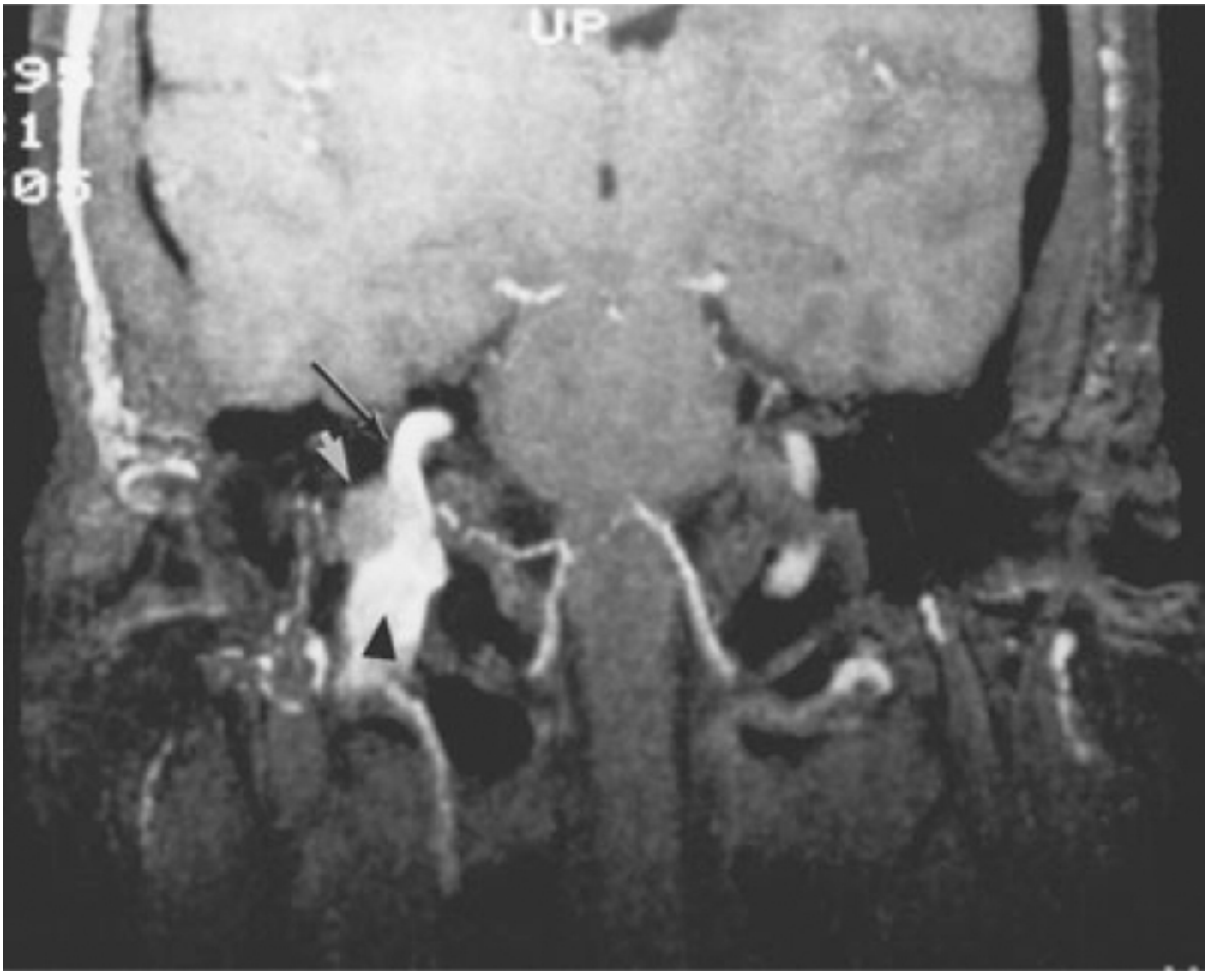

Figure 125-4 Magnetic resonance angiography (MRA) scan showing a tumor *(white arrow)* in the right skull base compressing the vertical portion of the carotid artery *(black arrow)*. The carotid artery and jugular vein are superimposed *(black arrowhead)*.

Until the techniques and interpretation of MRA become consistently reliable and comprehensively informative, four-vessel cerebral angiography remains necessary. Angiography should be obtained when CT imaging identifies a large tumor or when carotid artery involvement is suspected or identified. The data gained from this study provide critical information needed to institute a management plan. The location and extent of tumor are identified by the vascular blush. The four-vessel study also identifies other paragangliomas that may be present in the head and neck area (Fig. 125-5). Patients with multiple paragangliomas have a higher incidence of similar tumors being identified in other family members.

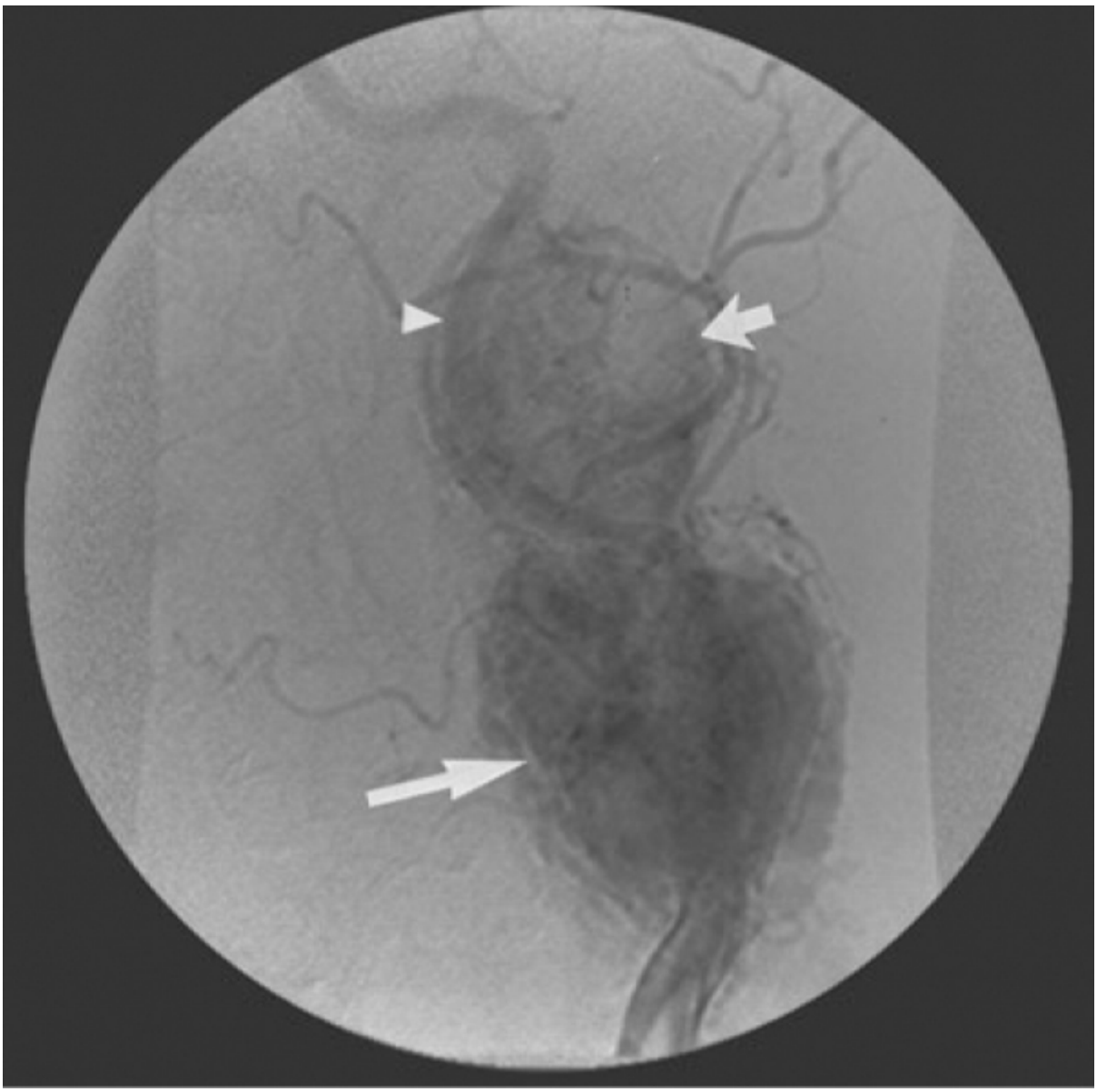

Figure 125-5 Left common carotid arteriogram of a patient with contralateral glomus jugulare demonstrating carotid body *(large arrow)* and glomus vagale *(small arrow)* tumors. The internal carotid artery is anteromedially displaced *(arrowhead).*

An effective method of screening these individuals is with a contrast-enhanced CT scan of the temporal bone (skull base) and neck, down to the level of the carotid bifurcation. It is imperative to know whether multiple paragangliomas or carotid body tumors are present on the ipsilateral or contralateral side in order to avoid the potential devastating complications related to bilateral lower cranial nerve compromise. Arteriography demonstrates the blood supply to the lesion. The major blood supply of glomus jugulare tumors is from the ascending pharyngeal artery. When a significant feeding vessel is identified, selective angiography and embolization using polyvinyl alcohol or Gelfoam is performed. Large lesions (glomus jugulare) are routinely embolized 1 to 2 days before surgical resection (Fig. 125-6).

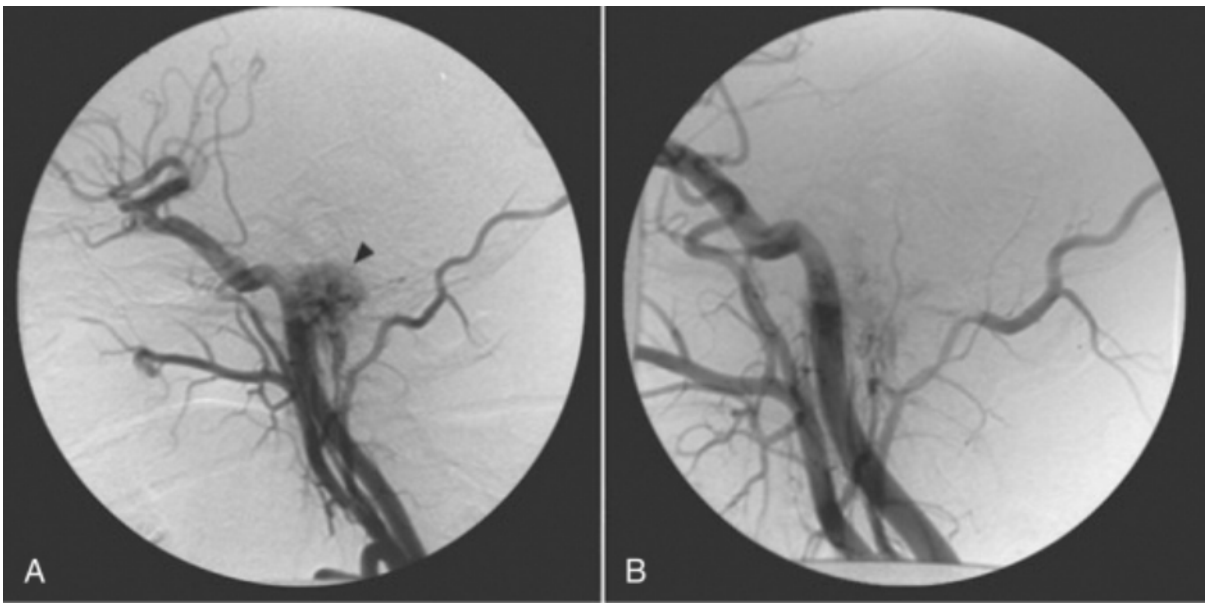

Figure 125-6 **A,** Preembolization arteriogram revealing right glomus jugulare tumor with characteristic blush *(arrowhead).* **B,** Postembolization study confirms effective occlusion of major feeding vessels. Tumor blush is significantly diminished.

Based on CT scan findings demonstrating extensive involvement, intraoperative carotid artery injury, repair, or resection may be anticipated. In this situation, at the time of the arteriogram, a balloon catheter is also used to functionally test the competency of the contralateral cerebral blood supply and circle of Willis. Balloon test occlusion (BTO) of the ipsilateral internal carotid artery (ICA) helps determine whether the patient can tolerate carotid artery occlusion or sacrifice. Xenon blood flow studies during BTO provide an objective and quantitative measure of cerebral blood flow. This information helps the surgical team determine whether carotid artery sacrifice will be tolerated. Failure to successfully tolerate BTO or if inadequate blood flow is demonstrated indicates that carotid artery repair or bypass will be necessary if injury or sacrifice occurs. Other members of the skull base team (neurosurgeons, vascular surgeons) are alerted if intracranial tumor is present or vascular repair is likely.

Finally, the venous phase of the arteriogram also reveals whether the sigmoid sinus is patent or occluded, demonstrates other venous outflow pathways, and indicates the status of the contralateral sigmoid sinus and jugular vein (Fig. 125-7). If no venous obstruction is shown, then the venous drainage of the dominant side is also demonstrated. The relationship of the ipsilateral vein of Labbé to the transverse sinus should be defined. Although tumor does not usually extend this far proximal in the venous drainage system, this information should be confirmed.

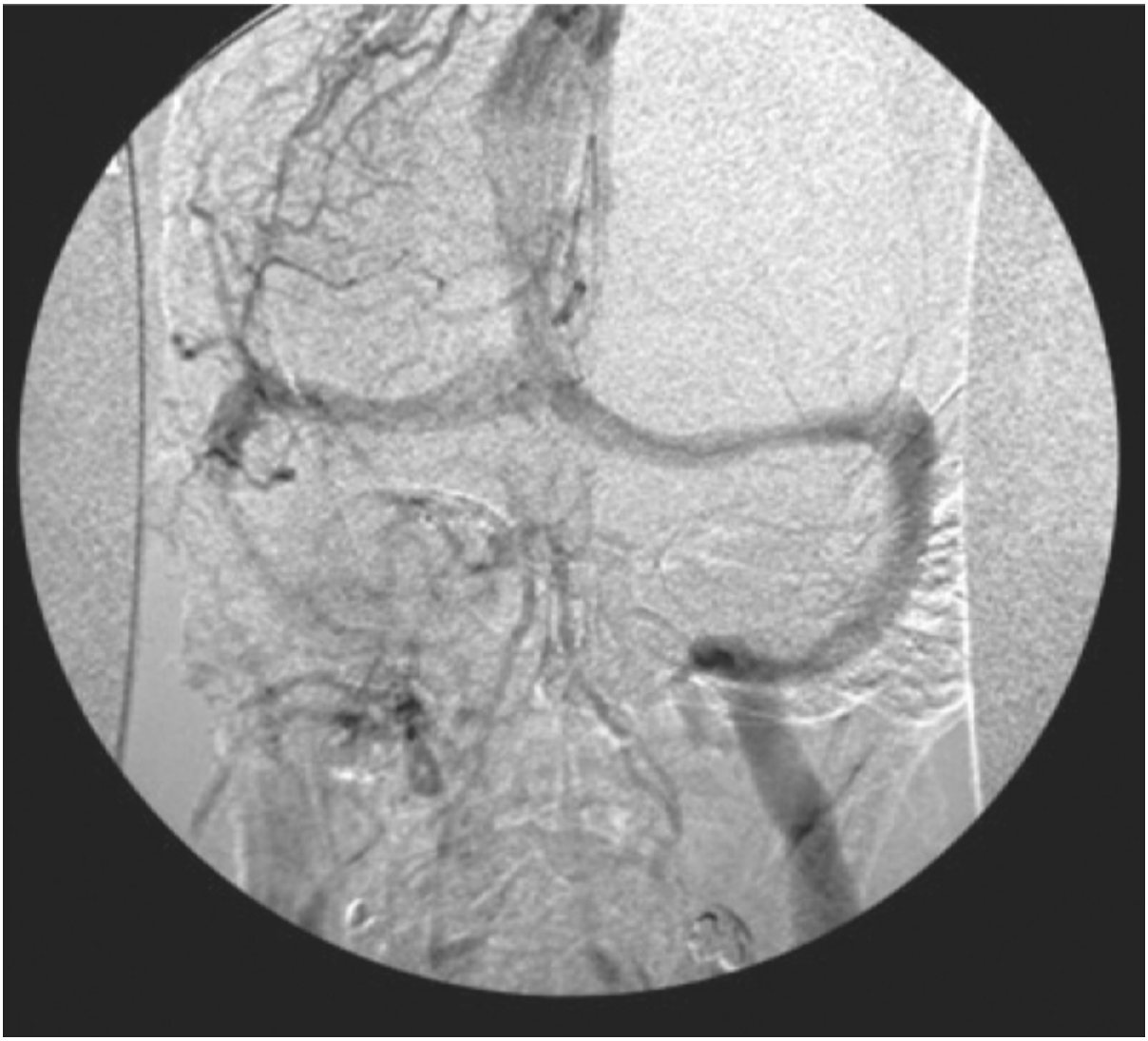

Figure 125-7 Venous phase of a carotid arteriogram showing marked attenuation of right sigmoid sinus and jugular vein flow with patency of the contralateral side.

Large glomus jugulare tumors are more likely to encroach on the ICA and the lower cranial nerves. The option of subtotal resection of a large tumor could be considered if cranial nerve function is normal preoperatively and compromise or sacrifice is anticipated to achieve complete resection. Determination is made based on imaging, assessing whether the pars nervosa (medial aspect of the jugular bulb) is involved. Intradural extension of tumor in this area would define the need for resection of the entire jugular bulb and pars nervosa. This approach is planned preoperatively based on the patient's preference for maintaining normal physiologic function of phonation, respiration, and swallowing. Similar strategy for planned subtotal resection can be given if the carotid artery is involved.

Perioperative systemic antibiotics are not given for glomus tumors isolated to the middle ear or mastoid space. Prophylaxis is provided if cerebrospinal fluid (CSF) exposure is anticipated. Patients with large lesions (glomus jugulare) should have blood held for type and screen. Autologous donation for potential transfusion can be planned in advance.

The importance of a comprehensive and understandable informed consent cannot be overemphasized. It is most important to discuss with the patient and family the location of the tumor and its relationship to the surrounding vascular, dural, and neural structures. The treatment options of observation, surgical removal, or radiation therapy are reviewed. The risks, benefits, and outcomes are explained. The discussion must be delivered to assure that the patient understands the significance of potential loss of lower cranial nerve function. The potential need for additional surgical procedures to manage complications following tumor removal should be discussed. These include eye protection with a gold weight and possible lower lid tightening, tracheotomy, vocal cord medialization procedures, gastrostomy tube, or CSF lumbar drainage. Delivery of a comprehensive informed consent should be documented in the patient's medical record.

STEREOTACTIC RADIOSURGERY

Fractionated XRT has been the conventional method for radiation treatment. Newer techniques using a frame-based linear accelerator, gamma radiation from cobalt delivered through the gamma knife, or photon beam from a robot-mounted mini-linear accelerator such as the CyberKnife are available for single or multiple focused treatments (fractions) providing submillimeter accuracy in treatment delivery. This author prefers the CyberKnife system for various reasons. It is a robotic-assisted mini-linear accelerator delivering 6 megavolts of photon beams with 6 additional degrees of freedom compared with conventional gantry-mounted accelerator (Fig. 125-8A). It is a frameless localization and delivery system that avoids the minor morbidity of placing a head frame on the patient, making it ideal for fractionated treatment. More important, the system allows for treatment of tumors that extend below the skull base, such as glomus jugulare tumors that grow inferiorly within the jugular vein or glomus vagale tumors located high in the neck. The gamma knife system has its field of delivery limited at the skull base.

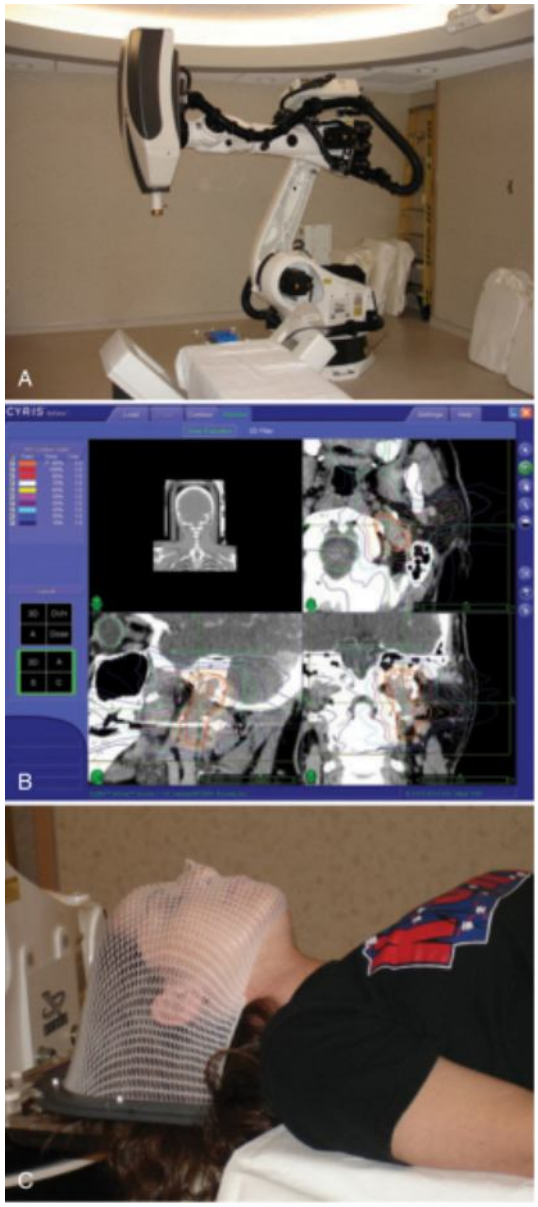

Figure 125-8 **A,** CyberKnife robotic radiosurgery system manufactured by Accuray, Sunnyvale, California. **B,** Contour and isodose treatment plan for a left jugular paraganglioma extending into the upper neck. **C,** Patient positioned on the CyberKnife treatment table secured by Aquaplast mask.

A variety of imaging modalities such CT, positron emission tomography, and MRI can be fused and displayed by overlapping the images. The tumor map is contoured, outlining the field of interest using information from the fused scan, although treatment is based on CT confirmation. The soft tissue signal from the MRI scan allows the tumor to be outlined. The bony anatomy of the temporal bone is also used to verify the location of the tumor and identifies other important structures to be protected from the full dose of delivered radiation. These are called *critical structures* and serve as avoidance structures that can be preferentially protected during treatment. At our institution, contouring the tumor is done both by the surgeon and radiation oncologist. The planned treatment is

then created and reviewed with the radiation physicist and ultimately approved by the radiation oncologist. The tumor is radiated with 18 or 21 Gy to the 80% isodose line in three fractions. The dose at the margin of the tumor is usually close to the prescription dose of 18 or 21 Gy, unless there is a critical structure immediately adjacent to the tumor. Minimizing treatment to a critical structure would lower the dose at the margin of the tumor. The maximum tumor dose is 22.5 or 26.25 Gy at the 100% dose (see Fig. 125-8B). A thermoplastic mask (Aquaplast) is created at the time of simulation and tightly immobilizes the patient's head and neck, and offers similar degrees of accuracy to the more invasive gamma knife frame. Each of three treatments is given for approximately 30 minutes delivered every other day. No sedation, intravenous fluids, or hospitalization are necessary (see Fig. 125-8C).

Radiation therapy may incur long-term complications such as osteonecrosis, chronic otitis externa, breakdown of skin, eustachian tube dysfunction, and the potential development of malignant tumor. Acute cranial nerve dysfunction (hearing loss, imbalance or vertigo, facial paresis, vocal fold weakness, dysphagia) can occur and may be transient.

SURGICAL TECHNIQUES

Unless the lesion is small and clearly limited to the middle ear promontory, all procedures are performed with the patient under general endotracheal anesthesia using inhalation and intravenous techniques. Muscle relaxants are avoided, so that intraoperative monitoring of cranial nerves VII, IX, X, XI, or XII can be performed.

Glomus Tympanicum Tumors

The surgical approach for glomus tympanicum tumors is dictated by the size of the lesion. Small tumors limited to the mesotympanum and hypotympanum without involvement of the jugular bulb are removed through a transcanal approach. Larger lesions usually require a postauricular approach to provide greater exposure. A postauricular approach is also necessary when the inferior annulus is not adequately visualized through the external auditory meatus. This is often due to a prominent bulge from the anterior wall or a high floor of the ear canal.

Under the operating microscope, local anesthesia is injected as a four-quadrant block into the skin of the external auditory meatus. The same local solution is infiltrated into the skin of the postauricular sulcus. Lateral canal incisions at 12 o'clock and 6 o'clock are made and connected with a vertical incision approximately 5 mm from the annulus. The posterior canal skin flap that is based laterally is back-elevated within the canal. The flap is also known as the *vascular strip.* A postauricular incision is made and carried down to cortical mastoid bone. The spine of Henle is a good landmark for the slope of the posterior bony ear canal. The postauricular and canal flap incisions are connected and the ear is retracted anteriorly with a self-retaining retractor.

A tympanomeatal flap based on the anterior third of the medial ear canal is created. This incision extends from the 12-o'clock to the 4-o'clock position in a right ear or from the 12-o'clock to the 8-o'clock position in a left ear. The incision should preserve 5 mm of skin lateral to the annulus. The tympanomeatal flap is elevated, preserving the chorda tympani nerve. The margins of the tumor are dissected and elevated to isolate the vascular supply to the lesion (Fig. 125-9). Excision of a tumor that involves the hypotympanum requires greater exposure by removal of the inferior tympanic ring with a microear drill (Fig. 125-10). When the inferior margin of the tumor cannot be visualized, the tympanic bone can be taken down to the floor of the hypotympanum. If tumor infiltrates the trabeculated air cells of the hypotympanum, these cells and tumor are removed using a diamond burr. The use of microbipolar forceps facilitates hemostasis during removal of the tumor (Fig. 125-11). Often a pedicled vessel feeding the tumor can be found and should  be electrocoagulated (see Video 125-1).

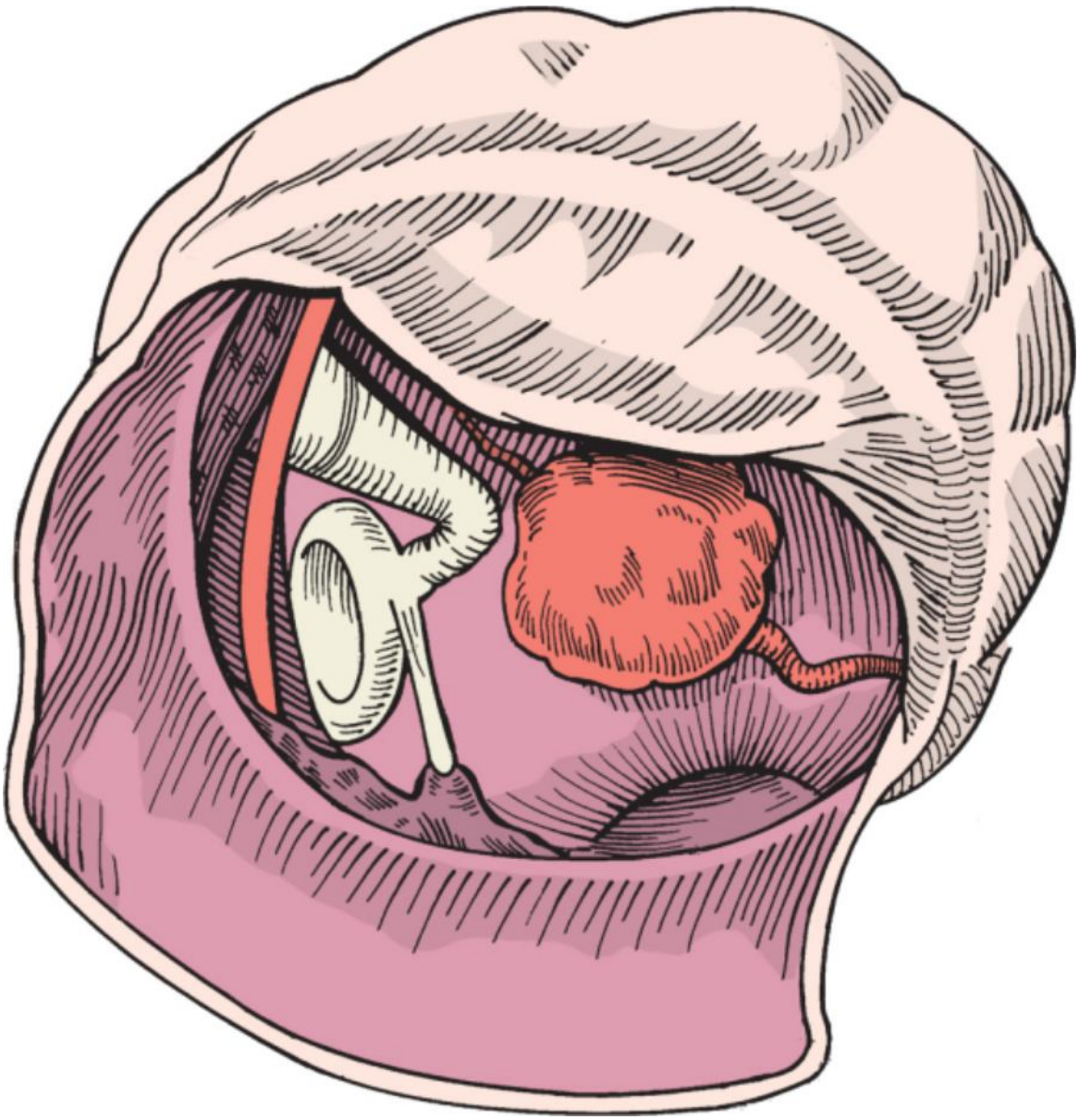

Figure 125-9 Transtympanic approach to a glomus tumor isolated within the middle ear.

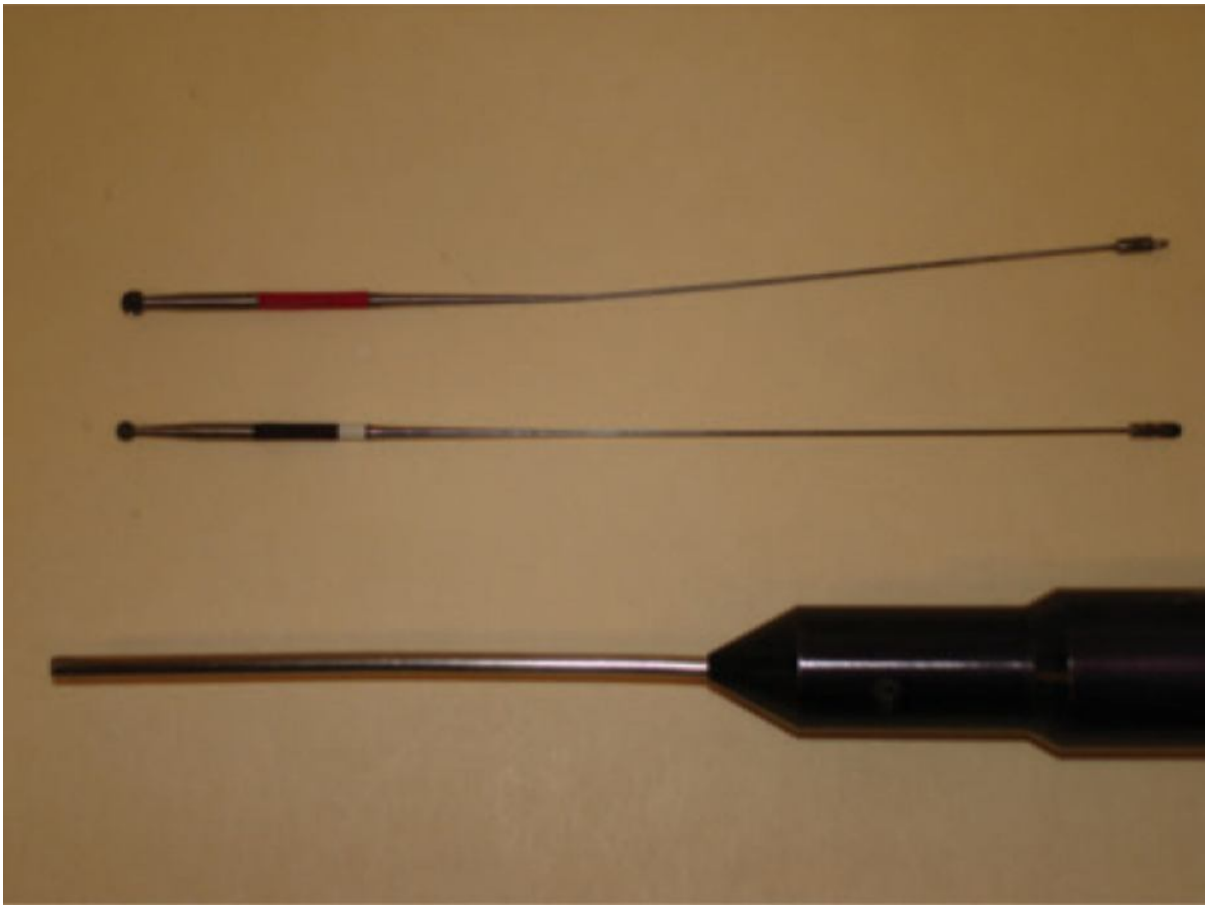

Figure 125-10 A microear drill is used to access narrow or small areas. Drill bits are available as cutting and diamond burrs.

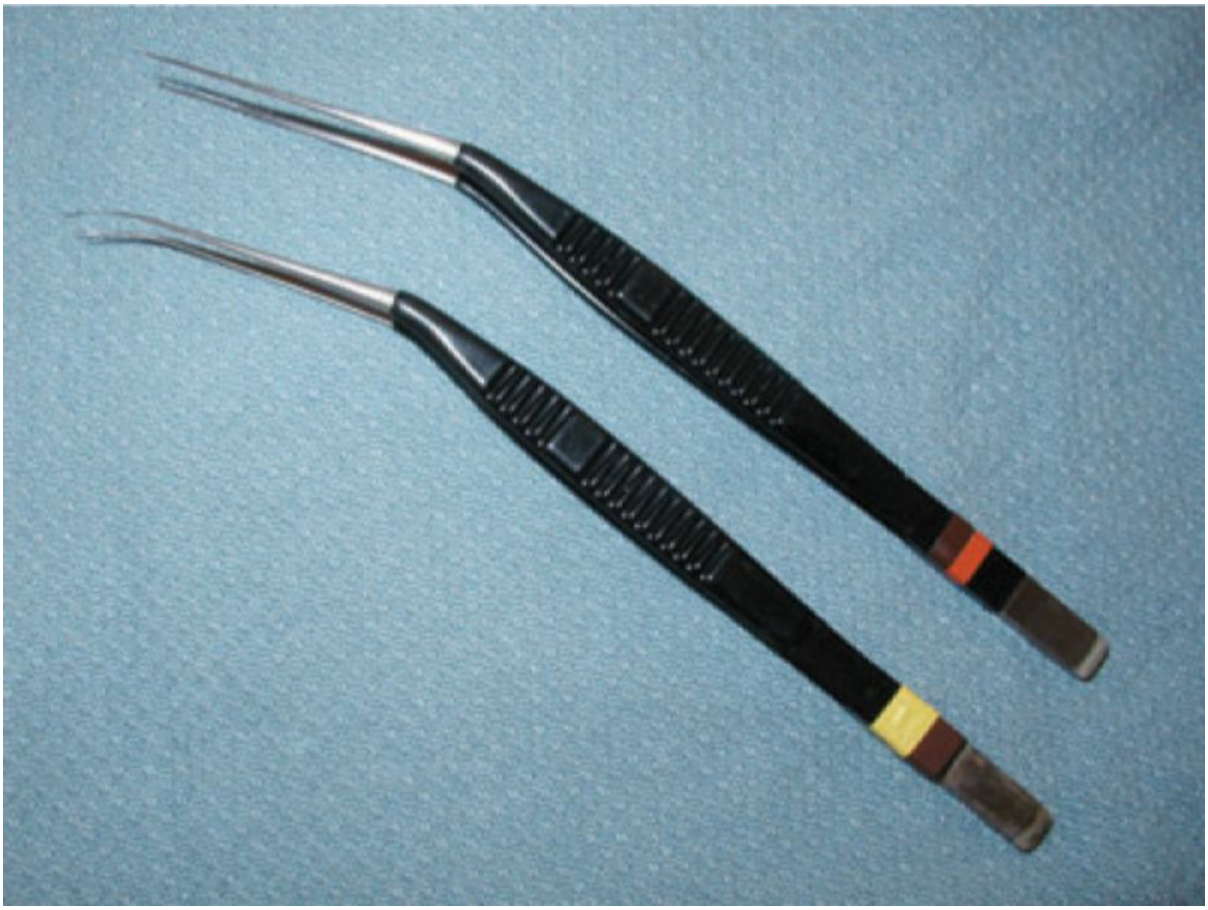

Figure 125-11 Microbipolar forceps facilitate cautery in the middle ear and can be used through an ear speculum.

The vascular supply from the tympanic plexus is isolated and cauterized. If this is not possible, then the tumor is coagulated with the bipolar cautery and removed, and subsequent hemostasis is obtained. Small cotton balls soaked with 1 : 1000 epinephrine are useful to temporarily compress sites of bleeding during tumor removal from the middle ear. Oxidized cellulose compressed over the tumor is another effective method for obtaining temporary hemostasis. After the tumor is removed and hemostasis is obtained, the tympanomeatal flap is returned to its anatomic position. Depending on the extent of hypotympanic exposure, it may be necessary to support the inferior aspect of the flap with a medially placed temporalis fascia graft or Gelfoam placed in the hypotympanum. A silk sleeve supported with cotton balls or Gelfoam is used for a canal dressing.

Jugulotympanic Glomus Tumors

Jugulotympanic tumors that involve the middle ear, jugular bulb, and hypotympanic air cells are usually too large to remove through a transcanal approach. The tumor may erode bone near the carotid artery (Fisch type C_1, Glasscock-Jackson tympanicum type IV, or jugulare type I). It is usually not necessary to develop the exposure of the infratemporal fossa (ITF) and upper neck required for large glomus jugulare tumors. The initial transcortical mastoid approach is similar to that described for mastoidectomy (see Chapter 115). An incision is made 1 cm posterior to the postauricular crease. The skin, subcutaneous tissue, and fibroperiosteal layer over the mastoid cortex are incised and elevated from a posterior and anterior direction. Dissection is taken to the spine of Henle and the posterior aspect of the external auditory meatus. A transcortical mastoidectomy is performed with cutting burrs and suction irrigation. Landmarks to be identified are the antrum, mastoid tegmen, horizontal semicircular canal, fossa incudus, and short process of the incus. The sigmoid sinus is skeletonized to provide maximal exposure between the sigmoid sinus and the vertical portion of the facial nerve. The tumor may be encountered in the infralabyrinthine retrofacial air cells. Following an imaginary line drawn along the body of the incus and short process, the facial recess (posterior tympanotomy) is developed with cutting and then diamond burrs. Extending the facial recess toward the tympanic ring greatly improves exposure to the posterior mesotympanum and hypotympanum. It is necessary to sacrifice the chorda tympani nerve with an extended facial recess approach.

Attention is then turned to the external auditory canal (EAC). A tympanomeatal flap is developed from the 12-o'clock to the 5-o'clock position (for a right ear). Elevation of the tympanic membrane exposes the middle ear portion of the tumor. At this point, the surgeon must determine whether tumor can be removed with the given exposure. If extensive tumor involves the tympanic bone and hypotympanum, the infratympanic facial recess approach is extended (Fig. 125-12).[13] It may be necessary to create a “fallopian bridge” by skeletonizing the distal vertical portion of the facial nerve in order to remove all tumor. The margins of the tumor are dissected and defined. Hemostasis and tumor shrinkage are obtained with bipolar or microbipolar forceps. Tumor is removed with cup forceps (see Video 125-2). A cotton ball with epinephrine is used to temporarily control bleeding (see Video 125-3). If the exposure is still inadequate, the posterior canal wall may need to be taken down. If the tumor is more extensive than anticipated, management follows the description for glomus jugulare tumors (see Glomus Jugulare Tumors [Fisch types C_1 and C_2] later in this chapter).

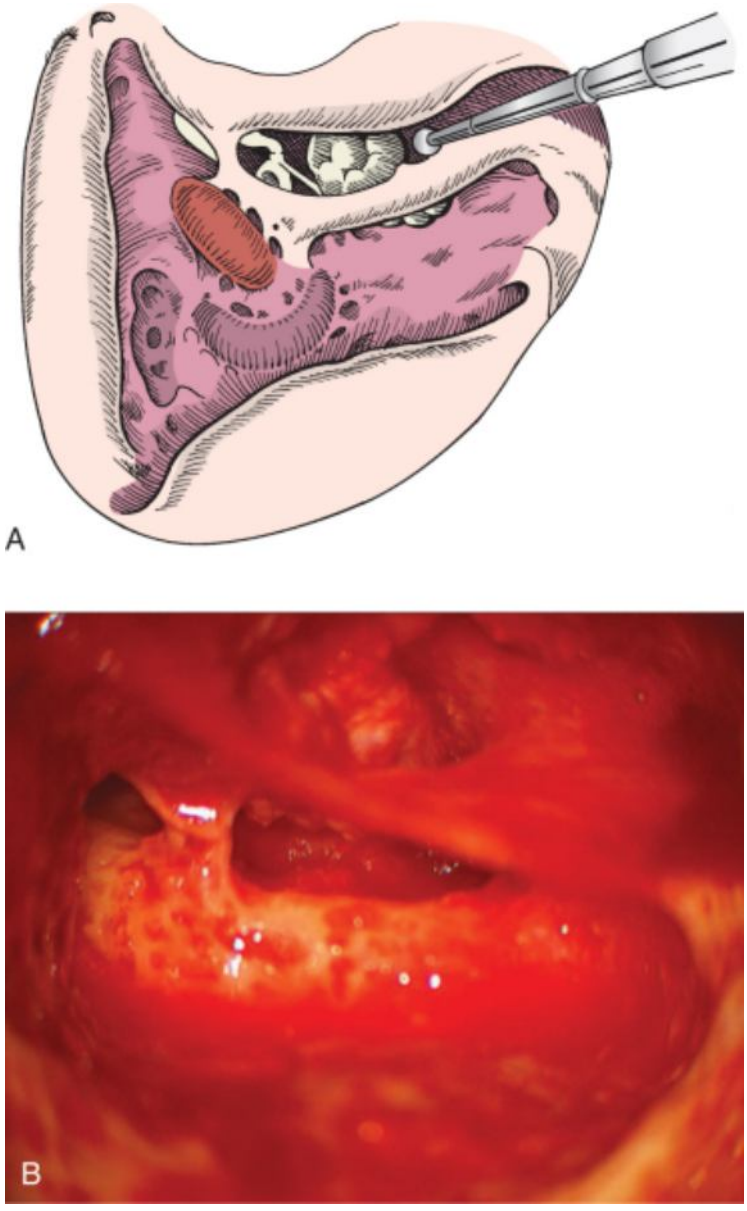

Figure 125-12 Access to the glomus tumor in the middle ear and hypotympanic space is achieved by extending the facial recess approach. Opening the retrofacial infralabyrinthine air cells provides exposure of the posterior margins of the tumor.

Whether the canal wall is left up or taken down, tympanic membrane and ossicular chain reconstruction are undertaken as needed. When the incus is removed in order to facilitate tumor removal from the area around the stapes, an incus interpositioning is performed. If the malleus is absent, then stapes augmentation is achieved with either a partial ossicular replacement prosthesis or an ossicular remnant of the incus body or malleus head (see Chapter 114).

If the posterior canal wall is intact, the posterior canal skin flap (vascular strip or Koerner's flap) is replaced along the posterior bony wall and the ear is returned to its anatomic position. A pack is placed in the external ear canal. Options for this include a silk sleeve, rosebud packing, Gelfoam, continuous gauze, or ointment. The author's group's preference is to use a silk sleeve bolstered with antibiotic-impregnated cotton balls.

If the posterior canal wall is taken down, a medial rosebud packing is placed (see Chapter 113). (Gelfoam may also be used to secure the tympanic membrane or graft.) The pinna is then returned to its anatomic position, placing the posterior canal wall skin flap into the mastoid cavity. A packing of antibiotic-impregnated continuous gauze is placed through the external auditory meatus and into the mastoid cavity. For both approaches, the postauricular incision is closed using 4-0 Nurolon suture. A sterile mastoid dressing is applied.

Glomus Jugulare Tumors (Fisch Types **C**$_1$ **and C**$_2$)

Skin Incision

Preoperative determination of tumor extension provides information for the operative approach. Jugulare tumors with limited carotid involvement (Fisch types C_1 and C_2) may permit preservation of the posterior bony canal wall. A postauricular mastoidectomy incision located 3 cm posterior to the pinna extends over the lower border of the mastoid tip, through an upper neck skin crease, toward the greater cornu of the hyoid bone (Fig. 125-13). A second, deeper incision positioned 1 cm anteriorly through the musculoperiosteum layer over the mastoid cortex creates a “stepped” flap that facilitates closure at the end of the procedure. The musculoperiosteum layer cortex is elevated anteriorly toward the EAC. The neck incision goes through the subcutaneous tissue and platysma muscle, with preservation of the greater auricular nerve, which is harvested to its maximal length.

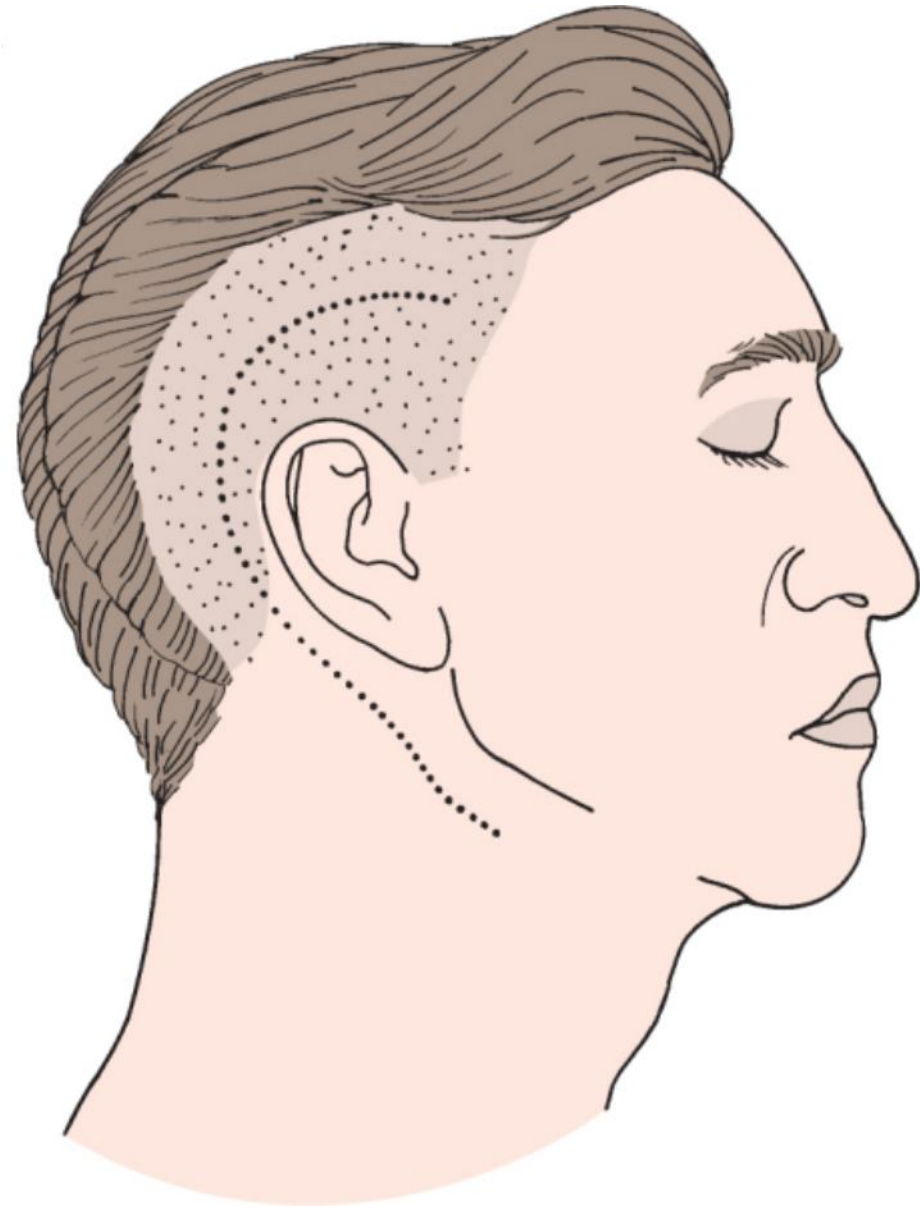

Figure 125-13 Skin incision for large glomus tumor when the carotid artery, jugular vein, and lower cranial nerves must be identified.

Neck Dissection

Dissection in the upper neck provides identification and isolation of cranial nerves X, XI, and XII and the great vessels. The neck dissection isolates the tail of the parotid from the sternocleidomastoid muscle (SCM). The spinal accessory nerve (CN XI) is identified at the anterior border of the SCM. Using cutting cautery, the SCM is released from its mastoid attachment and retracted posteriorly. The 11th cranial nerve is followed superiorly toward the jugular vein, identifying the 10th and 12th cranial nerves and the common, internal, and external carotid arteries. The tail of the parotid is elevated superiorly and anteriorly in order to identify the posterior belly of the digastric muscle. The posterior belly of the digastric muscle is dissected from the digastric groove and retracted superiorly in order to expose the inferior contents of the jugular fossa and stylomastoid foramen (Fig. 125-14). The jugular vein, common carotid artery, ICA, and cranial nerves are isolated and tagged with appropriately colored vascular loops. The ascending pharyngeal and occipital arteries are usually encountered and need to be ligated. The lower

cranial nerves, jugular vein, and carotid artery are further dissected superiorly to the inferior aspect of the tumor.

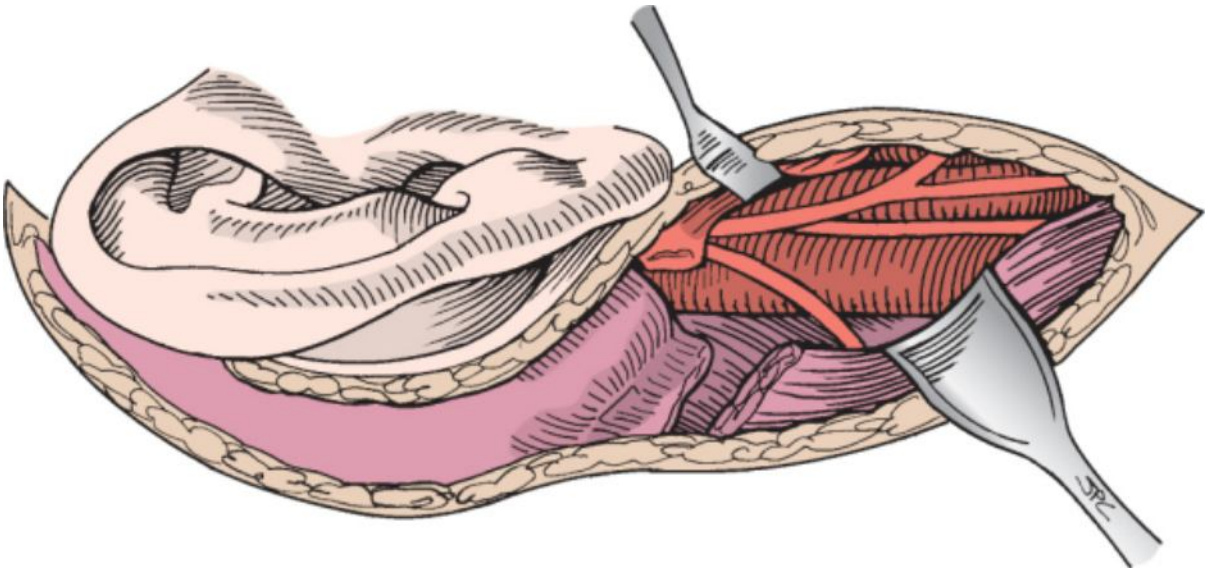

Figure 125-14 The postauricular incision is carried into the neck. The carotid artery, jugular vein, and lower cranial nerves are isolated. The sternocleidomastoid and digastric muscles are retracted.

Transtemporal Dissection

A transcortical mastoidectomy is performed. The pertinent landmarks to be identified are the horizontal semicircular canal, incus, mastoid tegmen, sinodural angle, and sigmoid sinus. Management of the posterior bony canal wall depends on the extent of the tumor. Tumors with extensive involvement of the carotid artery (Fisch types C_3 and C_4), extension into the clivus, invasion into the cochlea (anacousis), or destruction of lateral perifacial air cells often require a canal wall down procedure, along with transposition of the facial nerve. Tumors that are less extensive may be removed through an extended facial recess approach. Similar to the approach for a small jugulotympanic glomus tumor, the extended facial recess approach begins superiorly at the chorda facial recess. Using the vertical portion of the facial nerve as the medial dissection limit, the tympanic bone is drilled from a superior-to-inferior-to-anterior direction. The chorda tympanic nerve is sacrificed. The lateral limit of the extended facial recess is the tympanic ring. Retrofacial and infralabyrinthine air cells and bone are removed to the stylomastoid foramen. The facial nerve is skeletonized, leaving eggshell-thin coverage surrounding a “fallopian bridge” (see Fig. 125-12).

The digastric ridge is delineated within the mastoid cavity. Dissection is continued anteriorly toward the stylomastoid foramen. Thinning the bone to eggshell thickness along the lateral inferior aspect of the digastric ridge facilitates removal of the mastoid tip using a rongeur. It is necessary to expose the posterior fossa dura both posterior and anterior to the sigmoid sinus. Some bone is left over the proximal sigmoid sinus near the sinodural angle (above Donaldson's line). The sigmoid sinus is carefully dissected free from the overlying roof of bone. This exposure provides access to extraluminal packing or suture ligation of the sinus. Dissection with the drill is continued along the sigmoid sinus toward the jugular bulb. All bone must be removed from this area. The tumor is now removed.

This approach provides exposure of, but limited controlled access to, the infratemporal carotid artery (Fig. 125-15). Preoperative imaging determines whether distal and proximal control of the petrous carotid artery is necessary to safely remove all tumor. If the tumor does not erode the bone surrounding the carotid artery, this canal wall-up, extended facial recess approach provides appropriate exposure.

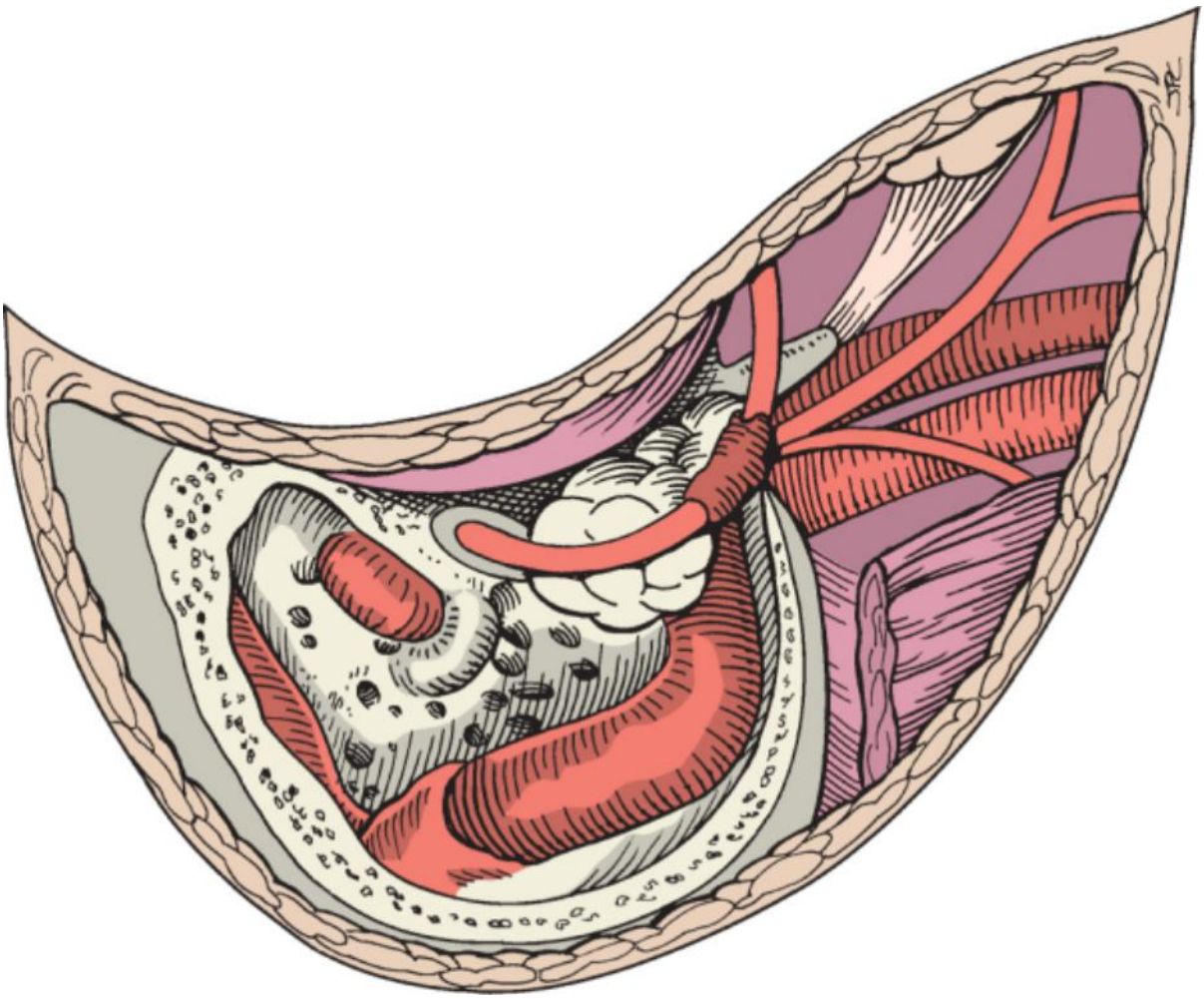

Figure 125-15 Transmastoid tumor dissection isolating the sigmoid sinus, vertical facial nerve along the extended facial recess, digastric ridge, neck vessels, and lower cranial nerves.

Tumor Isolation and Removal

Venous bleeding may occur from the sigmoid sinus, jugular vein, condylar vein, or inferior petrosal sinus. The first two are controlled at the onset of tumor removal. The sigmoid sinus may be occluded by either extraluminal packing or ligation. Extraluminal occlusion is performed with oxidized cellulose that is packed under a retained shelf of bone covering the proximal sigmoid sinus. If this is not successful or feasible, ligation of the sinus is performed by making small openings in the dura anterior and posterior to the sigmoid sinus. An aneurysm needle is passed from posterior to anterior deep to the sigmoid sinus. The needle is blindly passed medial to, but hugging, the sigmoid sinus, avoiding injury to the intracranial contents (cerebellum). A long 2-0 silk ligature is passed through the aneurysm, needle up to its mid length, and the aneurysm needle is withdrawn. The suture is then cut in half, so as to leave two separate ligatures (Fig. 125-16). A small piece of muscle is harvested and placed over the sigmoid sinus, and the first suture is tied and secured. The second suture is similarly tied, thus providing double ligation and occlusion of the sigmoid sinus.

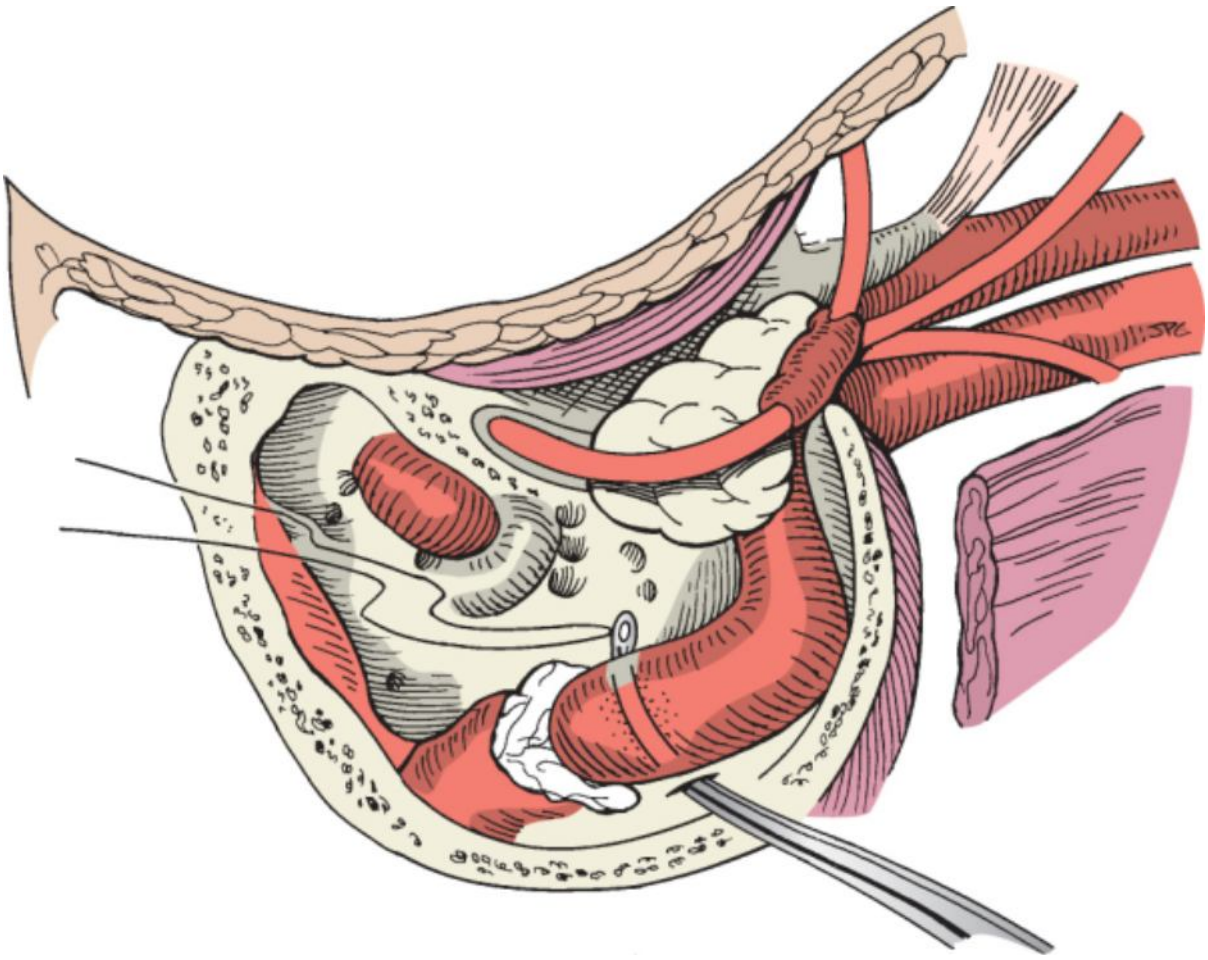

Figure 125-16 The extended facial recess approach exposes the tumor in the hypotympanum. The vertical portion of the facial nerve remains in place. The proximal sigmoid sinus can be isolated by extraluminal packing or suture ligation.

The jugular vein in the neck is doubly ligated at this time. The proximal stump of the jugular vein in the neck is dissected superiorly toward the jugular bulb and fossa (Fig. 125-17). Care is taken to preserve the spinal accessory nerve. It is usually necessary to pass the proximal stump of the jugular vein medial to the spinal accessory nerve in order to facilitate the superior dissection. The distal extratemporal ICA should be in direct view as the jugular vein is dissected toward the jugular bulb. The styloid process and its muscular attachments may need to be removed in order to gain access to the skull base.

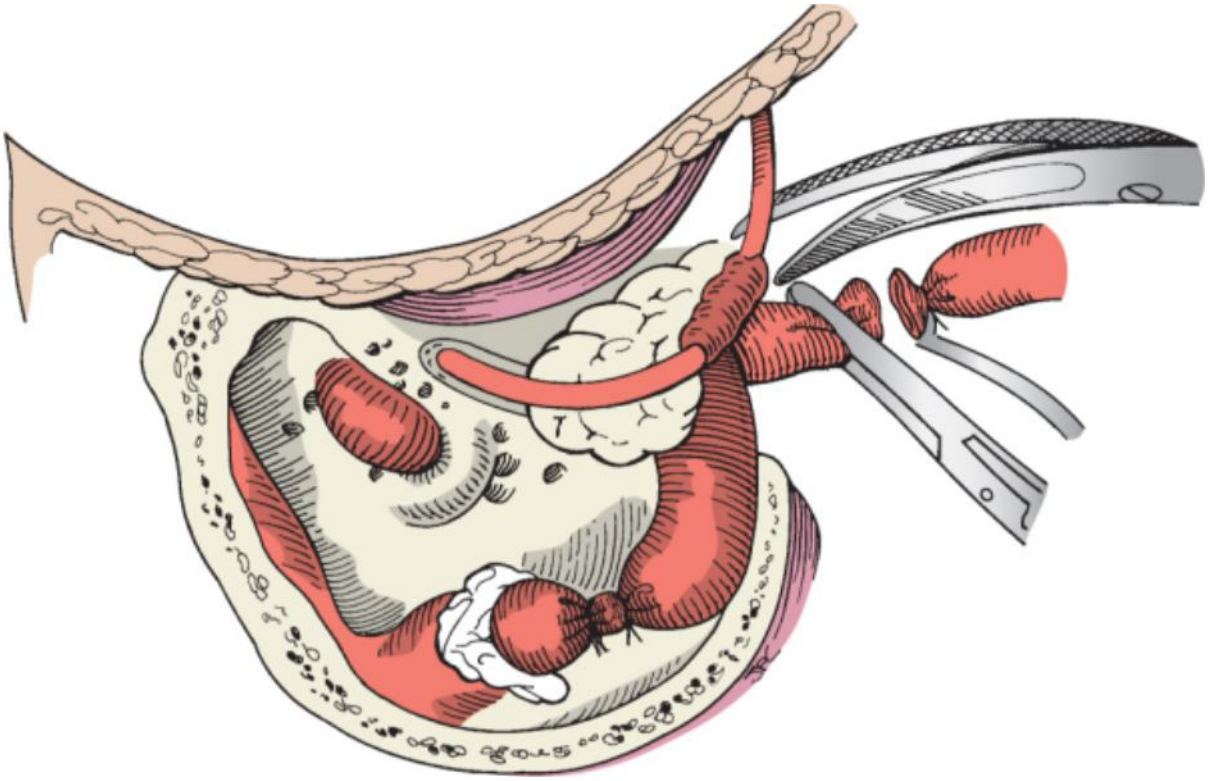

Figure 125-17 Proximal control of the sigmoid sinus is achieved. Surgicel packing can also be used to occlude the proximal sigmoid sinus. The jugular vein is divided, and the proximal segment is dissected into the jugular fossa. The anterior plane of dissection is along the carotid artery, which is not shown (see Fig. 125-16).

The sigmoid sinus is opened distal to the ligatures in order to inspect the intraluminal contents. Usually, the medial wall of the sigmoid sinus is not invaded by tumor unless there is intracranial extension. The inferior aspect of the sigmoid sinus is incised in a longitudinal direction toward the jugular bulb. The tumor is sequentially isolated from a posterior-to-anterior direction, dissecting the superior, lateral, and inferior walls of the sigmoid sinus from the medial wall. Bipolar coagulation is usually needed to obtain hemostasis and shrink the tumor. The lateral and superior aspect of the jugular bulb are dissected toward the superior extent of the tumor. Tumor often occludes the jugular bulb. However, once the tumor is removed from this area, brisk bleeding may be encountered from the multiple orifices of the inferior petrosal sinus and condylar vein. The medial jugular fossa is packed with oxidized cellulose. This dissection is performed medial (deep) to the vertical portion of facial nerve and stylomastoid area. Care must be taken to avoid trauma or pressure to the medial (deep) surface of the facial nerve. Tumor often remains in the hypotympanum and middle ear. This is removed by working both medial and lateral to the facial nerve.

Certain intraoperative findings may necessitate a canal-wall-down procedure. The first is the need for increased exposure of tumor invading the carotid artery. Also, limited space in the retrofacial and infralabyrinthine areas may require transposition of the facial nerve in order to provide adequate exposure of the tumor. Reconstruction of the tympanic membrane and ossicular chain can still be performed as needed.

Unless the medial wall of the jugular bulb or sigmoid sinus is removed, CSF is not encountered. However, the dead space created by the removal of bone and tumor requires partial obliteration with abdominal fat. Packing of the ear canal is identical to that described previously. A Hemovac drain is placed deep to the neck skin flaps. The postauricular wound is closed in three layers, reapproximating the musculoperiosteum, subcutaneous tissue, and skin.

Large Glomus Jugulare Tumors (Fisch Type **C**$_2$ or Greater)

A large postauricular C-shaped incision begins 3 cm superior to the pinna. If it is known that an ITF dissection is necessary to gain access to the distal ICA, then the incision begins anterior and superior to the pterion. The incision extends posteriorly and inferiorly 3 to 4 cm posterior to the helical rim, continues inferiorly across the lower border of the mastoid tip, and through upper neck skin crease toward the greater cornu of the hyoid bone in anticipation of isolation of the great vessels and lower cranial nerves. This creates an anteriorly based flap, which will include the pinna. The superior incision and dissection are taken down to the level of the temporalis fascia.

The EAC is transected medial to the cartilaginous bony junction. The skin of the proximal canal is dissected away

from the underlying cartilaginous canal, and the cartilage is removed. This leaves a cuff of external auditory meatus skin that will be everted by placing superior and inferior sutures in the canal skin. The ends of these sutures are passed retrograde through the EAC. This facilitates eversion of the cuff of the external auditory meatus skin, which is reapproximated in a linear fashion with interrupted sutures of 4-0 Dexon (Fig. 125-18). A deep flap of fibroperiosteum is developed in order to create a second layer for closure of the external meatus. This 2.5- × 2.5-cm flap is based on the posterior aspect of the subcutaneous tissue of the EAC. It is sutured anteriorly to the subcutaneous tissue of the skin just deep to the tragus.

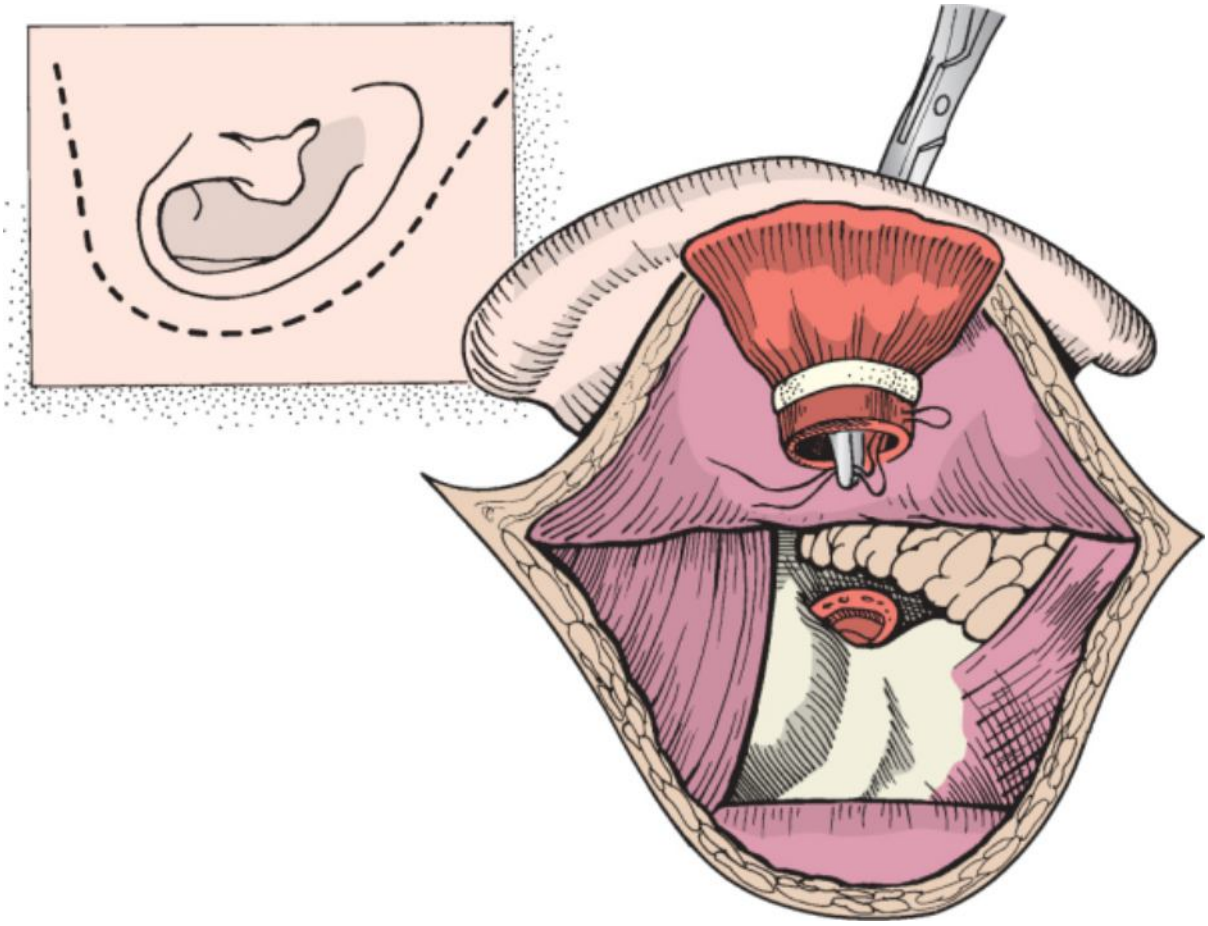

Figure 125-18 The ear canal is transected. A fibroperiosteal flap based on the posterior canal has been created. The ear canal skin is everted with a horizontal suture passed through the distal cuff of canal skin.
(Redrawn from Gantz BJ, Fisch U: Modified transotic approach to the cerebellopontine angle. Arch Otolaryngol 109:253, 1983. Copyright 1983, American Medical Association.)

Neck Dissection

The approach and dissection in the neck are identical to that previously described for smaller glomus jugulare tumors. The great vessels and cranial nerves are isolated and tagged with vessel loops. A moistened lap sponge is left in the wound during the infratemporal fossa dissection.

Infratemporal Fossa Dissection

Preoperative evaluation will determine the degree of involvement of the carotid artery or clivus. Extensive tumors involving the horizontal portion of the carotid artery and petrous apex require dissection of the ITF. This approach provides necessary exposure for control of the distal ICA. The upper limb of the skin incision must be extended toward the forehead, just superior to the pterion. The superior skin flap is elevated superficial to the temporalis fascia down to the level of the zygomatic arch. Care is taken to go medial to the deep cervical fascia in order to avoid injury to the temporal branches of the facial nerve. Dissection with electrocautery facilitates removal of the attachments of temporalis fascia to the arch and body of the zygoma. Similarly, the fascial attachment of the masseter muscle is removed from the inferior surface of the zygomatic arch, which is circumferentially isolated from surrounding tissue. Using the reciprocating saw, osteotomy cuts permit removal of the zygomatic arch. The periosteum at the origin of the temporalis muscle is cut, and the muscle is dissected from the temporal fossa.

The dissection is continued in a subperiosteal plane into the ITF (Fig. 125-19). Venous bleeding is often

encountered and controlled with bipolar coagulation and oxidized cellulose. Inferior and anterior retraction of the mandibular condyle widens the exposure. However, should complete exposure and control of the carotid artery be necessary, then resection of the mandibular condyle improves surgical access to the vertical segment of the carotid. The attachment of the pterygoid muscle to the neck of the condyle is divided with electrocoagulation. The condyle may be removed with a reciprocating saw. Dissection in the ITF is directed toward the foramen ovale (trigeminal nerve V3) and foramen spinosum (middle meningeal artery). Control of the carotid artery is obtained by performing an inferior frontotemporal craniotomy. A craniotomy is performed by placing burr holes through which the underlying dura is elevated and bone flap removed. Additional exposure of the petrous apex and carotid artery at the infratemporal skull base is accomplished with a dissecting drill and rongeurs. The horizontal segment of the petrous ICA is identified and isolated by this extradural middle fossa dissection. This approach facilitates the subtemporal ITF exposure of pertinent petrous artery landmarks: V3, greater superficial petrosal nerve, and middle meningeal artery.

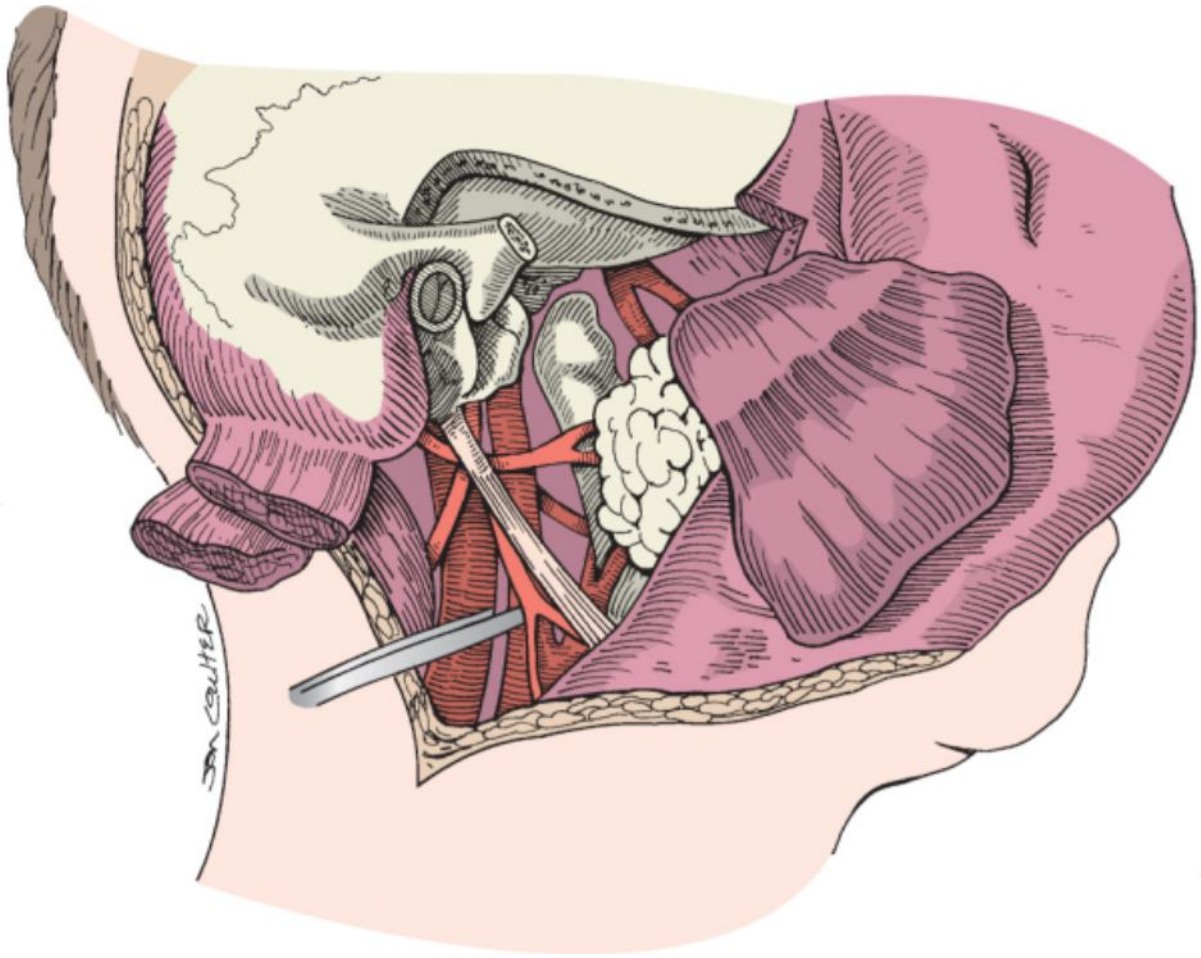

Figure 125-19 Removing the zygomatic arch permits mobilization of the temporalis muscle, exposing the infratemporal fossa. Soft tissue dissection isolating the extratemporal facial nerve, great vessels, and lower cranial nerves is demonstrated. The tumor has not been exposed in the temporal bone.

Transtemporal Dissection

Complete mastoidectomy is performed. The approach and dissection are identical to that previously described for smaller glomus jugulare tumors (Fisch type C_2 or greater). Exposure of the dura of the posterior fossa anterior and posterior to the sigmoid sinus must be performed in anticipation of opening the dura, possible resection of the dura, and removal of the tumor. Most tumors require complete exposure of the middle ear and necessitate removing the posterior canal wall along with the epithelium of the medial EAC and tympanic membrane. Reconstruction of the middle ear is usually not feasible owing to the extent of the lesion and the need to obliterate the eustachian tube to avoid CSF rhinorrhea. If function of the cochlea is to be preserved, the incudostapedial joint is separated through the facial recess approach. The ossicles lateral to the stapes and the tympanic membrane may be removed. The posterior bony canal wall is taken down to the level of the horizontal and vertical portion of the facial nerve. Similarly, the anterior canal wall must be removed to the level of the parotid gland and periosteum of the glenoid fossa. Tumor extending into the middle ear is encountered at this time. The vertical segment of the facial nerve interferes with access to large tumors filling the jugular fossa and invading the carotid artery. Facial nerve transposition is usually necessary to achieve optimal exposure. The facial nerve is decompressed from the

geniculate ganglion to the stylomastoid foramen with cutting and then diamond burrs. Favorable anatomy may permit more limited mobilization of the facial nerve from its second genu (Fig. 125-20).

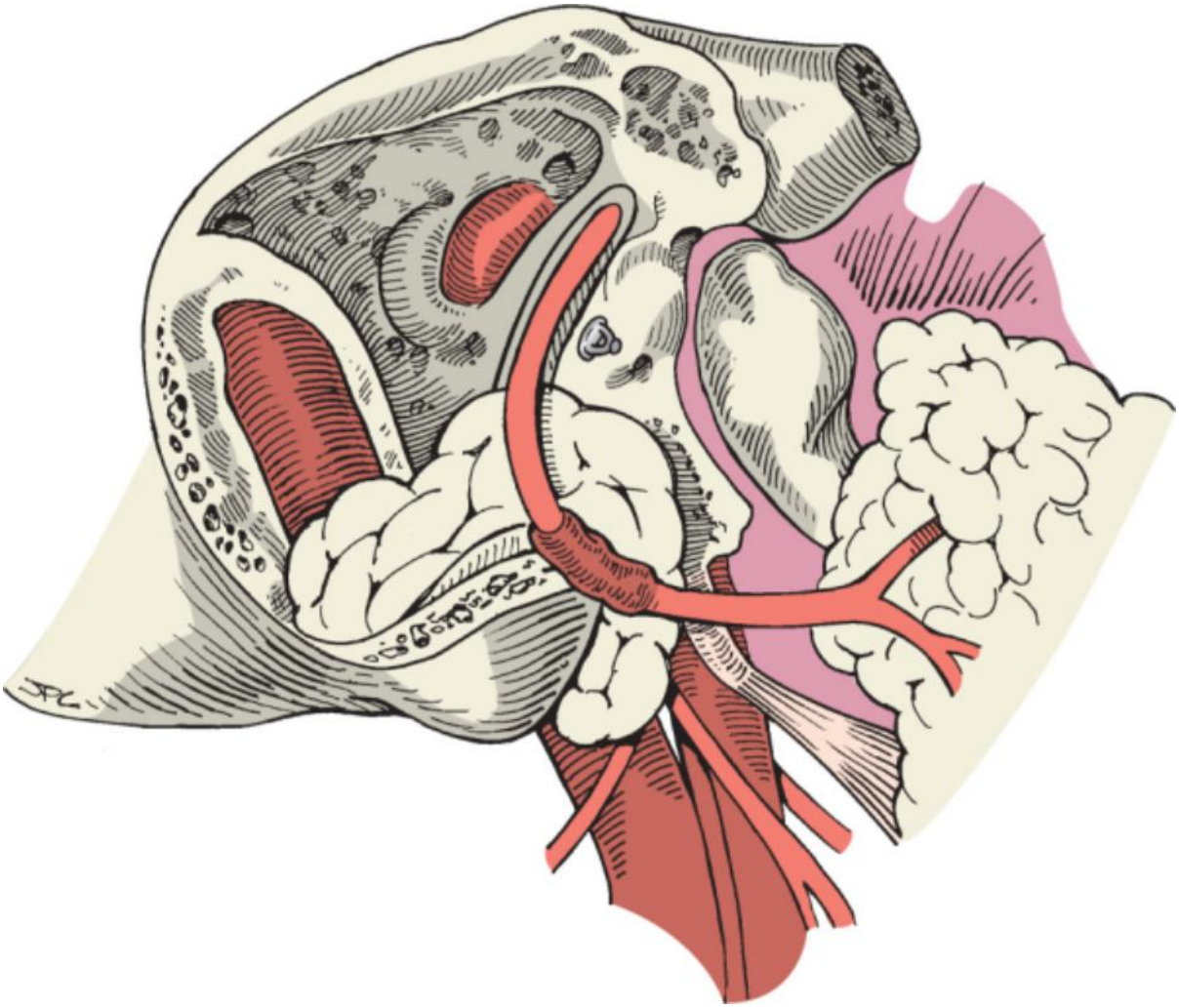

Figure 125-20 Radical mastoidectomy has been performed by removing the posterior canal wall medially to the skeletonized facial nerve. The digastric ridge is dissected in preparation for removing the mastoid tip and mobilizing the facial nerve. The tumor in the middle ear and mastoid is evident.

Precise dissection and isolation of the facial nerve in the stylomastoid foramen is difficult. The transposition is facilitated by cutting the remaining tendon and fibers of the digastric muscle within the mastoid proximal to the stylomastoid foramen. The muscle, along with the contents of the stylomastoid foramen, is elevated from the underlying bone in a posterior-to-anterior direction. The facial nerve is then dissected away from the fallopian canal. The cuff of fibrous tissue surrounding the facial nerve is sutured anteriorly to the periparotid fascia, providing retraction of the vertical and horizontal facial nerve. Complete removal of the tympanic ring and remaining mastoid tip can then be accomplished. The styloid process is identified and removed by releasing the stylohyoid and styloglossal tendons from the tip of the styloid (Fig. 125-21).

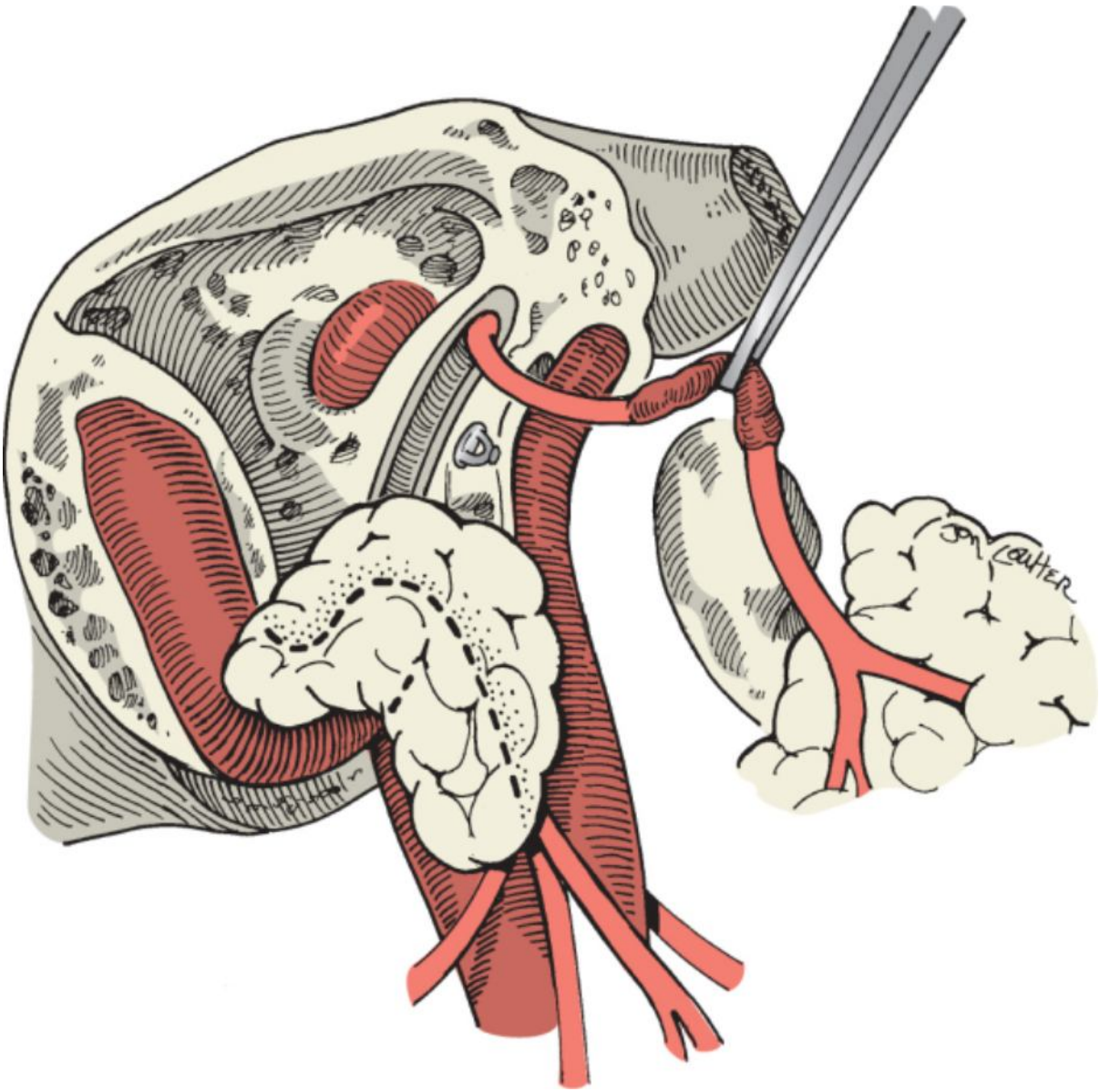

Figure 125-21 The facial nerve has been mobilized anteriorly, exposing the tumor in the mastoid, middle ear, and jugular bulb areas. The tendons attached to the styloid process have been divided. Retraction of the mandible permits isolation of the carotid artery.

Tumor Isolation and Removal

The tumor is removed. The sigmoid sinus is isolated proximally, and the jugular vein is ligated in the upper neck (Fig. 125-22). Resection of the tumor follows the same sequence as that previously described for less extensive glomus jugulare tumors. Tumor found superior to the jugular bulb and into the middle ear is dissected away from the otic capsule. If erosion into the inner ear occurs, further drilling of the otic capsule facilitates removal of the tumor.

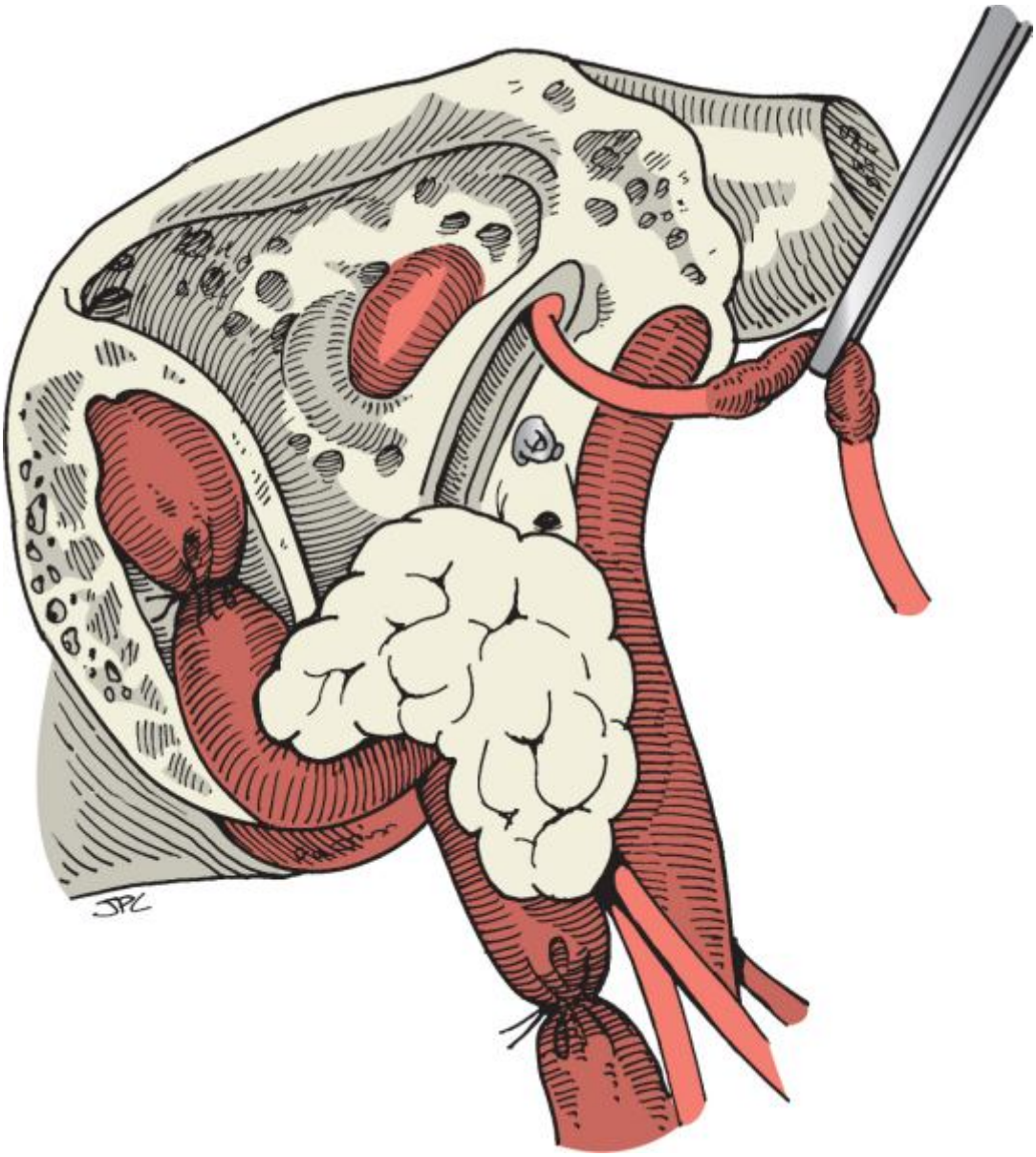

Figure 125-22 The proximal sigmoid sinus and jugular vein are suture ligated. The tumor is carefully dissected off the carotid artery. The lateral sigmoid sinus is opened.

Remaining tumor is carefully dissected off the carotid artery in a subadventitial plane, which completes the extradural dissection and removal of the tumor. The fibrocartilaginous ring surrounding the carotid artery at the skull base must be divided to achieve this plane (Fig. 125-23).

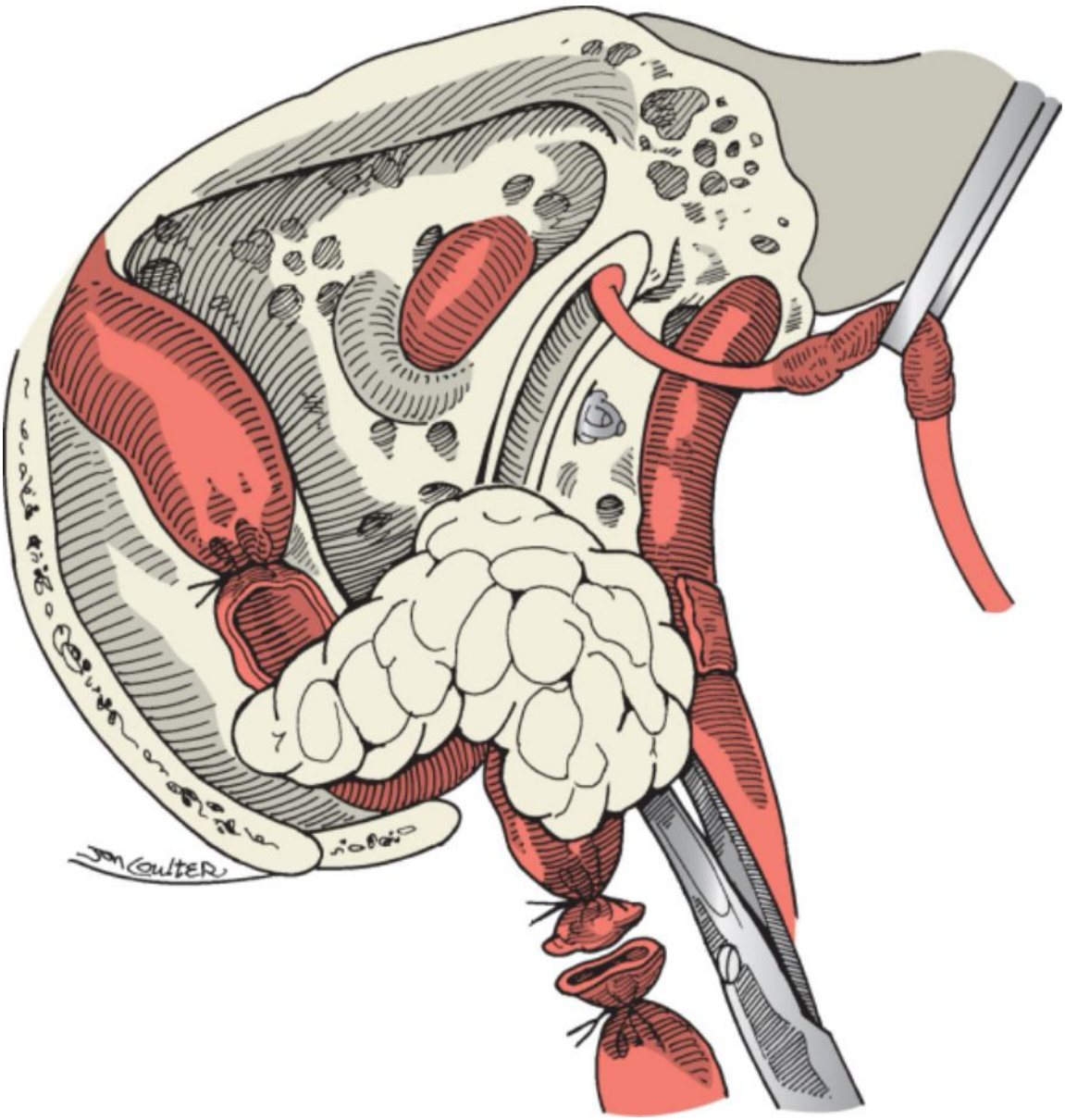

Figure 125-23 The sigmoid sinus and jugular vein have been divided. The inferior aspect of the tumor is dissected off the carotid artery in a subadventitial plane.

If intracranial tumor is found, the dura of the posterior fossa is opened. The medial wall of the sigmoid sinus and jugular bulb typically require resection. Care is taken to preserve the neural contents of the pars nervosa. Significant intracranial extension of tumor and jugular bulb involvement obviate successful nerve preservation. Tumor is removed from the cerebellopontine angle (Fig. 125-24). Often the intracranial portion of the tumor has been devascularized by the skull base dissection. Should a large amount of intracranial tumor remain with limited exposure, then a retrosigmoid craniotomy is performed. This additional exposure would be anticipated based on preoperative imaging and planning. A second-stage procedure may be required if a large amount of residual tumor is present. When all tumor has been removed and hemostasis achieved, the wound is then closed. Care is taken to ensure obliteration of the eustachian tube to avoid postoperative CSF rhinorrhea. The body of the incus, a small piece of muscle, and oxidized cellulose are packed into the eustachian tube. Large defects in the dura are repaired primarily with grafts of temporalis fascia, although watertight closure in this area can be difficult. Other fascial tissue available include galea, pericranium, and fascia lata. It also may be necessary to close the small openings in the dura where the sigmoid sinus was ligated. Small pieces of muscle are bolstered over these defects with figure-of-eight sutures in the dura. Fat is harvested from the abdominal wall. The wound is copiously irrigated with a bacitracin solution, and the fat is placed into the temporal bone defect area.

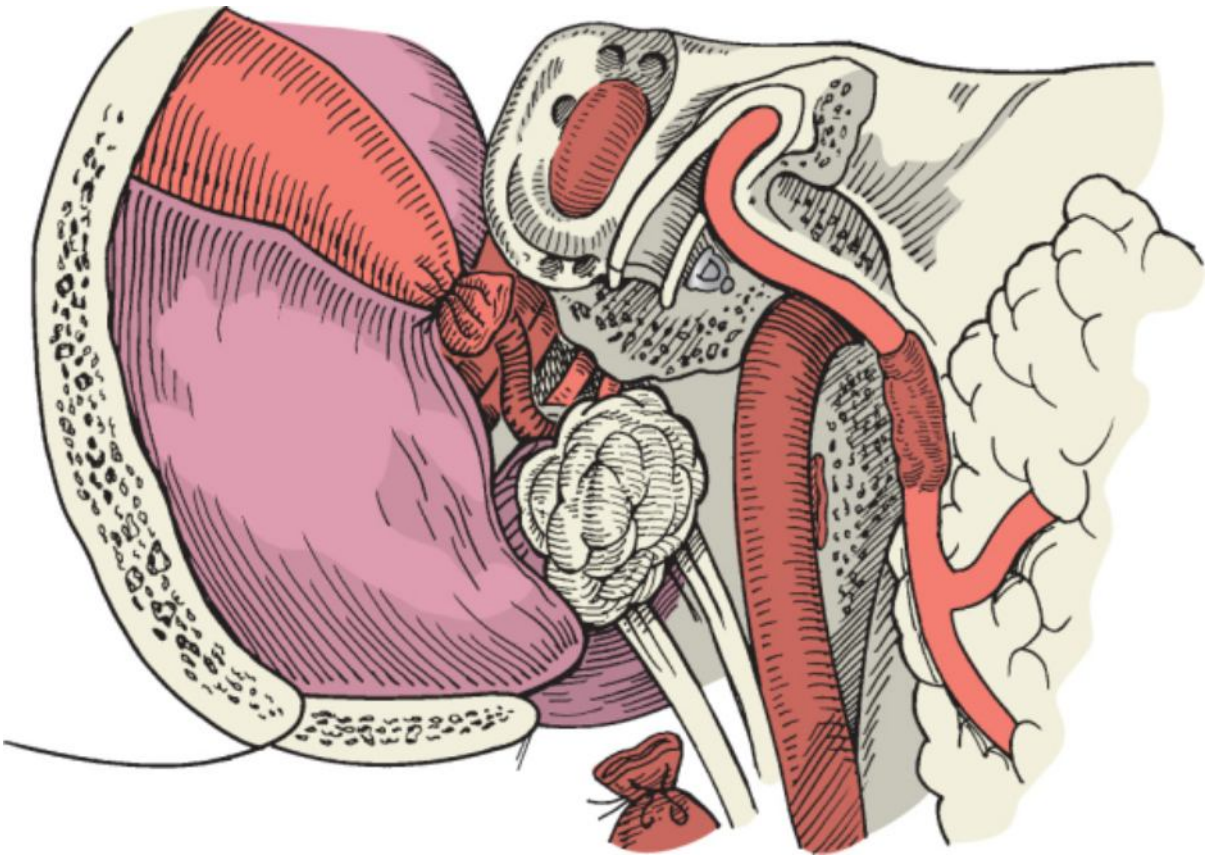

Figure 125-24 The dura is resected when tumor invades the posterior fossa and additional exposure is needed. Tumor in this area often requires sacrifice of lower cranial nerves. Dural grafting is necessary.
(Redrawn from Sekhar LN, Janecka IP [eds]: Surgery of Cranial Base Tumors. New York, Raven Press, 1993.)

Patients with large lesions requiring removal of posterior fossa dura and grafting are at greater risk for CSF leaks. Additional tissue may be necessary to provide watertight closure of the wound. The temporalis muscle can be elevated and transposed posteriorly and inferiorly in order to facilitate this closure. It is usually necessary to remove the root of the zygoma in order to pedicle and rotate the entire temporalis muscle. The edges of the temporalis muscles are approximated to the SCM and semispinalis and splenius capitis muscles (Fig. 125-25). Extensive tumors that require a transotic capsule approach with large areas of temporal bone resection create considerable dead space that may not be adequately obliterated with abdominal fat. In some cases, it may be necessary to fill the defect with a free flap of rectus abdominis muscle. The SCM and proximal posterior belly of the digastric muscles are also reapproximated to the fibroperiosteal tissue on the skin flap that covered the mastoid cortex. A small Hemovac drain is tunneled through a stab incision in the posterior neck and placed in the inferior aspect of the neck dissection and left to gravity drainage overnight. The skin flap is returned to its anatomic position and closed in a three-layer fashion. A running locking suture reapproximates the skin. Staples may also be used. A sterile compression mastoid and neck dressing are applied.

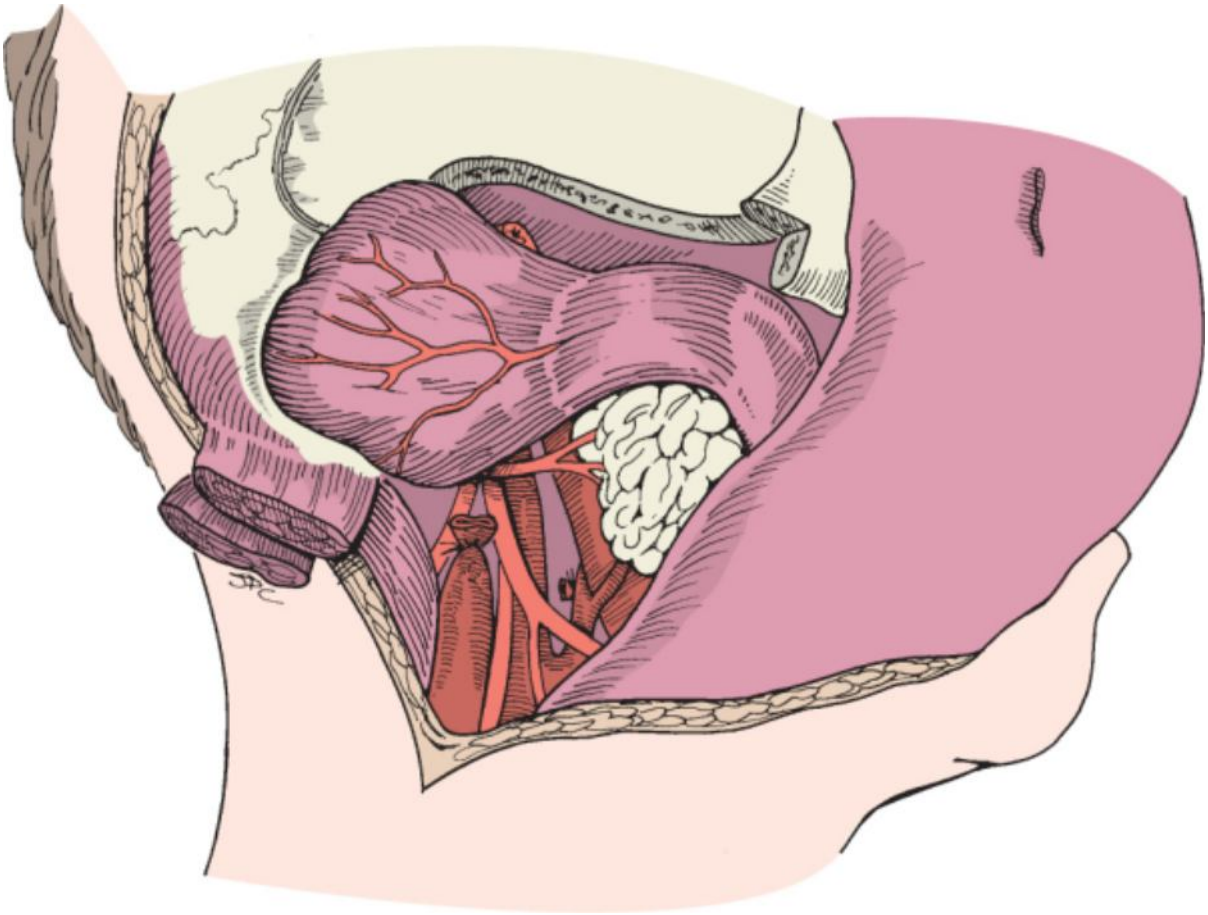

Figure 125-25 The temporalis muscle is rotated posteriorly and inferiorly to provide a vascularized graft, facilitating wound closure.

POSTOPERATIVE CARE

The level and intensity of postoperative monitoring and management are dictated by the extent of the surgical procedure and the neurovascular structures encountered. Small lesions that were approached through an endaural incision receive basic postoperative care. Vital signs are monitored on a routine basis. The ear packing is changed as needed. The patient should be ready for discharge the following morning.

Tumors excised by a combined transmeatal postauricular approach are managed in a similar fashion. A mastoid dressing that was placed postoperatively is removed the following morning. Patients are instructed to change the external canal cotton ball as needed. The postauricular incision is cleaned with peroxide or alcohol, and a topical antibiotic ointment is applied twice a day. Patients are seen 7 to 10 days postoperatively. Those patients with an endaural approach have the packing removed and the tympanic membrane inspected for integrity. Those with a combined approach have their sutures and outer ear canal packing removed. The patient is instructed to use topical ear drops twice a day, and the inner packing is removed 1 week later.

Patients with more extensive tumors requiring resection of dura frequently have the external ear canal closed as a blind sac. If a Hemovac neck drain is placed, it is removed the day following surgery. A pressure dressing remains for 3 days. Patients are carefully assessed for function of the lower cranial nerves. Oral intake is commenced if dysphagia and aspiration do not occur. Corneal protection with a moisture chamber, artificial tears, and ointment at night are provided if facial paresis is present. Patients are encouraged to be out of bed the following day. Management of complications or cranial nerve injury is detailed in the following section. Sutures are removed 7 to 10 days postoperatively. In order to assess the status of the excised tumor, a baseline scan is obtained postoperatively at 3 months. MRI with fat suppression technique helps differentiate the varied tissue densities within the operative site. Any suspicious areas identified on the scan prompt a repeat scan in 6 months. Otherwise, imaging is obtained in 1 year. If this is negative for tumor, a scan is obtained 1 year later.

COMPLICATIONS

The potential complications following glomus tumor surgery are numerous and depend on the location and extent of the tumor. The postoperative complications following glomus tumor surgery can be divided into those that pertain to problems with the wound, bleeding, CSF leak, neurologic, and cranial nerve deficits.

A seroma or hematoma may compromise wound healing. Glomus jugulare tumors require a large skin flap and dissection into the neck. Obtaining a watertight closure may be difficult with regard to the integrity of the dura. For this reason, a Hemovac drain remains only until the following day. It is kept to gravity drainage as opposed to negative pressure. A pressure dressing is also applied to the wound to minimize the possibility of seroma or hematoma. A collection of CSF (pseudomeningocele) is managed by maintaining a pressure dressing and elevating the head. If the CSF collection continues to expand, then lumbar drainage is instituted for 3 to 5 days.

Injury to cranial nerves results in functional deficits that must be addressed. Transposition of the facial nerve usually results in temporary facial paresis or paralysis. It is difficult to predict when the return of facial function will occur. Corneal protection is necessary if the patient has inadequate eye closure. Topical artificial tears are used throughout the day. Ophthalmic ointment is placed in the lower conjunctival sulcus at night. Additional daytime protection with a clear plastic moisture chamber provides comfort and minimizes corneal desiccation. Long-term eye protection is effectively provided with placement of a gold weight in the sub orbicularis oculi muscle/pretarsal space (see Chapter 121). A lower lid–tightening procedure is recommended for patients with laxity of the lower lid or those demonstrating ectropion.

Complications related to the tympanic membrane and ossicular chain can occur when excising tympanic or jugulotympanic glomus tumors. During an endaural approach with the tympanomeatal flap elevated, inadvertent contact with a microbipolar forceps to the tympanic membrane may result in ischemia and necrosis. A tympanic membrane perforation may result, requiring a patch or formal myringoplasty (see Chapter 113). Persistent conductive hearing loss as a result of ossicular chain reconstruction can be subsequently revised or treated with a hearing aid. The maximal conductive hearing loss that results from obliteration of the middle ear space and closure of the ear canal is more difficult to manage. Unless a deep cuff of tissue is constructed to create an external auditory meatus that would fit an ear mold, the use of a hearing aid will not be successful.

Injury or sacrifice to the ninth and tenth cranial nerves in patients who previously were asymptomatic results in problems of dysphagia, aspiration, and hoarseness. Despite the patient being under general anesthesia, it is reasonable to perform an intraoperative vocal cord medialization procedure (thyroplasty type I) while the neck is open (see Chapter 41). If the 10th cranial nerve is anatomically intact, then postoperative assessment is determined when the patient is awake.

Injury to the spinal accessory nerve results in shoulder drop and dysfunction. Postoperative physical rehabilitation therapy should be requested once the patient is ambulatory.

Tumors that extend inferiorly into the neck may put the hypoglossal nerve at risk. This would be unusual unless there is an associated large glomus vagale or carotid body tumor. Dysmotility of the tongue may result in mild dysarthria and problems with deglutition. Functionally, this injury alone is usually well tolerated. However, in combination with injury to the ninth and tenth cranial nerves, hypoglossal injury puts the patient at greater risk for dysphagia and aspiration. Thus patients who are clinically symptomatic postoperatively should be evaluated for a vocal cord medialization procedure. An arytenoid adduction procedure augments the closure of the posterior glottic chink, which further minimizes the potential for aspiration. Tracheotomy may still be necessary for uncontrolled aspiration and pulmonary toilet. Cricopharyngeal myotomy further relaxes the upper esophageal sphincter, which facilitates swallowing.

Watertight closure may be difficult when the dura is resected and repaired. Avoidance of a CSF leak is accomplished by meticulous obliteration of the eustachian tube, packing of the mastoid cavity with fat, and three-layer closure to the muscle and fibroperiosteum, subcutaneous tissue, and skin. Due to the critical structures adjacent to glomus tumors including the lower cranial nerves, carotid artery, jugular vein and venous sinuses, the dura, brain and spinal fluid serious complications of lateral skull base surgery do occur. The mortality from management of the carotid artery has resulted from cerebrovascular complications. In a large series of glomus tumors of the temporal bone treated surgically, the rate of CSF leak was 11.6% and incidence of aspiration pneumonia was 5.3%.[5] The incidence of postoperative CSF leak dropped to 4.5% when dural closure or reconstruction involved a vascularized local, regional, or free flap.[14]

If CSF leak occurs through the skin or nose despite a pressure dressing, lumbar drainage is instituted. This is maintained at 6 to 10 mL/hr over 3 to 5 days. Although rarely necessary, persistent CSF leak may warrant wound reexploration.

Ongoing problems with aspiration may pre-vent resumption of oral intake. In the immediate postoperative period, nutritional support is provided by nasogastric feeding. Despite efforts to provide glottic competence with swallowing therapy, a gastrostomy or jejunostomy feeding tube may be necessary.

Management of intraoperative injury to the ICA is dictated by results of BTO and xenon blood flow studies. Demonstration of inadequate contralateral perfusion requires preservation of the ipsilateral carotid. Laceration or tear is either primarily repaired or replaced with a saphenous vein graft. Despite data providing assurance of

adequate contralateral flow, ligation of the carotid artery may result in progressive thrombus formation and possible emboli. Patients in whom a carotid artery was repaired or grafted should have postoperative studies, such as Doppler ultrasound, MRA, or transfemoral arteriography, to determine the status of the ICA. Patients exhibiting ischemic neurologic compromise should be considered for embolectomy or heparinization.

PEARLS

- Arteriography and venous outflow studies are helpful for large tumors by identifying the blood supply of the tumor, the proximity of the major vessels, occlusion of the ipsilateral jugular vein, patency of the contralateral venous drainage system, and the opportunity for tumor embolization before surgical resection.
- Stereotactic radiotherapy may provide effective control of tumor when given as the primary modality of treatment, following recurrence of tumor, or as part of the planned strategy for subtotal resection.
- Small cotton balls soaked with 1 : 1000 epinephrine are useful to temporarily compress sites of bleeding during tumor removal from the middle ear.
- Rotation of a vascularized temporoparietal fascia/muscle flap is an effective reconstructive technique for closure of a cranial and dural defect, minimizing the risk of CSF leak.
- The potential morbidity of newly acquired cranial nerve dysfunction (CN VII-XII) following resection of large glomus jugulare tumors demands a comprehensive discussion with the patient and family and documentation of the informed consent containing the options, risks, and benefits of the treatment.

PITFALLS

- Failure to recognize potential compromise of the lower cranial nerves following resection of large glomus jugulare tumors may incur significant postoperative morbidity and changes in the quality of life.
- Intraoperative severe and difficult to control hypertension may occur in patients with unrecognized preoperative symptoms and signs of a vasoactive-secreting tumor.
- Incomplete assessment of patients with a family history of paraganglioma may proceed toward subsequent growth of multicentric tumors resulting in more difficult management decisions affecting the cranial nerves and great vessels of the head and neck.
- Following resection of a glomus jugulare tumor, sacrifice of a patent sigmoid sinus and jugular vein when the contralateral central venous outflow is insufficient may result in intracranial hypertension.
- Unexpected facial nerve injury can result if involvement of the vertical portion of the facial nerve from posterior tumor extension in the middle ear and hypotympanum is not recognized.