A rare presentation and challenging reconstruction of an extensive cystic orbital schwannoma

Abstract

A 57 year-old woman presented an irritated right eye and right-sided enophthalmos with hypoglobus for the past year. The patient previously had an extensive right-sided cystic orbital schwannoma removed with orbital reconstruction 5 years ago. Imaging revealed an enlarged right orbit with medial wall collapse and inferior displacement of a floor plate implant. An anterior orbitotomy with placement of an enophthalmos implant, three 1 mm Medpor sheets, an orbital rim implant, and an oral mucosa graft in inferior fornix with a lateral strip were used to reconstruct the ocular malposition and right lower lid retraction. We report a successful case of a complex orbital reconstruction in a patient with orbital deformity subsequent the removal of a massive cystic schwannoma.

Key Words schwannoma, orbital reconstruction, enophthalmos, orbital implant, lid retraction

Introduction

Primary orbital schwannoma (POS) comprises 1% of all orbital tumors and are typically slow growing and benign. They arise from the peripheral nervous system and are asymptomatic when small but if large can present as an anterior orbital mass with optic neuropathy, proptosis, diplopia, and sinusitis. Massive tumors involving the orbit are often in close proximity to the cranium and sinuses, requiring extensive and meticulous dissection along with challenging reconstruction. This case highlights an extensive POS and the subsequent reconstruction after primary intervention by neurosurgery and otolaryngology.

Case Report

A 57 year-old woman presents with an irritated right eye surface discomfort, severe right enophthalmos, and hypoglobus evaluation. Her past medical history is significant for a massive well-circumscribed right orbital cystic schwannoma, that extended from the right cavernous sinus into the superior orbital fissure superiorly, the foramen rotundum inferiorly, and through the lamina papyracea into the sinuses medially (Figure 1a). Surgical removal of the tumor with only partial orbital reconstruction was performed 5 years prior with no evidence of tumor recurrence; unfortunately the patient’s right eye went blind before surgery. On examination, the right eye had no light perception, a relative afferent pupillary defect, and optic nerve atrophy. There was significant ocular surface dessication, in keeping with a 10 mm lagophthalmos and cicatricial lower lid retraction. In addition, she had a large angle right sensorial exotropia, as well as limitation in adduction and elevation of the right globe. Naugle exophthalmometry measurements revealed 13mm on the right and 28 mm on the left, with 7mm right hypoglobus. Extraocular movement and anatomical position was normal in her left eye.

An orbital CT and MRI showed a partially reconstructed right orbit displaying significant orbital volume discrepancy due to an enlarged and disrupted orbit. Orbital abnormalities included: a metallic mesh in the right frontal bone after the craniotomy resulting in a disrupted roof, a posterior displaced lateral orbital rim with most of the greater wing of the sphenoid absent (Figure 1b), a metallic mesh acting as the orbital floor was placed lower than the anatomical position of the left orbit, and a collapsed medial orbital wall.

A management plan was designed in a step-wise fashion and the patient underwent four surgical procedures to address the different problems. Globe position was initially addressed by an anterior orbitotomy with an inferior fornix approach. A large right enophthalmos Medpor implant (22x31x7mm) was inserted along with three trimmed 1mm thickness Medpor sheaths positioned between the enophthalmos implant and the previously reconstructed metallic floor mesh (Figure 2a-b). Care was taken to ensure no worsening of the restriction on the elevation of the globe. Post-operative exophthalmometry readings...


Discussion

POS are rare tumors and their origin can be identified only half the time, arising from the branches of the oculomotor, trochlear, trigeminal, and abducens nerves, as well as, from sympathetic and parasympathetic fibers, but most originate from first division of the trigeminal ganglion (supratrochlear and supraorbital branch). Clinical presentation is often dictated by tumor size and includes: slowly progressive proptosis, globe displacement, dysfunction of ocular motility, and diplopia. Progressive growth can cause compression of the optic nerve with papilledema or optic atrophy. Although mostly benign, POS can cause bone erosion with widening of the superior orbital fissure, expansion of the orbit, as well as destruction of the orbital wall invading the maxillary sinus and nasal cavity in the case of local extension.

CT and MRI provide useful information on tumor size, characteristics, location within the orbit and extension into neighboring tissues. CT normally shows a round, encapsulated, well-defined hypodense lesion. However, cystic degeneration is not uncommon in long standing lesions. MRI is generally limited by anatomical variation of the lesion, but has use for soft tissue delineation and changes in the lesion over time. Ultimately, and as most tumors in the orbit, the key in the diagnosis is the histopathology confirmation. Classically, the tumor shows two distinct patterns of spindle cell arrangement: the solid and cellular Antoni type A, and the less cellular Antoni B pattern with myxoid areas. Degenerative cystic changes in old lesions are secondary to coalescence of mucinous or microcystic areas in a predominance of Antoni B tissue.

The treatment of choice POS is the complete surgical resection, however, equally of importance is the reconstruction. The presentation of this case is unique because of the wide involvement of several anatomical structures of the orbit and ocular adnexa. Our patient was blind in the right eye after the initial tumor resection which subsequently led to a large angle sensory exotropia. Furthermore, a limited reconstruction resulted in an enlarged orbit positioned posteriorly and that left significant lower lid scarring producing lid retraction and lagophthalmos. The challenging and intricate reconstruction required consecutively staged surgeries led to a remarkable cosmetic outcome and resolution of the enophthalmos and the malposition of the eyelids and globe (Figure 3).

Our case focuses on the presentation of an extensive POS and the critical importance of subsequent reconstruction of the orbit and ocular adnexa after primary surgical intervention. Management POS should involve an ophthalmologist, and ideally an orbital specialist, who can address and manage symptomatic and cosmetic complications that may arise after tumor resection. A multi-disciplinary approach in the treatment of POS would maximize treatment results and avoid the need for future reconstructive measures.

Figure 2: (a) A coronal CT after her first orbital surgery, showing a Medpor enophthalmos implant and three Medpor sheets piled on top of it on the right orbital floor (arrow). (b) An axial CT showing a Medpor enophthalmos implant in the right orbit. (c) Intra-operative image of the orbital rim Medpor implant insertion in the patient’s third reconstructive surgery.

Figure 3: Initial presentation of the patient to demonstrating severe right enophthalmos, exotropia, and hypoglobus. Post-operative photo, one week after her last surgery, showing a good cosmetic result and resolution of orbital and lid abnormalities.