Chapter 97 – Orbital Exenteration

S. Tonya Stefko

PATIENT SELECTION

The radical surgery of orbital exenteration is occasionally necessary in the management of head and neck cancer. Most frequently for the otolaryngologist, it will be due to spread of malignant tumor from adjacent paranasal sinuses or the skull base. Some cancers arising in the skin will also require total or subtotal removal of the orbital contents. Invasive fungal infections are indications for aggressive débridement, sometimes including orbital exenteration. Rarely, a benign but locally aggressive tumor or intractable pain will be an indication.

The most clear-cut indications for orbital exenteration are those involving malignant tumors with no hope of salvaging vision in the affected eye (Fig. 97-1). These situations most often include squamous cell carcinoma and other epithelial malignancies. In the largest reported series of orbital exenteration, squamous cell carcinoma (originating in the sinus, skin, or conjunctiva) accounted for one third of exenterations.[1] A more recent series found basal cell carcinoma to be the most frequent diagnosis, followed by melanoma and sebaceous cell carcinoma.[2] Conversely, a review of 70 consecutive cases of maxillectomy found that 40% of these patients required concurrent orbital exenteration.[3] Imaging is likely to underestimate the extent of tumor invasion into the orbit, and the decision to proceed may need to be made in the operating room. The recent trend, particularly with squamous cell carcinoma, has been preservation of the orbit except in instances of proven invasion of the periorbita (by frozen section at the time of surgery). In several retrospective series there has been no difference in disease-free survival between patients treated with orbit-sparing procedures and those whose surgery included orbital exenteration.[4]

Figure 97-1 Patient with malignant melanoma destroying the globe, adnexa, and orbital contents.

Malignant tumor of the orbit or eyelids, although not strictly invading the eye or periorbita itself, may require removal of the orbital contents because of insufficiency of the remaining tissue for protection of the globe (Figs. 97-2 and 97-3).

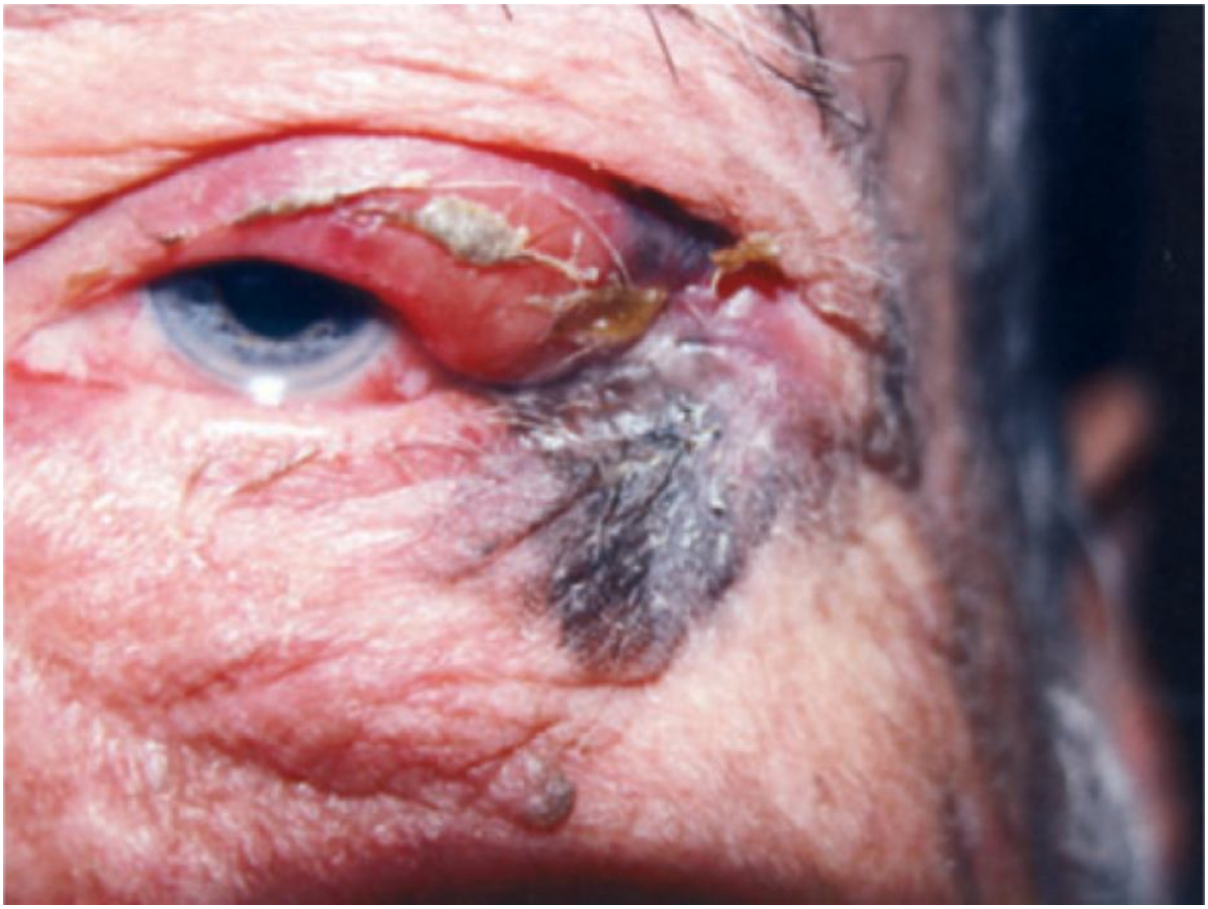

Figure 97-2 Advanced cutaneous melanoma of the eyelid with orbital extension.

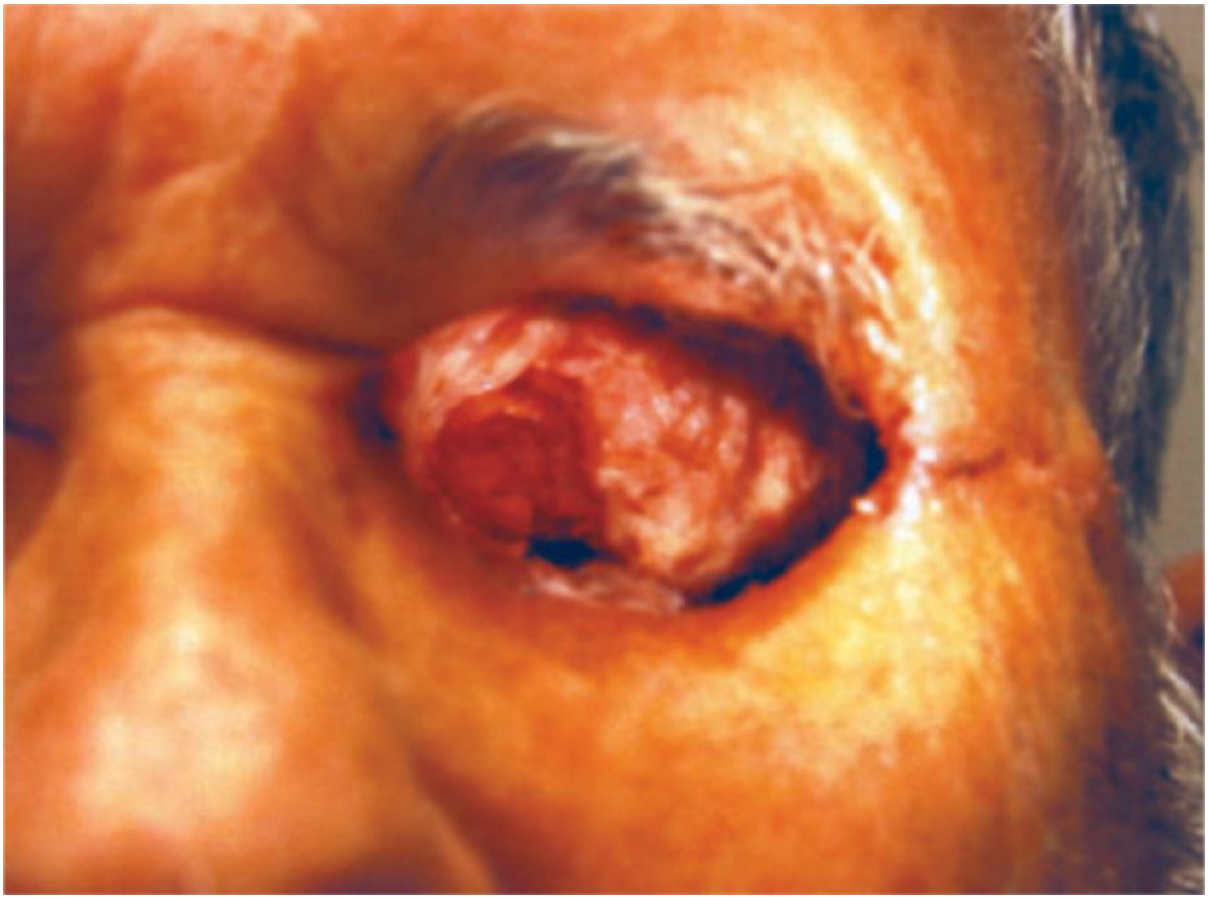

Figure 97-3 Patient from Figure 97-2, 12 days after exenteration with a split-thickness skin graft in the orbit.

Surgical decision making in the setting of aggressive infection is difficult. Most fungal infections of the orbit spread from adjacent sinuses, and two main types occur. *Aspergillus* affects both immunocompromised and immunocompetent hosts. In the former patients, the prognosis is extremely poor if immune status cannot be improved and if intracranial involvement is present.[5] In the latter setting, exenteration is likely to be necessary in cases of apical involvement, but not when the infection is confined to the anterior, inferomedial orbit,[6] where limited débridement may suffice.

Rhino-orbital-cerebral mucormycosis is a rare but serious infection caused by *Rhizopus* species (occasionally *Mucor* or *Absidia*). It affects immunocompromised patients, most often those with a concurrent metabolic acidosis. These patients require aggressive surgical débridement to the level of bleeding tissue. This has frequently included orbital exenteration in the presence of orbital signs, but one series showed no difference in survival between patients with orbital involvement who underwent exenteration and those who did not.[7] However, the overall mortality in patients with orbital involvement was higher (33% versus 14%). The operation may be followed by hyperbaric oxygen therapy, which improves survival.[8]

Three percent to 17% of patients requiring exenteration have benign orbital disease.[1,9–12] The various associated diagnoses include inflammatory disease, Stevens-Johnson syndrome, lymphangioma, meningioma, and others. These diseases are either painful, inadequately controlled medically, and “grossly disfiguring” or, although histologically benign, tend to invade adjacent tissue.[13] The pain associated with the disease most often resolves immediately after surgery.

PREOPERATIVE EVALUATION

Patients who may be candidates for orbital exenteration should, at the very least, be evaluated by computed tomography or magnetic resonance imaging (or both) of the orbits, sinuses, and brain. In the majority of cases that involve malignancy, positron emission tomography is also indicated. When neck dissection is anticipated,preoperative imaging of the neck and scintigraphy may be considered. The choice of imaging will be guided by the patient's diagnosis. In patients with malignancies contiguous with the orbit, it is likely that imaging will underestimate the extent of disease and intraoperative histopathologic monitoring will be necessary.

Before the operation, coagulation studies, a complete blood count with platelets, and typing and crossmatching of two units of packed red blood cells should be performed.

In many instances, given the radical nature of the surgery, pathology of the lesion should be independently confirmed by pathologists experienced in the field of head and neck or orbital disease.

Before scheduling surgery, a complete ophthalmologic examination of the remaining eye is indicated both to guide surgical decision making and to maximize the remaining vision.

In most otolaryngologists’ practices, exenteration will be undertaken in conjunction with maxillectomy or during aggressive sinus débridement (e.g., for invasive fungal infection). During the informed consent process, if the decision whether to exenterate the orbit will be made at surgery, it must be made very clear to the patient and family that although every effort will be made to preserve the orbit and eye, this goal will not preclude complete eradication of disease.

SURGICAL APPROACHES

Total or subtotal exenteration should be planned. Subtotal exenteration most often means a partially lid-sparing technique (Fig. 97-4A), which is appropriate in most cases of benign disease and with tumors that primarily affect the posterior or deep portions of the orbit. It is inappropriate in any tumors affecting the skin, conjunctiva, or lacrimal gland. Lid-sparing exenteration gives an excellent color match to surrounding skin and heals relatively quickly. Globe-sparing exenteration has been described, with purported advantages of easier prosthetic fitting later.[14]

Figure 97-4 **A,** Patient with mucoepidermoid cancer of the maxillary antrum who underwent orbital exenteration and radical maxillectomy. A lid-sparing incision is outlined. **B,** Postoperative appearance with the lids sewn together, thus eliminating view of the open orbit. **C,** Oblique view. **D,** Patient wearing special glasses that obscure view of the orbit.

Primary Reconstructive Options

Reconstructive options should be considered before surgery. The simplest option is healing by secondary intention, or granulation. The advantages of such healing are shorter operating time, good color match to surrounding tissue, excellent visibility for monitoring of possible recurrence of disease, and a tendency to produce a shallow socket (easier for eventual patient care).[15] Disadvantages include a long healing time and the necessity for several painful dressing changes.

Split-thickness skin grafts, most commonly from the thigh, are frequently used to line the orbital cavity when total exenteration with removal of periosteum is performed (see Fig. 97-3). Although this leads to shorter recovery time and excellent visual monitoring for recurrence, a deeper cavity is formed, for which care and prosthetic fitting are somewhat more difficult. The pain encountered after this reconstructive option is generally from the donor site.

Local or free tissue flaps are most commonly performed at our institution, particularly for defects resulting from concurrent partial or complete maxillectomy. Temporalis flaps,[16] with or without cervicofacial,[17] radial forearm,[18] and rectus abdominis flaps, are most frequently mentioned. These flaps have the advantage of ample tissue and good vascularity, which is desirable in a field likely to require radiation therapy. Disadvantages of longer operating time, donor site morbidity/disfigurement, and a requirement for highly specialized microsurgical techniques (in the case of free flaps) discourage their use in very sick patients. These options also render monitoring for local recurrence of disease more difficult.

It has been reported that most patients are likely to wear an occlusive eye patch regardless of attempts at reconstruction.[19] Prostheses may be fashioned by aprosthodontist and may be affixed by topical adhesive. Alternatively, prostheses may be fixed magnetically to osseointegrated dental implants.[20,21]

Surgical Procedure

The patient is placed under general anesthesia, and the surgical margins are marked (see Fig. 97-4A); in the case of concurrent maxillectomy, that procedure is performed first. A suture tarsorrhaphy is usually helpful. If the exenteration is to be a lid-sparing technique, an incision line several millimeters proximal to the eyelid margins and parallel to them is marked. A knife is used to cut through skin and orbicularis, and blunt dissection in the pretarsal/preseptal plane is then carried out to the level of the orbital rim. Periosteum is incised, and the surgery proceeds as described in the following text.

If the skin is to be included in the resection, the excision margins are drawn at the orbital rim and at least 1 to 2 cm from the cutaneous lesion. The marked areas may be infiltrated with local anesthetic with epinephrine to help with hemostasis. A no. 15 scalpel is used to cut through skin to the periosteum. Bleeding should be expected at the supraorbital vessels. The periosteum is elevated with a Cottle or Freer elevator, beginning at the superotemporal portion of the orbit where the bone is thickest. Dissection in this plane continues circumferentially and posteriorly, with cautery of the zygomaticofacial and zygomaticotemporal vessels encountered in the lateral wall. Resistance will occur in the area of the trochlea (superomedially) and at the canthal ligaments and may require sharp

dissection in these areas. The posterior limb of the medial canthal tendon should be elevated from bone, and the lacrimal sac should be divided at its junction with the nasolacrimal canal. The inferior oblique muscle originates just lateral to the sac and should be elevated with the periosteum.

Dissection proceeds posteriorly toward the orbital apex, with special care directed to the following areas: first, elevation of the periosteum over the lamina papyracea must be carried out very gently to not create dehiscences in the bone. The ethmoidal vessels must be divided and bleeding controlled with bipolar cautery and bone wax if needed. Second, areas of the roof, particularly posteromedially, are often quite thin or contain frank dehiscences. It is here that an iatrogenic cerebrospinal fluid leak may be created, either by direct trauma or by the use of monopolar cautery, which may travel through emissary foramina or dehiscences.[22] Monopolar cautery should be avoided except in the anterior-most portion of the orbit.

The contents of the superior and inferior orbital fissures and the apical stump must now be divided, which may be accomplished with a curved knife, curved enucleation scissors, or a snare. The latter is tightened to a soft stop and tightened sequentially over a period of several minutes to assist with hemostasis. The snare may then be tightened completely to transect the apical stump, or curved scissors may be used to cut just anterior to it. The orbit should immediately be packed with gauze and pressure applied for 5 to 10 minutes. The apex should then be explored cautiously with suctionand bipolar cautery to localize and control any bleeding.

If the orbit is to heal by granulation, it should now be lined with a nonadherent dressing such as Telfa followed by Xeroform gauze, packed with fluffed gauze, and bandaged tightly. A small pledget of cellulose (Surgicel) or other hemostatic agent may be placed over the apical stump.

It may be possible in some patients to preserve the eyelids. After a skin graft has been placed in the orbit, the margins of the eyelid are sewn together to provide a good appearance or at least a better appearance than if the orbit were open to view (see Fig. 97-4B to D).

If the lid skin has been spared, it is draped over the rim and spread posteriorly as far as possible. The remainder of the orbit may be left to granulate or may be covered with a split-thickness skin graft. If a split-thickness graft is used, it is harvested from a non–hair-bearing area of the inner surface of the thigh or the abdomen and the donor site covered with an adherent dressing. Dimensions of the graft are approximately 5–10 cm before meshing. The graft is carefully draped into the orbital cavity, with care taken to ensure that the epithelial side remains exterior. The graft is sutured in place at the rim with interrupted 7-0 chromic gut or silk suture. The wound is then dressed as discussed earlier.

If a local or free flap is to be used for reconstruction, it is prepared and placed as dictated by the specific procedure. The patient in Figure 97-5A underwent maxillectomy for sarcoma of the maxilla. A recurrence was treated with radiation therapy. The tumor recurred once again and destroyed the orbital contents and the orbital wall (see Fig. 97-5B). The orbital defect (see Fig. 97-5C) was reconstructed with a rectus abdominis free tissue transfer (see Fig. 97-5D).

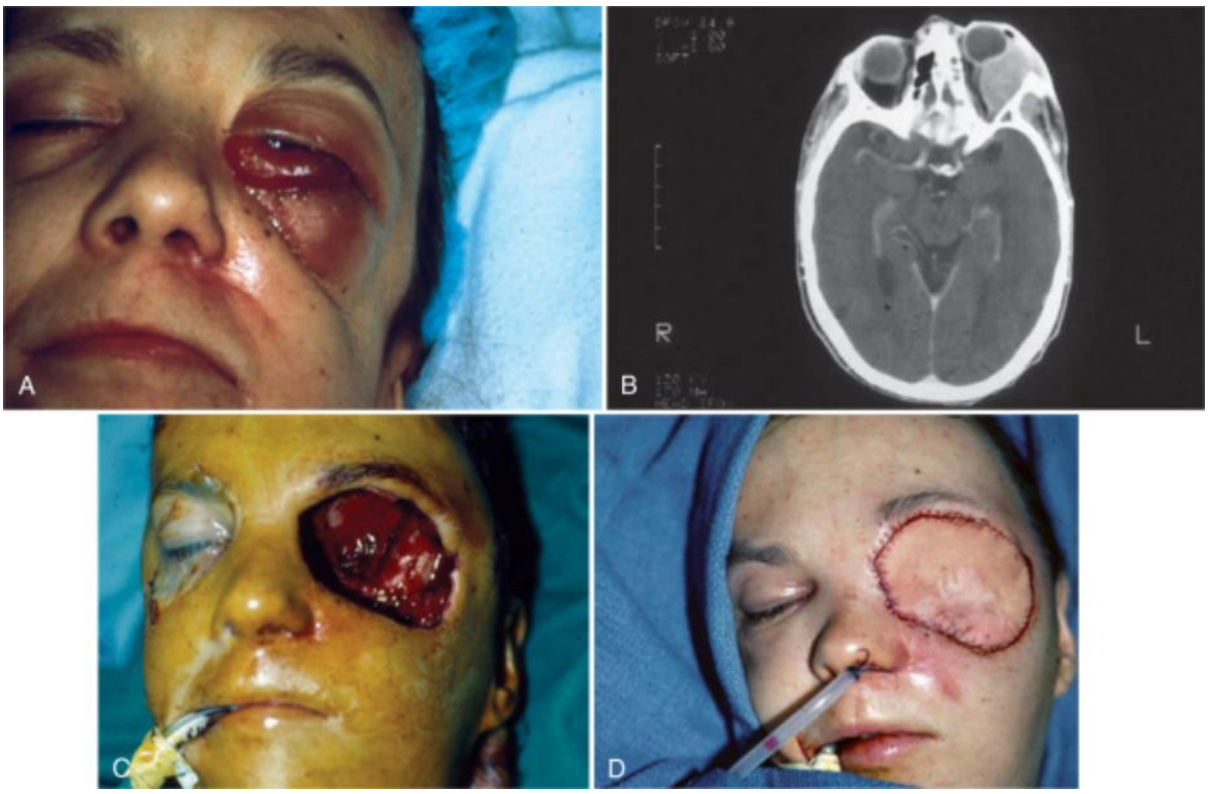

Figure 97-5 **A,** Patient with a sarcoma of the maxilla treated by maxillectomy. Recurrence in the orbit was treated with radiation therapy. The tumor recurred once again and destroyed the orbital contents. **B,** Computed tomography scan demonstrating invasion of the orbit and destruction of the bony orbital wall. **C,** Defect after orbital exenteration. **D,** Reconstruction with a rectus abdominis free tissue transfer.

When flap reconstruction is not used, the pressure dressing remains undisturbed for 3 to 5 days. During this time the patient should have antibiotic coverage. After this time, the pressure dressing is gently removed, and the area is soaked with saline. After 5 to 10 minutes, the lining layer (nonadherent dressing such as Telfa or Xeroform) may be gently loosened from the underlying tissue. Minimal débridement should occur at this stage because areas of skin that appear devitalized may heal surprisingly well. The area is coated lightly with antibiotic ointment, and the patient and family are instructed to repeat the application of ointment twice daily. Ointment should also be applied to the light dressing placed over the orbit to prevent adherence. Vigorous cleaning of the area should be avoided.

The patient is examined in the office 1 week postoperatively and then at regular intervals afterward as dictated by disease process. Weekly or biweekly visits are appropriate in the first 4 to 6 weeks for cleaning of the socket and monitoring for infection. In most cases in which radiation therapy is not necessary, the patient is ready for prosthesis fitting in 3 months (Fig. 97-6).

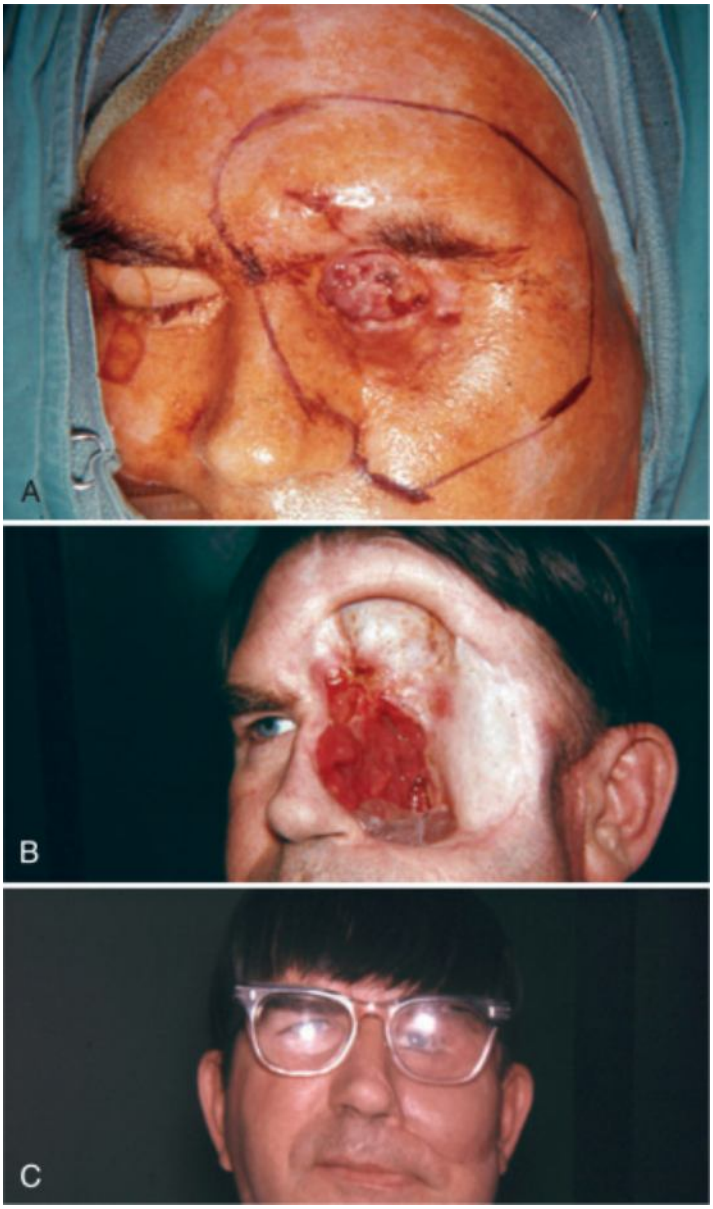

Figure 97-6 **A,** Patient with recurrent basal cell carcinoma in a previously exenterated orbit. **B,** Postoperative defect. **C,** Postoperative appearance with a prosthesis and glasses.

COMPLICATIONS

Possible complications of the operation include intraoperative bleeding, postoperative hematoma, postoperative infection, and dehiscence of the surgical wound. Intraoperative bleeding is treated by packing and pressure, prothrombotic agents applied to the orbital apex, and transfusion of packed red blood cells if necessary. Postoperative hematoma requires therapy if it occurs under a skin graft or in a free flap. The former may be drained by making a small incision in the graft, gently evacuating the hematoma, and then replacing the pressure dressing for 2 to 5 days. Free flaps must be monitored carefully by Doppler ultrasound to ensure patency of the artery and vein, and a pressure dressing is contraindicated. Postoperative infection must be vigorously treated with intravenous and then oral antibiotics, with judicious surgical débridement of devitalized tissue or drainage of purulent collections as needed. Dehiscence of the surgical wound should raise suspicion of infection, which must be treated aggressively before considering secondary repair.

Fistulas resulting from orbital exenteration may communicate with either the sinuses or dura. If a bone or dural defect is noted intraoperatively, it should be repaired. If the orbit is to heal by granulation or a skin graft is used, a small flap of extraocular muscle and fat may be spared and used to cover either the bone window or the dural repair.[23]

PEARLS

- Orbital exenteration is appropriate for patients with tumor invasion of periorbita proved by frozen section examination.
- Patients with invasion of orbital bone only will probably have comparable survival if the orbital contents are spared.
- In patients with malignant tumors of the lacrimal gland, exenteration is the procedure of choice.
- Aggressive débridement of skin grafts early in the postoperative period is to be avoided.
- Postoperative infection must be treated early and completely.

PITFALLS

- Sino-orbital fistulas are most easily created over the lamina papyracea and result in chronic discharge.
- Use of monopolar cautery in the deep or superior aspect of the orbit is to be avoided because cerebrospinal fluid leak may result.
- Resection of most of the orbital floor frequently leads to loss of integrity of the eye because of dystopia, diplopia, or ocular surface problems. This may be compounded by postoperative radiation effects on the eye.[24]