Chapter 8
Exenteration and Multidisciplinary Approaches

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Indications

Orbital exenteration is most commonly performed with the goal of achieving local disease control in malignancies and severe infections. Examples of primary orbital tumors that may lead to exenteration include adenoid cystic carcinoma, rhabdomyosarcoma, retinoblastoma, and mucoepidermoid carcinoma [1–8]. Eyelid or ocular adnexal tumors, such as melanoma, basal cell carcinoma, squamous cell carcinoma, and sebaceous cell carcinoma, as well as sinonasal tumors such as esthesioneuroblastoma may also lead to exenteration in cases of extensive invasion into the orbit [1–8]. In case of malignancies, the inherent characteristics of orbital anatomy and orbital tissues make it challenging to obtain distinct tumor margins for complete resection once intraorbital tumor infiltration is present. In such cases, exenteration may be the only viable option for ensuring complete resection and for decreasing the risk of metastasis [1–4]. When advanced metastatic disease is already present, exenteration is sometimes offered as a palliative measure to relieve intractable pain of significant mass effect [1–8].

Local disease control via exenteration for nonmalignant condition is indicated in severe life-threatening orbital fungal infections such as *mucormycosis* and aspergillosis with risk of intracranial extension [1–8] and rare cases of severe periocular necrotizing fasciitis (group A β-hemolytic *Streptococcus*) unresponsive to medical therapy and debridement [9]. Congenital deformities or benign tumors and trauma with severe disfigurement and irreversible vision loss may also be candidates for exenteration [10, 11].

For all of the above malignant and nonmalignant conditions, exenteration is a radical surgery that is offered as a last-resort option. Needless to say, the permanent

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vision loss and facial disfigurement come with significant physical and emotional burden for the patient and his or her family. In cases of some elderly or severely debilitated patients with complex medical comorbidities or limited life expectancy, a realistic discussion regarding the expected benefit of exenteration for the particular pathology in question should be held.

Preoperative Evaluation

Every patient must be evaluated with a complete ocular examination. Special attention is directed to the eyelid and adnexal structures, including the nasolacrimal system and adjacent sinonasal anatomy depending on the underlying etiology. Any lymphadenopathy or facial sensory deficits can provide additional clues with regard to the extent of disease. These findings can be confirmed on orbital imaging with CT and/or MRI. If not already available at this stage, an incisional biopsy is obtained for a definitive pathological diagnosis on permanent section prior to consideration of subsequent steps.

A multidisciplinary approach may be needed if involvement of the intracranial space or sinonasal cavity is noted. A combined approach with neurosurgery and/or otolaryngology is sometimes chosen to allow for complete resection of adjacent structures in addition to orbital exenteration. For malignant lesions, the team may also include radiation oncology and medical oncology for coordination of neoadjuvant or adjuvant chemotherapy and radiation. For infectious etiologies, tailored medical therapy guided by infectious disease specialists is essential.

A comprehensive preoperative evaluation can determine the extent of exenteration to be performed: total, eyelid-sparing, or subtotal. Diffusely infiltrating disease involving significant areas of eyelid skin and conjunctiva, as well as orbital structures, is best treated with total exenteration. A posteriorly located disease with no surface or skin involvement, on the other hand, can be approached via the eyelid-sparing technique. Rarely, pathology with more localized involvement of the orbit can be treated with subtotal exenteration with partial removal of orbital contents.

Operative Technique

Antiplatelet and anticoagulation agents are discontinued prior to surgery in the absence of major medical contraindications.

- *Incision: total exenteration vs. eyelid-sparing exenteration*

Under general anesthesia, an injection of local anesthetic with epinephrine (i.e., 2% lidocaine with 1:100,000 epinephrine) is given for hemostasis. A tarsorrhaphy

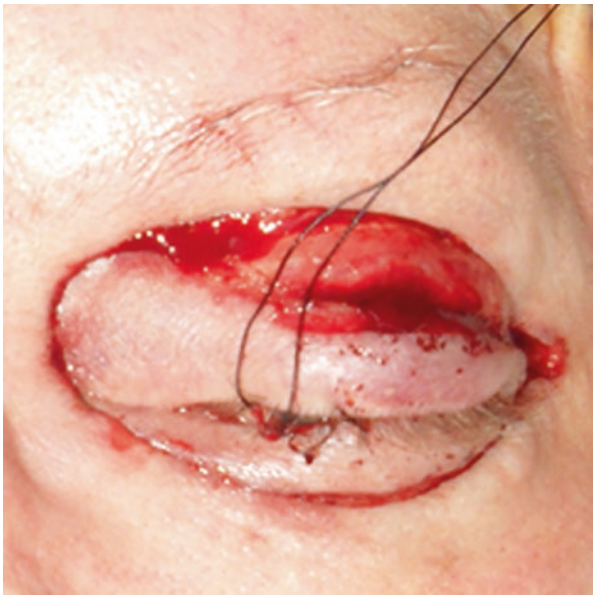

Fig. 8.1 Intraoperative photo demonstrating suture tarsorrhaphy and incision for a partial eyelid-sparing exenteration

can be placed through the central lid margins to help with traction. For total exenteration, a #15 blade or monopolar cutting cautery is used to incise the skin 2–3 mm outside the arcus marginalis down to periosteum. In an eyelid-sparing exenteration, the skin incision is made outside the upper and lower eyelid lash lines (Fig. 8.1). Dissection is then carried out in either the pre-orbicularis plane or the pre-tarsal plane toward the orbital rim, which is then incised as above.

- *Dissection*

A subperiosteal dissection is carried out with a Freer elevator toward the orbital apex. The extent of the posterior dissection depends on the location of the primary pathology (subtotal vs. total exenteration). Controlled, gentle dissection is used near the delicate medial wall, inferomedial floor, and the neurovascular bundles (supraorbital, supratrochlear, anterior and posterior ethmoidal, zygomaticofacial, and zygomaticotemporal), in combination with sharp dissection around the firm attachments of medial and lateral canthal tendons, trochlea, lateral orbital tubercle, and the origin of the inferior oblique muscle. The nasolacrimal sac-duct junction and the supraorbital and infraorbital fissure contents are transected. Dissection in the area of primary pathology is typically performed last in order to allow for uncomplicated dissection of all other areas and improved access and visualization of the area of interest, which may involve bony erosion or abnormal anatomy. Bleeding during the dissection can be controlled with cautery, bone wax, vascular ligation clips, or prohemostatic agents.

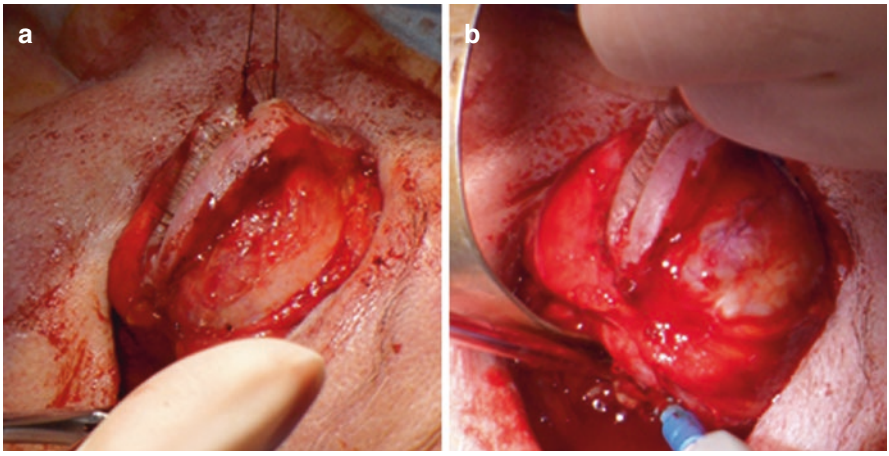

Fig. 8.2 (**a**) A large hemostat placed around the apical structures of the orbit following complete subperiosteal dissection. (**b**) Monopolar cutting cautery is used to transect the apical tissues above the hemostat

- *Removal of orbital contents*

Following adequate dissection, one or two large hemostats are placed posteriorly across the apical structures (Fig. 8.2a). It should be noted that some patients may develop significant bradycardia during this maneuver, and the anesthesiologist should be informed ahead of time for appropriate monitoring. Using scissors or monopolar cutting cautery, the apical tissues are transected above the hemostat and the orbital contents removed in toto (Fig. 8.2b). Any arterial bleeding from the ophthalmic artery is controlled with cautery or surgical clips. If posterior extension of the main lesion or pathology is suspected, intraoperative frozen sections of the remaining apical tissues can be taken to clear the margin. Concurrent bony resection may be necessary in the setting of bony erosion, which may involve a multidisciplinary approach (e.g., craniotomy, maxillectomy).

- *Reconstruction of exenterated socket*

A number of techniques are available for primary reconstruction of an exenterated socket: healing by secondary intention, skin graft, dermal substitute, myocutaneous advancement flap, or free flap [8, 12–14].

Healing by Second Intention

Healing by second intention is the simplest option in which the exenterated socket is left to granulate on its own. At the end of surgery, the bare bone of the exenterated socket is covered with iodoform gauze and antibiotic ointment. This dressing is then

changed on a regular basis (every 2 days) as granulation tissue forms and slowly epithelializes over 3–4 months. Despite perhaps having the shortest surgical time and good tissue color match with the rest of facial skin, the surface is typically irregular and can mask recurrence in cases of malignancy. The lengthy healing time is also generally uncomfortable for the patient and too long for those in need of adjuvant radiation.

Split-Thickness Skin Graft

Lining the exenterated socket with split-thickness skin graft allows for faster healing and a smooth surface [12]. Non-hair-bearing skin of the thigh is a commonly used donor site. A dermatome is used to harvest an even-thickness (0.3–0.6 mm) 3 × 5 inch graft. The split-thickness skin graft is then fed through a 1:1 or 1:2 ratio mesher, which cuts the graft in a mesh-like pattern allowing it to be stretched to cover a larger surface area. The perforations also serve as premade vents that prevent formation of hematomas under the graft (Fig. 8.3a, b). The graft is molded to the socket and sutured to the edges of the circumferential incision around the orbit with interrupted and running dissolvable sutures. Complete contact between the graft and the recipient bed is ensured by lining the graft with antibiotic ointment, non-adherent dressing, and pressure packing (e.g., surgical sponges, gauze, iodoform dressing). A firm pressure patch is fashioned over the packing and maintained for 5–7 days. The donor site for the skin graft is covered with non-adherent dressing.

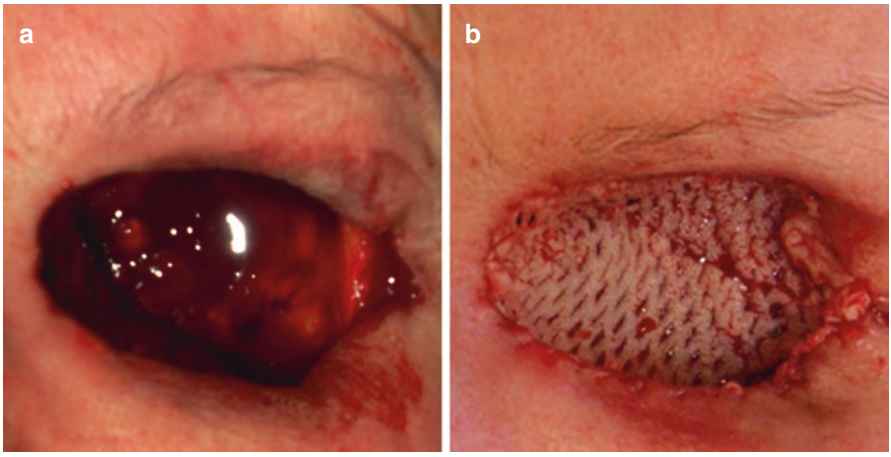

Fig. 8.3 (**a**) Exenterated socket following removal of orbital contents. (**b**) A split-thickness skin graft lining the exenterated socket

Dermal Substitute

Using a commercially available dermal substitute avoids the need for a skin graft and the associated donor site morbidity. An example of such product is an acellular extracellular matrix scaffold made of biodegradable cross-linked bovine tendon collagen and glycosaminoglycan layered on a silicone membrane (Integra®) [13, 14]. The dermal substitute is positioned over the exenterated socket and sutured to the skin edges. The scaffold provides a supportive environment for rapid granulation and epithelialization. The silicone membrane is removed by 3–4 weeks, and healing is typically complete by 6–8 weeks. While effective, the major limitation of such product is the cost.

Myocutaneous Advancement Flap or Free Flap

When exenteration is combined with concurrent resection of adjacent structures (e.g., maxillectomy) leading to large soft tissue and bony defects, there may not be any contiguous bony surface to cover with a skin graft or dermal substitute. In such cases, myocutaneous advancement flaps or free flaps are used to cover the resection bed [7, 8]. Options for local transposition flaps include the temporalis, frontalis, galea-frontalis flap, and the temporoparietal fascial flap [7, 8]. Some commonly utilized free flap sources include the radial forearm, latissimus dorsi, rectus abdominis, lateral arm, or anterolateral thigh [7, 8]. In contrast to reconstructive strategies discussed above, exenterated sockets covered by flaps cannot be fitted with a prosthesis and typically have a bulky, less aesthetically satisfactory outcome. The relative thickness of the flap can also mask tumor recurrence, and close surveillance via imaging is therefore needed.

Potential Complications

Potential complications of exenteration may arise from specific surgical techniques as well as from the nature and extent of the underlying pathology. Some of the most notable complications include hematoma formation, postoperative infection, cerebrospinal fluid leak, sino-orbital fistula, and recurrence.

Hematoma Formation Sources of bleeding include the ophthalmic artery, ethmoidal arteries, and neurovascular bundles coursing through the superior and inferior orbital fissures and other perforating foramens. Hematoma can form under a skin graft or a flap in the setting of poor hemostasis and cause graft failure or delayed healing. The hematoma should be promptly evacuated, the source of bleeding controlled, and any necrotic tissue debrided. Limited areas of graft dehiscence or flap failure can be left to heal by secondary intention, but larger areas may require repeat grafting.

Postoperative Infection True surgical site infection is rare with appropriate sterile technique. In cases of an underlying infectious etiology leading to exenteration (invasive fungal or bacterial infection), concurrent local and/or systemic administration of antifungal or antibiotic agent is usually given. If frank purulence is noted in the exenterated socket, gram stain and cultures should be obtained to guide therapy and any necrotic tissue debrided. A povidone-iodine wet-to-dry dressing can be applied for further debridement and wound care.

Cerebrospinal Fluid (CSF) Leak Penetration of dura with subsequent CSF leak can happen via the cribriform plate, orbital roof, or the greater wing of the sphenoid. Small CSF leaks can close spontaneously, but larger leaks may require direct repair using tissue graft and/or tissue adhesive in conjunction with neurosurgery.

Sino-orbital Fistula Full thickness penetration of orbital walls with direct communication into the adjacent sinuses can result in fistulas. The defect in the orbital wall can be from unnecessarily forceful dissection at the time of surgery, direct bony erosion from the primary pathology, or from tissue breakdown secondary to adjuvant radiation therapy. While small fistulas can be observed and the edges allowed to granulate, large fistulas can lead to chronic discharge from the sinus mucosa and difficulty with breathing and phonation. Options for closure of fistulas include additional skin graft or vascularized tissue flap.

Recurrence In cases of malignancy, the risk of tumor recurrence secondary to positive surgical margins or residual areas of microscopic disease requires ongoing surveillance for all patients. While a thorough margin clearance via frozen sections at the time of surgery decreases the potential for this complication, it does not guarantee complete tumor clearance or prevent microscopic metastasis (Fig. 8.4).

Postoperative Care

Appropriate postoperative care of an exenterated socket depends on the method of reconstruction used. For healing by secondary intention, the socket can be packed with wet-to-dry dressing of 4 × 4 gauze soaked in 10% povidone-iodine solution to allow for debridement of dried blood and keratin debris, while the granulation tissue forms over the bony surface. As noted above, this can be a lengthy process that requires daily wound care by the patient or the caregiver over 3–4 months.

If a split-thickness skin graft was placed, the pressure packing can be removed 5–7 days after surgery and a wet-to-dry dressing with a 50:50 mixture of 10% povidone-iodine solution and hydrogen peroxide initiated. The frequency of dressing change is gradually decreased until the socket is fully epithelialized. Dermal substitute-based reconstruction should be managed according to the manufacturer's instructions for optimal healing. In the setting of adjuvant radiation therapy, the socket will be more prone to epithelial breakdown, and additional dressing change or treatment with antibiotic ointment and topical emollient may be necessary.

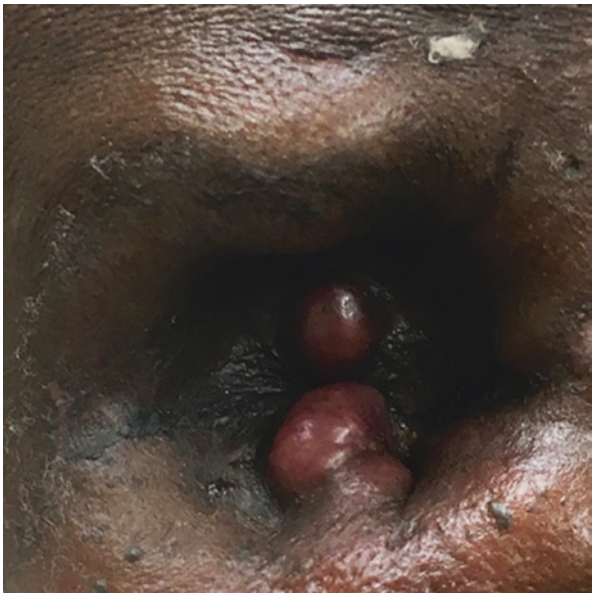

Fig. 8.4 Patient with a history of eyelid-sparing exenteration for invasive squamous cell carcinoma presenting with large nodular growths, concerning for recurrence

In cases of myocutaneous advancement flap or free flap, maintaining flap perfusion and survival is the primary goal of postoperative management. Early detection of any hypo-perfused areas with timely debridement of any necrotic tissue is recommended.

After the socket has been completely epithelialized, the patient can be fitted with an oculofacial prosthesis. The traditional method of prosthesis construction involves taking an impression of the orbit with a plaster cast to make a mold, with which a silicone template is made and painted to resemble the other eye. The prosthesis can be attached to thicker glasses frame to help camouflage the prosthesis-skin interface or directly attached to periorbital skin with skin adhesive or magnetic posts (see chapter on osseointegrated prosthesis) [15]. More recently, advancements in 3D-printing technology are being applied to making custom-printed prostheses at a lower cost than the traditional methods. Along with the prosthesis, all patients are required to wear polycarbonate glasses to protect the remaining eye, with lifelong monocular precautions.

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