Chapter 6 – Juvenile Angiofibroma

Ricardo L. Carrau,
Carl H. Snyderman,
Amin B. Kassam

Juvenile nasopharyngeal angiofibroma (JA) is a rare benign tumor found exclusively in males, most often during adolescence or young adulthood. The tumor accounts for less than 0.5% of all tumors of the head and neck and is found in 1 in 16,000 hospital admissions to an ear, nose, and throat service.[1] It is believed that this tumor has been recognized since the time of Hippocrates.[2]

The origin of JA remains undefined; however, its exclusive occurrence in males suggests a hormonal influence. The site of origin is most likely at the basisphenoid, near the sphenopalatine foramen, at the junction of the orbital and sphenoid processes of the palatine bone and the body of the sphenoid bone. This site of origin explains the frequent extension of the tumor into the pterygopalatine fossa. JA also extends into the nasal cavity, the nasopharynx, and the ethmoid and sphenoid sinuses. JA may invade the orbit and cranial cavity by direct extension following preformed pathways or by destroying bone.

PATIENT SELECTION

History and Physical Examination

Nasal obstruction and epistaxis are the most common complaints of patients with JA. Any adolescent male presenting with these symptoms should be suspected of having JA. As the tumor extends into other areas such as the nasal cavity, orbit, and intracranial cavity, other signs and symptoms may develop, such as proptosis or displacement of the globe, diplopia, pain in the cheek, headache, swelling of the cheek, sinusitis, headaches, and hearing loss (e.g., middle ear effusion). The findings on physical examination depend on the extent of the tumor. Endoscopic nasal examination and cranial nerve testing provide clues regarding the extent of the tumor. Typically, the JA is a gray-red, smooth, sessile, polypoid mass, although it can be irregular and lobulated (Fig. 6-1A-C). Physical examination may reveal signs of orbital extension such as proptosis, displacement of the globe, and limitations of extraocular muscle movement (see Fig. 6-1D). Otoscopic evaluation may demonstrate serous effusion or a retracted tympanic membrane.

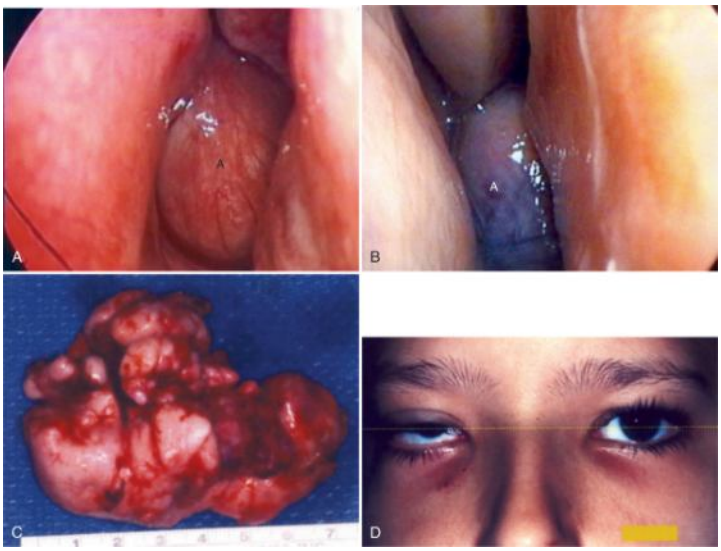

Figure 6-1 Fiberoptic examination demonstrating a polypoid juvenile nasopharyngeal angiofibroma (JA) occupying the right **(A)** and left **(B)** nasal cavities. A, angiofibroma. **C,** Surgical specimen of a large multilobulated JA. **D,** Patient with epiphthalmos caused by a massive angiofibroma.

Imaging

JA may be identified on plain films as a soft tissue mass occupying the posterior nasal cavity with or without extension into the nasopharynx or paranasal sinuses. The most suggestive finding is anterior bowing of the posterior wall of the maxillary sinus observed on a submentovertex view. Computed tomography (CT) scanning, however, is the preferred imaging technique for the initial evaluation of a patient with a suspected JA. CT scanning defines the bony architecture of the sinonasal tract and skull base, and the use of contrast demonstrates the vascularity of the tumor (Fig. 6-2A). Magnetic resonance imaging (MRI) complements the CT scan in those cases that present with intracranial, infratemporal, or intraorbital soft tissue extension (see Fig. 6-2B). MRI better defines the soft tissue planes, demonstrating the interface between the tumor and the soft tissue of the infratemporal fossa (muscles or neurovascular structures), or intracranial structures, and helps differentiate tumor extension from retained secretions in the sinuses.

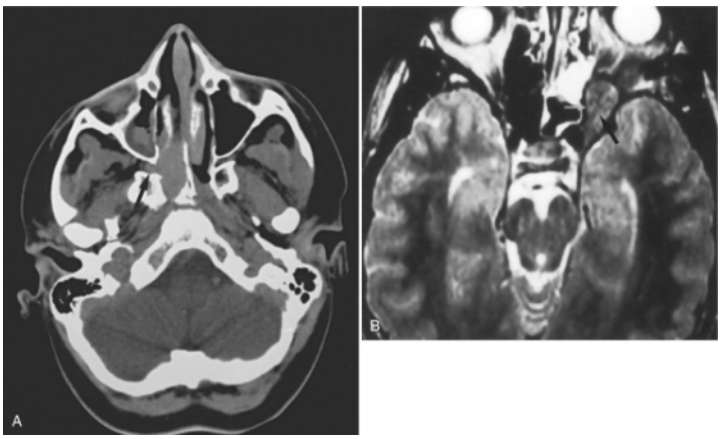

Figure 6-2 **A,** Computed tomography scan, axial view. Juvenile angiofibroma occupying the right nasal cavity (soft tissue algorithm). The *arrow* points to a tumor expanding the pterygopalatine fossa. The right maxillary sinus contains secretions. **B,** Magnetic resonance imaging with T2 sequence demonstrating extension of the angiofibroma into the middle cranial fossa *(arrow)*.

Angiography demonstrates the blood supply of the tumor, which is usually derived from branches of the internal maxillary and ascending pharyngeal arteries. Contributing branches from the internal carotid artery are common in tumors that extend to the nasopharynx, sphenoid sinus, orbital cavity, and infratemporal fossa and are the rule in tumors with intracranial extension. Therefore the angiographer should evaluate the circulation of the external and internal carotid arteries bilaterally. Angiography, however, is invasive and is associated with a defined, albeit low, number of complications. Its use is reserved for preoperative embolization.

Embolization should be considered as part of the preoperative preparation and should be performed shortly before the surgical procedure (<24 hours), because JA is known to achieve rapid

revascularization. This diagnostic angiography and the therapeutic angiography (e.g., embolization) are scheduled as a single-stage procedure. Other factors, such as limitations on the maximal volume of contrast, patient anxiety, and length of the procedure, may necessitate a modification of this plan (Figs. 6-3 and 6-4).

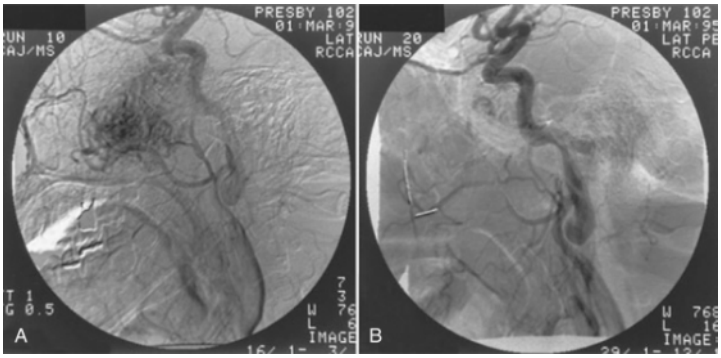

Figure 6-3 **B,** Angiography with injection in the right common carotid artery demonstrating the tumor blush. Branches of the external and the internal carotid arteries supply blood to the tumor. **B,** Postembolization view.

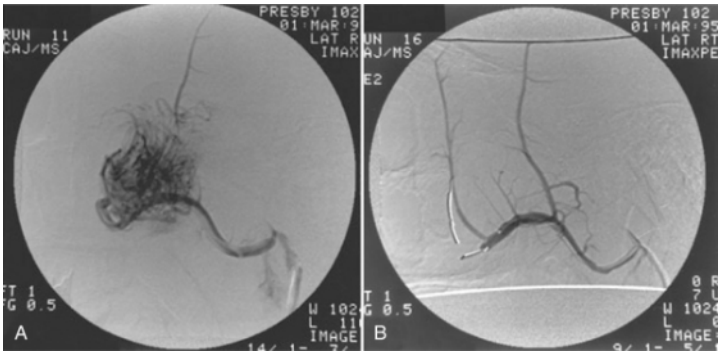

Figure 6-4 **A,** Superselective angiography with injection of contrast in the internal maxillary artery producing an intense tumor blush. **B,** Postembolization view. To obtain this degree of blood flow reduction, the patient required bilateral superselective embolization of the distal internal maxillary arteries (to preserve the blood supply to the masseter and temporalis muscles) and ascending pharyngeal arteries.

Biopsy

Histologic confirmation is not usually necessary for the diagnosis of JA. A definitive diagnosis is usually made with clinical and imaging findings. Malignant tumors such as rhabdomyosarcoma and olfactory neuroblastoma can mimic JA; therefore, a biopsy is prudent in those patients who are to be treated with primary radiation (poor surgical candidates) or in patients in whom the diagnosis remains uncertain even after thorough evaluation. The biopsy should be performed in the operating room with the patient under general anesthesia (e.g., controlled airway). The characteristic histologic appearance is that of a thin-walled vascular network surrounded by fibrous stroma (Fig. 6-5).[1]

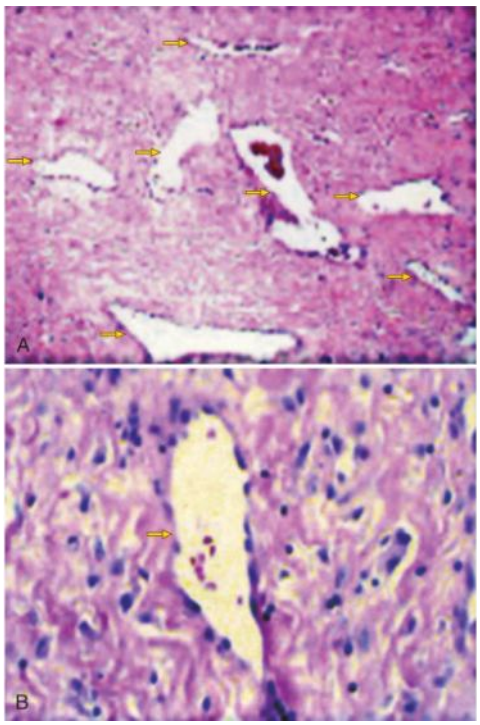

Figure 6-5 Histologic section of juvenile angiofibroma at ×10 **(A)** and ×40 **(B)** magnification, demonstrating the characteristic thin-walled vascular network *(arrows)* surrounded by fibrous stroma.

Staging

Different staging systems have been proposed for JA (Tables 6-1 and 6-2).[3,4] They facilitate communication among different institutions and allow comparisons of outcomes after different therapeutic modalities and surgical approaches. Nevertheless, these systems are rarely used prospectively and their usefulness is limited because most institutions have not adopted any single staging system.

Table 6-1 -- STAGING OF JUVENILE NASOPHARYNGEAL ANGIOFIBROMA

Stage I	Tumor limited to posterior nares and nasopharyngeal vault
Stage IIA	Minimal lateral extension into the pterygomaxillary fossa
Stage IIB	Tumor fills pterygomaxillary fossa, displacing posterior wall of antrum, and extends superiorly, eroding bone of orbit
Stage IIC	Extension into cheek and temporal fossa through pterygomaxillary fossa
Stage III	Intracranial extension

Table 6-2 -- STAGING OF JUVENILE NASOPHARYNGEAL ANGIOFIBROMA

Stage	Tumor Location
I	Nasopharynx
II	Nasal cavity and sphenoid
III	Ethmoid and maxillary sinuses, pterygomaxillary space, infratemporal fossa, orbit, and cheek
IV	Intracranial

PREOPERATIVE PLANNING

Preoperative preparation aims to diminish the risk of complications associated with massive bleeding and blood transfusions. Hormonal therapy, using androgens or estrogens, has been advocated in the past as a way to decrease the bulk and vascularity of JA. However, these drugs have significant side effects that are very undesirable for any adolescent male (e.g., testicular atrophy, feminization, gynecomastia). These side effects, as well as new developments in the instrumentation and techniques for angiographic embolization, make hormonal therapy undesirable and unnecessary.

Angiography and embolization should be completed in a single procedure. JAs may develop rapid revascularization after embolization; therefore, definitive surgery should ideally be scheduled within 24 hours after the angiography. Close communication with the interventional radiologist is essential. Details, such as preservation of the blood supply to the temporalis muscle if needed for reconstruction, should be discussed in advance.

Despite adequate embolization, blood in the form of packed red blood cells should be available for the surgery. We encourage the use of autologous blood banking to avoid the risks of mismatched blood transfusion and the transmission of infectious agents. Frequently, the surgery may be delayed for several weeks to allow for autologous donation of 2 to 3 units of blood. A cell saver device, which allows the safe recycling of the patient's blood despite contamination by the flora of the upper aerodigestive tract, can also be used during the surgery.

The extirpative surgery is considered to be clean-contaminated, for which perioperative prophylactic antibiotics with broad-spectrum coverage of the flora of the upper aerodigestive tract are recommended. The antibiotics are initiated at the time of surgery and continued for 24 hours postoperatively. In those cases in which there is communication of the cranial cavity with the upper aerodigestive tract, antibiotics are continued for 48 hours postoperatively.[5]

SURGICAL TECHNIQUE

Therapeutic Modalities

Historically, surgery and radiation therapy yield a similar control rate (80% cure) of JA.[6–10] However, surgery is favored for most cases owing to the long-term sequelae of radiation therapy (e.g., secondary malignancies, cataracts, pituitary insufficiency, mucous gland metaplasia) and the improved surgical outcome and morbidity brought about by advancements in embolization and surgical approaches.[6–14]

Radiation may be considered a primary therapeutic modality for the rare patient who is a poor surgical candidate or for those who refuse surgery. The recommended dose ranges from 35 to 45 Gy.[6,7] Adjunctive radiation is also recommended for those cases in which the intracranial component (or intracranial recurrence) is deemed unresectable. Many of these patients, however, do not need any treatment and may be followed with imaging (remnants of tumors may show involution).

Other therapeutic modalities, such as cryosurgery and sclerotherapy, are of historical importance only. Chemotherapy has been advocated by some authors for the treatment of recurrent or progressive nonresectable tumors that have failed to respond to radiation therapy.[15] Future advances in tumor biology may lead to new nonsurgical therapies using inhibitors of specific growth factors or angiogenesis.

SURGICAL APPROACHES

The surgical approaches to JA are tailored to the extent of the tumor but are also based on the surgeon's experience and familiarity with the different approaches (experienced surgeons may use the more limited approaches to remove even massive tumors). In many instances, the extirpative surgery requires the use of a combination of approaches to provide for complete visualization of the tumor and control of bleeding.

The surgical approaches can be divided into inferior, anterior, and lateral. Inferior approaches include transpalatal (i.e., through the hard palate) and transoral-transpharyngeal routes, with or without a soft palate split. These techniques provide access to the nasopharynx and the nasal cavity. Anterior approaches expose the nasal cavity (e.g., lateral rhinotomy and midfacial degloving) and can be extended through the medial maxilla to expose the antrum and ethmoid sinuses and the pterygopalatine fossa (e.g., LeFort I, Denker's, and medial maxillectomy) or even the sphenoid sinus and nasopharynx (e.g., swing maxillotomy and facial translocation). In addition, these latter approaches may be extended to expose the infratemporal fossa, infratemporal skull base, middle cranial fossa, or paramedian skull base (anterolateral exposure). Alternatively, the infratemporal fossa may be approached laterally through a preauricular subtemporal route (i.e., lateral approach).

Endoscopic techniques are our preferred approach for the extirpation of JAs.[11] Rod lens endoscopes are useful during the preoperative evaluation to visualize the anterior extension of the tumor and to complement the most limited approaches (e.g., transpharyngeal and transpalatal) by providing improved visualization of the ethmoid and sphenoid sinuses. Selected tumors extending to the nasal cavity, nasopharynx orbit, infratemporal fossa, or cranial cavity may be amenable to an endoscopic transnasal approach. The middle turbinate is removed and a wide nasoantral window (NAW) is opened to facilitate the removal of the posterior wall of the antrum (Fig. 6-6A and B). Septal and sphenoidal extensions are freed so that the tumor can be pushed into the nasopharynx. This gains space within the nasal cavity and allows an easier manipulation of the tumor. Extensions into the PtPF (pterygopalatine fossa) and ITF (infratemporal fossa) are reached by mobilizing the tumor from its bony attachments at these areas and then retracting the tumor into the nasoantral area (see Fig. 6-6C). The mucosa around the tumor is incised using an electrocautery, and the tumor is dissected off the pharyngobasilar and basioccipital fascia in toward the nasopharynx.

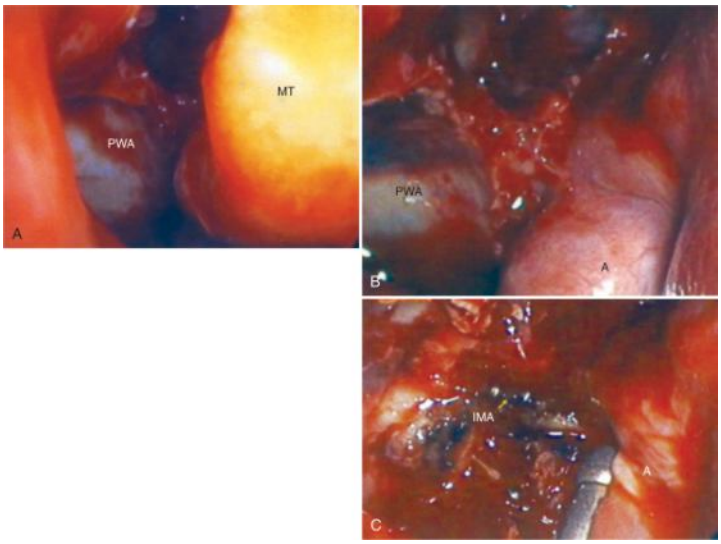

Figure 6-6 Intraoperative picture demonstrating the initial approach to the right middle meatus and opening of a large nasoantral window to expose the posterior wall of the antrum (PWA). **B,** Intraoperative picture demonstrating the augmented exposure of the tumor after removal of the middle turbinate. **C,** Intraoperative picture demonstrating the medial retraction of the tumor toward the nasal cavity after removing the PWA. A, angiofibroma, E, ethmoid sinuses; IMA, internal maxillary artery; LP, lamina papyracea; MT, middle turbinate.

Transpharyngeal or transpalatal approaches are viable alternatives. The soft palate may be retracted with the help of two red rubber catheters introduced through the nose and taken out through the mouth. If the tumor is adequately visualized, the surgeon proceeds with the resection; if not, a midpalate split is recommended. The midpalate split is performed using electrocautery following a lazy S pattern (Fig. 6-7). This type of incision prevents velopharyngeal insufficiency brought by contraction of a linear scar. Each half of the velum is retracted laterally to allow full visualization of the

nasopharynx and posterior choanae (Fig. 6-8).

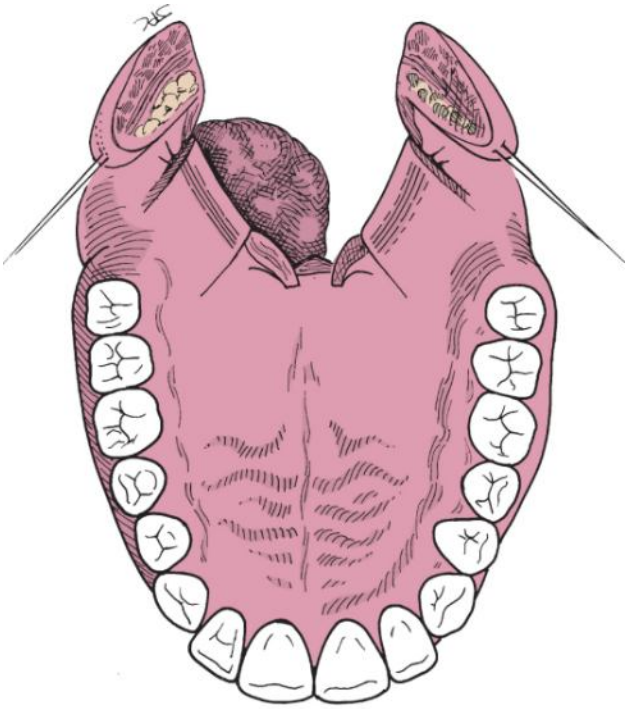

Figure 6-7 Incision for the soft palate split. The horizontal release incision should be performed anterior to the posterior edge of the hard palate to prevent the formation of an oral-nasal fistula at the trifurcation. Alternatively, this horizontal incision may be performed as a V-shaped incision and closed in a V-to-Y fashion to elongate the soft palate.

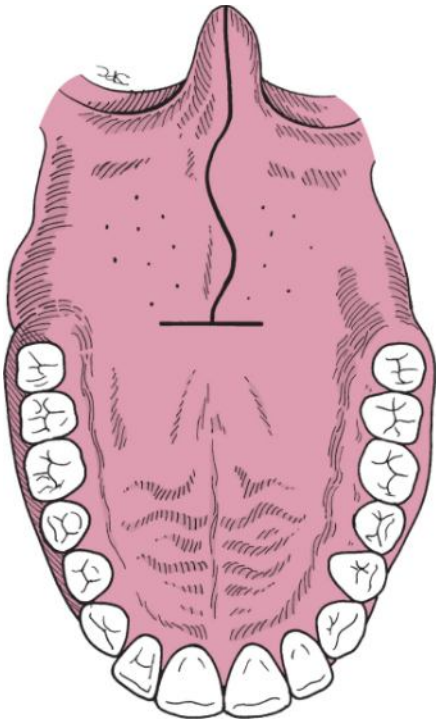

Figure 6-8 Retraction of the soft palate after the split, providing exposure of the tumor.

The exposure of the pterygopalatine fossa may be enhanced by resecting the bone of the hard palate. The mucoperiosteum of the hard palate is incised, creating a U-shaped, posteriorly based flap (Fig. 6-9). The mucoperiosteum is elevated, exposing the palatine bone back to the junction with the soft palate. Alternatively, the mucoperiosteum can be based on the contralateral greater palatine neurovascular bundle (sacrificing the ipsilateral nerve, artery, and vein) (Fig. 6-10). This upgrades the mobility of the flap and provides uninterrupted exposure from the nasal cavity to the nasopharynx. The bone is then removed with a Kerrison rongeur, exposing the mucosa of the floor of the nose, which is then incised (Fig. 6-11). The posterior septum may be removed to better expose the nasopharynx and rostrum of the sphenoid. The hamulus process and the inferior portion of the medial pterygoid plate can be removed for better exposure of the pterygopalatine fossa (Fig. 6-12).

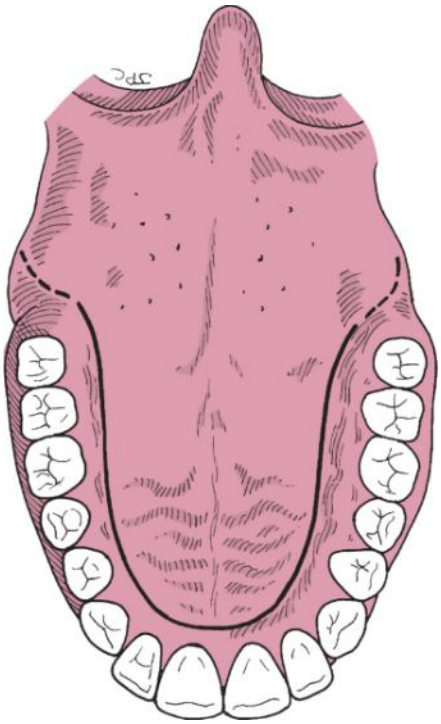

Figure 6-9 U-shaped mucoperiosteal incision. The *dashed line* corresponds to a release incision to provide further exposure to the pterygoid plates (the pterygopalatine fossa) and more mobility to the mucoperiosteal flap on the side ipsilateral to the tumor.

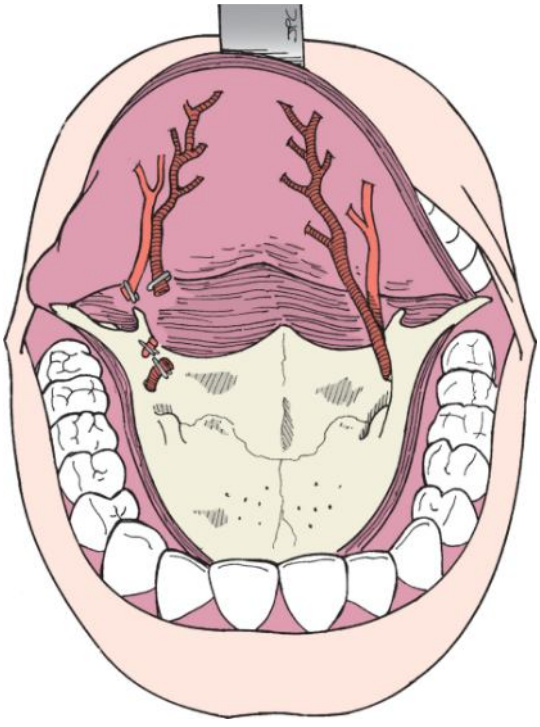

Figure 6-10 The mucoperiosteal flap has been elevated to expose the bone of the hard palate. The greater palatine neurovascular bundle, ipsilateral to the tumor, has been ligated and transected to enhance the mobility of the mucoperiosteal flap.

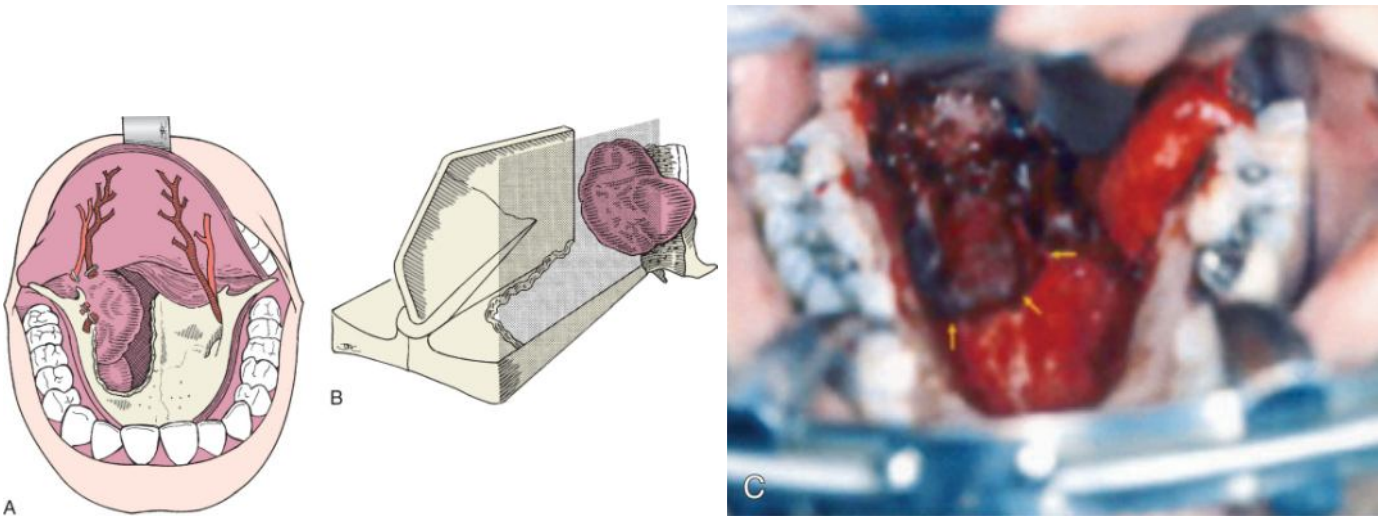

Figure 6-11 **A,** The palatine bone corresponding to the floor of the nose has been removed with a Kerrison rongeur (or drill), and the mucosa has been incised to provide exposure of the tumor. **B,** Three-dimensional illustration of the approach to a tum the posterior nasal cavity and with minimal invasion of the pterygopalatine fossa. **C,** Intraoperative view demonstrating the removal of the right palatine bone *(arrows)* to expose the mucoperiosteum of the nasal floor (n).

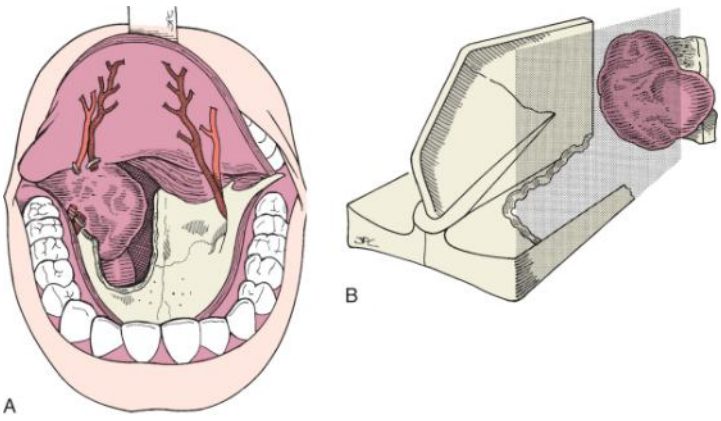

Figure 6-12 **A,** Exposure of the inferior aspect of the tumor extension into the pterygopalatine fossa after removal of the inferior aspect of the pterygoid plates. **B,** Three-dimensional illustration of the completed approach.

The final stage of the tumor resection may be described as a controlled avulsion. The mucosa around the tumor is incised using an electrocautery, and the tumor is dissected off the pharyngobasilar and basioccipital fascia in toward the nasopharynx. At this point, the tumor is avulsed, using blunt dissection. Bleeding is controlled with Yankauer suctions, bipolar cautery, and packing.

The floor of the nasal mucosa and the hard palate mucoperiosteum are repaired with interrupted absorbable sutures (e.g., chromic 3-0). The soft palate is repaired with a three-layer closure using chromic 3-0 interrupted sutures for the nasal surface and Vicryl 3-0 for the muscle, oral mucosa, and mucoperiosteum. The hard palate mucoperiosteum is pressed against the remaining bone using an acrylic splint (Fig. 6-13).

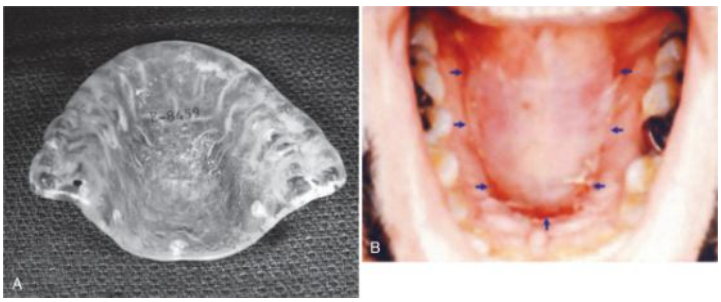

Figure 6-13 **A,** An acrylic splint. **B,** A well-healed palate incision.

The LeFort I osteotomy also provides exposure of the anterior nasal cavity, pterygopalatine fossa, and nasopharynx. In addition, the hard palate and premaxilla can be split sagittally to swing the ipsilateral maxilla as a medially based flap. The basic approach consists of a degloving incision that follows the horizontal plane from maxillary tuberosity to maxillary tuberosity approximately 5 cm above the gingival margins (Fig. 6-14A) and intranasal incisions (see Fig. 6-14B). The soft tissues of the midface are elevated in a subperiosteal plane, exposing the infraorbital neurovascular bundles, the body of the zygomatic bone, and the piriform aperture (Fig. 6-15). The mucosa over the floor of the nose and the mucoperichondrium and mucoperiosteum of the septum are elevated, exposing the septal cartilage, vomer, and maxillary crest, bilaterally. The nasal septum and vomer are separated from the maxilla using a 6-mm chisel or osteotome. The position for the LeFort I osteotomy is marked, and rigid osteosynthesis plates are bent to conform to the surface of the maxilla and are screwed in place, as will be required to repair the osteotomies (Fig. 6-16). The plates and screws are then removed and the LeFort I osteotomy is performed with a reciprocating saw, preserving the apices of the dental roots. The pterygoid plates are separated from the maxilla using a curved osteotome. Care is taken to avoid injury to the internal maxillary artery. (It is preferable to identify and clip the ipsilateral internal maxillary artery in order to secure hemostasis.) The junction with the lateral wall of the nose is separated with Mayo or double-action scissors, and the posterior wall of the antrum is fractured on mobilization of the maxilla in a caudal direction (Fig. 6-17).

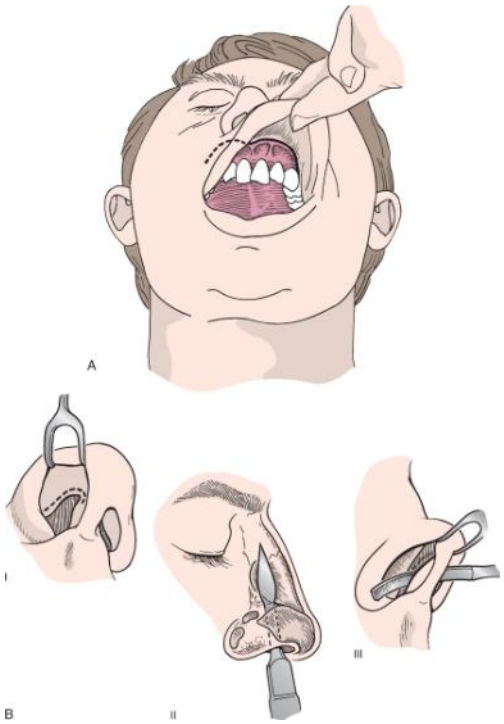

Figure 6-14 **A,** Gingivobuccal incision. **B,** Intercartilaginous incisions *(I)* are joined to a transfixion incision at the anterior edge of the nasal septum *(II)*. The soft tissue is elevated from the nasal bones *(III)*.

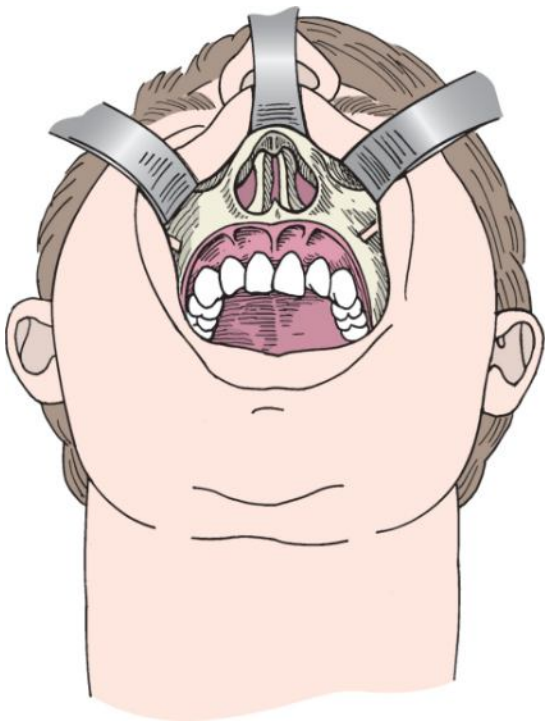

Figure 6-15 The soft tissues of the midface are elevated, providing access to the maxilla and nasal cavity bilaterally. The superior exposure is limited by the infraorbital neurovascular bundles.

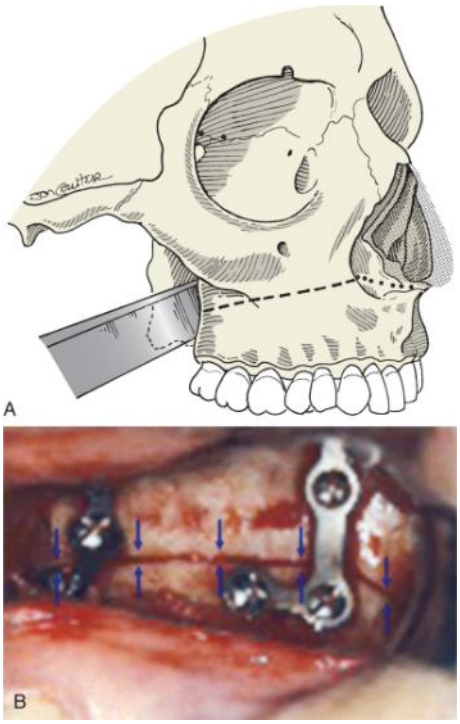

Figure 6-16 **A,** Level of LeFort I maxillary osteotomies. The attachments of the cartilaginous septum to the maxillary crest and the vomer bone are severed. It is greatly advantageous to pre-plate the maxilla before the osteotomies are performed. Pre-plating obviates the need for maxillomandibular wiring to restore occlusion before plating, saving considerable time. The attachment of the pterygoid plate to the maxilla is fractured with a curved osteotome. **B,** Intraoperative photograph demonstrating pre-plating of a Le Fort I osteotomy. The anterior osteotomy is performed and the maxilla is pre-plated before completing the posterior osteotomy.

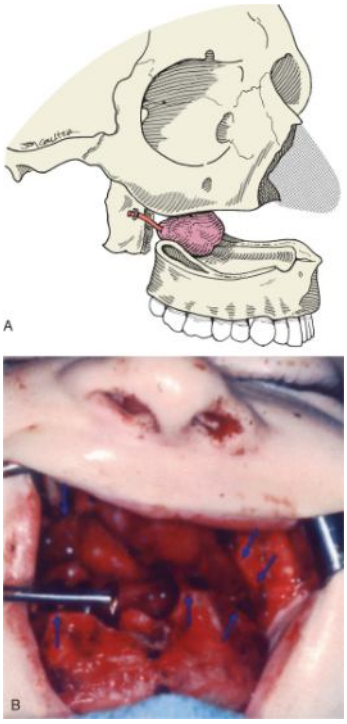

Figure 6-17 **A,** The LeFort I osteotomies have been completed, and the internal maxillary artery has been clipped and transected, exposing the tumor occupying the nasal cavity and pterygopalatine fossa. **B,** Intraoperative photograph demonstrating plating of the Le Fort I osteotomy *(arrows)*.

Hemostasis should be complete at the end of surgery. The routine use of postoperative anterior or posterior nasal packings or nasogastric tubes is discouraged. A nasal airway (e.g., nasal trumpet) may be used to secure the airway, especially in patients who develop significant soft palate swelling. A tracheotomy is indicated if the swelling involves the rest of the oropharynx or extends to the hypopharynx or larynx, as may be the case after extensive infratemporal fossa dissection.

POSTOPERATIVE CARE

Postoperatively, the patient is transferred to an intensive care unit for close monitoring of vital signs, oximetry, and neurologic status. As soon as the gastrointestinal tract is deemed ready, a clear liquid diet is started and advanced as the patient tolerates. A soft diet is continued for 10 to 14 days for those patients who undergo a soft palate split. Patients treated with anterior or lateral approaches may be advanced to a regular diet as tolerated. Patients with wounds over the hard palate benefit from a palatal splint to protect the suture line and to press the mucoperiosteum against the remaining bone.

Skin incisions are cleaned three times daily with normal saline solution and are then covered with an antibiotic ointment. Intraoral wounds mandate the use of antiseptic mouthwashes (e.g., chlorhexidine gluconate [Peridex]). An oral hygiene appliance (Water Pik) is recommended for dental care, because patients are advised not to brush their teeth for 10 to 14 days or until the wounds are healed.

Prophylactic antibiotics are continued as discussed under “Preoperative Planning” earlier in this chapter.

COMPLICATIONS

Postoperative bleeding after surgery for JA varies from simple oozing to massive hemorrhage that needs rapid volume replacement and control of the airway. This discussion concerns the latter

situation.

Management of significant hemorrhage should follow the ABCs of resuscitation. When at risk, the airway should be controlled through endotracheal intubation or tracheotomy, and the intravascular volume should be initially replaced with isotonic solutions (e.g., Ringer's lactate).

At the same time, measures are taken to control the bleeding. The method for controlling the hemorrhageis influenced by the extension of the surgery and the surgical approach. In most cases, the surgeon has a clear idea what vessels could have been injured during the operation. An emergency posterior packing will control most bleeding problems after JA surgery, providing time for a more thorough evaluation. After the initial stabilization, the patient is transferred to the operating room for identification and ligation of the bleeding vessel. Alternatively, in some cases an angiography with embolization can provide hemostasis without disrupting the surgical field. This may be the procedure of choice in patients in whom multiple approaches were used or in those who required a complex reconstruction.

Infection after surgery for JA is rare. Its treatment should follow the same sensible principles for treatment of postoperative infection anywhere else. Cellulitis is treated with intravenous antibiotics, abscesses should be drained, and necrotic tissue should be débrided. Bone grafts or flaps usually remain viable as long as the rigid fixation is stable.

Other complications such as cranial nerve deficit or intracranial problems are discussed in Chapter 101.

PEARLS

- Any adolescent male with significant and/or repeated nosebleeds should be evaluated for a juvenile angiofibroma.
- Preoperative embolization reduces operative bleeding.
- During an endoscopic transnasal resection, do not divide the juvenile angiofibroma within the infratemporal fossa because it will be more difficult to retract and dissect.
- Attention should be paid to the site of origin (medial pterygoid plate) and the vidian foramen because these are common places for recurrences.
- Follow-up should include a CT scan with contrast.

PITFALLS

- Biopsy of a juvenile angiofibroma will produce significant hemorrhage.
- Embolization may be associated with significant morbidity.
- Incomplete resection of the site of origin often results in a progressively growing recurrence.
- All surgical approaches have the potential to interfere with facial growth.
- Destruction of the nasal epithelium invariably leads to nasal crusting and subsequent scarring.