Management of a blinding orbital apex cyst in a 14-year-old girl

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ABSTRACT

This study was conducted to highlight the diagnostic and management challenges of orbital apex lesions. It is a retrospective report of a 14-year-old female who presented with no perception of light vision in her left eye following a 1-year history of left unilateral axial proptosis. Her computed tomography scan revealed a mass surrounding the optic nerve which was reported to be an optic nerve glioma. She had lateral orbitotomy and a cystic mass was dissected and excised. Systemic steroids and antibiotics were administered. Visual recovery was achieved in the immediate postoperative period. At discharge, her visual acuity was counting finger in the left eye. Visual recovery in the orbital apex lesions is possible if the patient is diagnosed early and managed promptly but delayed presentation and diagnostic challenges could lead to irreversible blindness.

Key words: Blindness, cyst, orbital apex, visual recovery

INTRODUCTION

The bony orbit is a pyramidal structure housing the eyeball and the rest of the orbital soft tissues.

It has a base anteriorly and the apex posteriorly. The bony orbit is composed of fairly strong rim and walls including the inferior wall referred to as the orbital floor, the medial wall, the superior wall referred to as the orbital roof, and the lateral wall. The volume of the orbit is about 30 ml and the anterioposterior diameter of the orbit is between 40 and 50 mm. The base of the orbit has a height of about 35 mm and a width of about 40 mm.¹

The orbital apex is the posterior end of the bony orbit and it is composed of a number of openings including the optic foramen, the superior and inferior orbital fissures which transmit quite a number of vital structures. The optic nerve passes through the optic foramen to the eyeball while the superior orbital fissure transmits the lacrimal nerve, the frontal nerve, the trochlear nerve, and the superior ophthalmic vein.¹

CASE REPORT

A 14-year-old female presented with left axial proptosis and loss of vision of 1-year duration [Figure 1]. The visual acuity in the left eye was no perception of light (NPL) while that of the right eye was 6/6 unaided. Her exophthalmometric readings were 14 mm by 22 mm in the right and left eyes, respectively, on the base of 102 mm. There was left exotropia of about 45° (Hirchberg’s staging) and computed tomography (CT)-scan revealed a cystic mass behind the eyeball but within the muscle cone [Figure 2]. The radiologist’s report suggested a cystic mass to rule out either dermoid cyst or optic nerve glioma. Exploratory supero-temporal orbitotomy was done [Figure 3] and a huge intraconal cystic mass was encountered but got ruptured in the process of removal with mainly serous fluid release. The walls of the cyst were excised in pieces and sent to the laboratory for histology. A thorough lavage with normal saline was done and the wound was closed with a drain in place and the eye padded for 72 h. The visual recovery in the immediate postoperative period. At discharge, her visual acuity was counting finger in the left eye. Visual recovery in the orbital apex lesions is possible if the patient is diagnosed early and managed promptly but delayed presentation and diagnostic challenges could lead to irreversible blindness.

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The patient was placed on systemic prednisolone 20 mg BID with ciprofloxacin 50 mg BID for 10 days. The sutures were removed after 6 days and patient was discharged home. Visual acuity in the left eye immediately at the opening of the pad after 72 h was counting finger at 1 m unaided. The histology report revealed features suggestive of dermoid cyst. At follow-up visit after 2 weeks, the vision improved to 6/36 unaided with minimal proptosis. Further follow-up 12 months later revealed vision had improved vision to 6/9 unaided with no proptosis or exotropia [Figure 4].

**DISCUSSION**

Lesions in the orbital apex invariably cause severe visual morbidity and esthetics depending on the size, duration of the lesions, the type of lesions, and the structures involved.¹ Some of the structures involved in pathologies of this area include the 3rd, 4th, 6th cranial nerves, the lacrimal and frontal nerves. Other vital structures in this area include the superior ophthalmic vein and the optic nerve.¹ Depending on the mass effect of the tumor in this area, various presentations are possible ranging from nerve palsies to visual loss. Compression of the optic nerve by tumors in the orbital apex will ultimately lead to blindness.² Some studies carried out have suggested the possibility of spontaneous visual recovery perhaps depending on the level of visual affectation when the patient presents.³ In our case report, the lesion was cystic and quite huge in size with presenting visual acuity of NPL. The lesion was a dermoid cyst which could have been there for years before attaining such a size (almost larger than the eyeball size) with ultimate NPL vision.

The orbital apex is a site that is predisposed to a number of lesions which will ultimately constitute the orbital apex syndrome. Included are (1) head and neck tumors such as nasopharyngeal carcinoma, adenoid cystic carcinoma, and squamous cell carcinoma. (2) Neural tumors such as neurofibroma, meningioma, ciliary neurinoma, and schwannoma. (3) Metastatic lesions from the lung, breast, renal cell, and malignant melanoma. (4) Hematologic tumors such as the Burkitt’s lymphoma, non-Hodgkin’s
The early diagnoses of orbital apex lesions pose a great challenge as a myriad of pathologies find their way to this area. The lack of the essential tools to diagnose the lesions particularly in the third world makes it even more challenging. Clinical assessment including detailed history, demography and clinical presentation, laboratory investigations including the full blood count and differentials together with radiological imaging techniques can go a long way in assisting in making prompt diagnosis of orbital apex lesions. In the history, it is possible to know how the disease started and progressed, the duration of the disease the accompanying symptoms such as pain, loss of sensation on the face, variability during and/or after exertions or sleep, and the age and sex of the patient. Clinical examination will reveal the nature of the disease including the presence of proptosis, the type of proptosis whether axial, nonaxial, whether pulsatile or not whether with visual impairment or not, whether there is relative afferent papillary defects, color desaturations, exposure keratopathy or not, and whether there is a loss of corneal sensation or not. The laboratory investigations including hemogram will be able to reveal whether or not there is an associated inflammatory component, parasitic infestations, lymphoproliferative diseases, or otherwise. It is worthy of note that the dermoid cysts are by far the most common orbital cysts in childhood, while the inflammatory cysts due mainly to parasitic infestations are common in the tropics.

The availability of high-resolution magnetic resonance imaging (MRI) will aid in resolving the nature of the orbital lesions and where injury with fractures of the bony orbit is suspected or MRI is contra-indicated the CT-scan will suffice. Surgical management of orbital apex tumors also poses great challenge. The space is tight and the structures here are very vital. Meticulous dissections are a must! The surgical option for lesions in very tight spaces such as the orbital apex will require advanced endoscopic instruments to overcome the challenges of visualization and reaching of such embedded tumors. Gamma knife radiosurgery has revolutionized surgery in such tight spaces as well. The gamma knife radiosurgery uses multisource gamma ray emitter that is able to focus accurately on the targets in concealed spaces like the intracranial tumors and the other spaces and thus replacing the need for open surgeries. In our own situation, we did not have such refined equipment and had to contend with bloody surgery and rupture of the cystic lesion though with a favorable visual and esthetic outcome!

**SUMMARY AND CONCLUSION**

The orbital apex is a sensitive space with a number of vital structures! Any careless assault surgically on this space will result in grave morbidities particularly visual loss in view of the optic nerve and other vital structures. The space also attracts a lot of pathologies including benign and malignant tumors. Prompt diagnoses and careful surgical removal of such tumors will go a long way to avoiding the aforementioned morbidities. The challenges in our environment however include inadequate availability of the high-tech diagnostic and surgical tools. We therefore recommend that there should be collaboration with the surgeons and equipment companies in the developed world in the area of capacity building both in terms of manpower training and making the high-tech equipment available at subsidized rates.

In our case, however, the NPL vision and the exotropia were reversed after the surgical intervention. This is probably due to the timely surgical intervention. We advise also in conclusion, therefore, that all orbital apex tumors be immediately operated upon irrespective of the presenting visual acuity.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

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