Case Report

A Misleading Orbital Cystic Lesion

Shruthi Tara, Shraddha Shah

Department of Orbit and Oculoplasty, Sankara Eye Hospital, Coimbatore, Tamil nadu, India.

Abstract

Orbital schwannomas are rare and arise from the Schwann cells of sensory nerves. Cystic degeneration has been known to occur with orbital schwannomas but those occurring intra-ocularly have not been reported till date. Authors here report a rare case of a 24 year old woman, blind in the right eye, presenting with right sided headache and progressive proptosis since four years. Computer tomography (CT) of right eye showed presence of a well-demarcated, retro-orbital cystic lesion with a solid component causing displacement of the globe-like structure and bony remodelling. From the clinical and radiological features a pre-operative diagnosis of a right congenital cystic eyeball and a left microphthalmos with cyst was made. The patient underwent enucleation of the right eye along with the cystic lesion as a whole. Based on histopathology, a final diagnosis of an orbital schwannoma with extensive cystic degeneration and intra-ocular extension was made

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Introduction

Schwannomas are benign tumours arising from schwann cells of sensory nerves, often seen in head and neck region. Orbital schwannomas are rare.[1] Few cases of cystic schwannomas have been reported in the brainstem, intracranial nerves, adrenals and orbit[1] but to our knowledge, no cases of cystic orbital schwannoma with intra-ocular extension have been documented till date. There are no pathognomonic clinical or radiological features and definitive diagnosis is based on histopathology.

Case History

A 24 year old woman, presented to us with headache and progressive proptosis in the right eye (RE) since four years which gradually increased causing horizontal dystopia (Figure 1). Neonatal history revealed a full term forceps delivery. On examination, the visual acuity was no perception of light and presence of a shrunken globe with rudimentary cornea was seen. Left eye (LE) showed features of microphthalmos with microcornea. Diminution of vision in the LE occurred after 15 years of age, which deteriorated to perception of light.

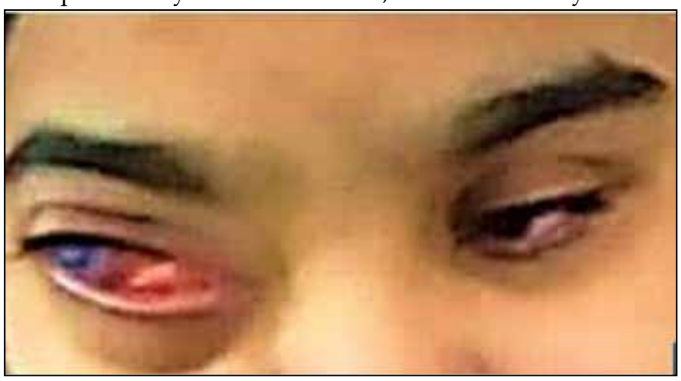

Figure 1: *Clinical picture showing proptosis and horizontal dystopia of the right eye.*

RE Computed Tomography (CT) of orbit with contrast showed a well-demarcated, complex lesion predominantly cystic with a small solid component, measuring 3.2 x 2.8 x 2.4cm in size. This was located retro-orbital and infero-nasal to the globe-like structure causing medial bowing of the lamina papyracea (Figure 2a). The solid component with calcification measured 1.4 x 1.2cm and was seen on the anterolateral aspect of the cystic lesion (Figure 2b). Shrunken globe-like structure, hyperdense and mildly enhancing, measuring 1.3 x 1.3cm with absent crystalline lens and foci of calcification within, was noted (Figure 2c). CT findings of LE showed an eyeball measuring 2.1 x 2.3 x 2.7cm with intact lens. Similar smaller cystic lesions were seen on the sclera, superior to the optic nerve. On T2 weighted Magnetic resonance imaging (MRI) of RE, the cyst was hyper-intense with a hypo-intense solid component (Fig.3b). Post contrast showed greater enhancement of the cystic lesion (Figure 3c). Anteriorly, the cystic component was seen attached to the globe-like structure (Figure 3d). LE showed multiple hyper-intense cysts projecting posteriorly from the globe (Figure 3a). Imaging of the superior and inferior orbital fissures, sinuses and brain were unremarkable. No systemic malformations were present. Probable diagnosis of a right congenital cystic eyeball and left microphthalmos with cyst was made.

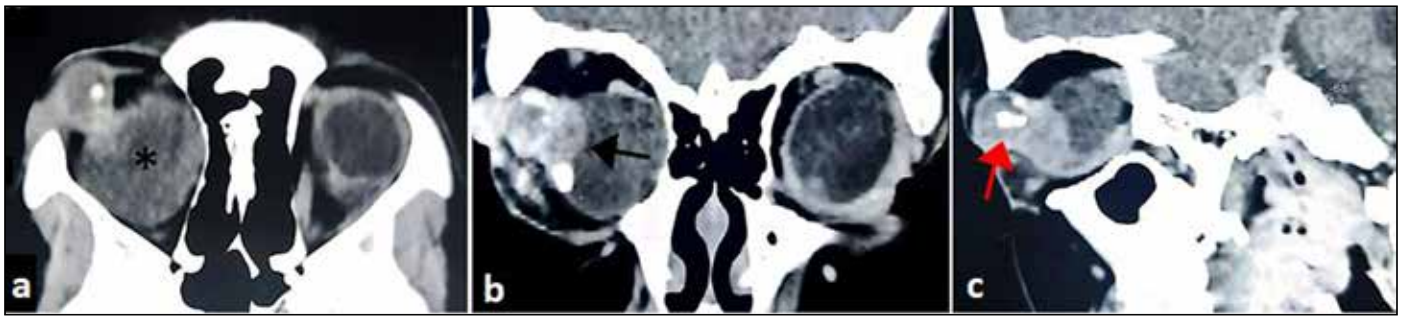

Figure 2: *Orbital Computed tomography (CT) scan of the right eye (a) Axial view showing a well-defined cystic lesion (asterisk) located in the retro-orbital plane. (b) Coronal view showing presence of a solid component (black arrow) within the lesion with calcifications (c) Sagittal view demonstrating enhancement of the calcifications (red arrow) within the shrunken globe.*

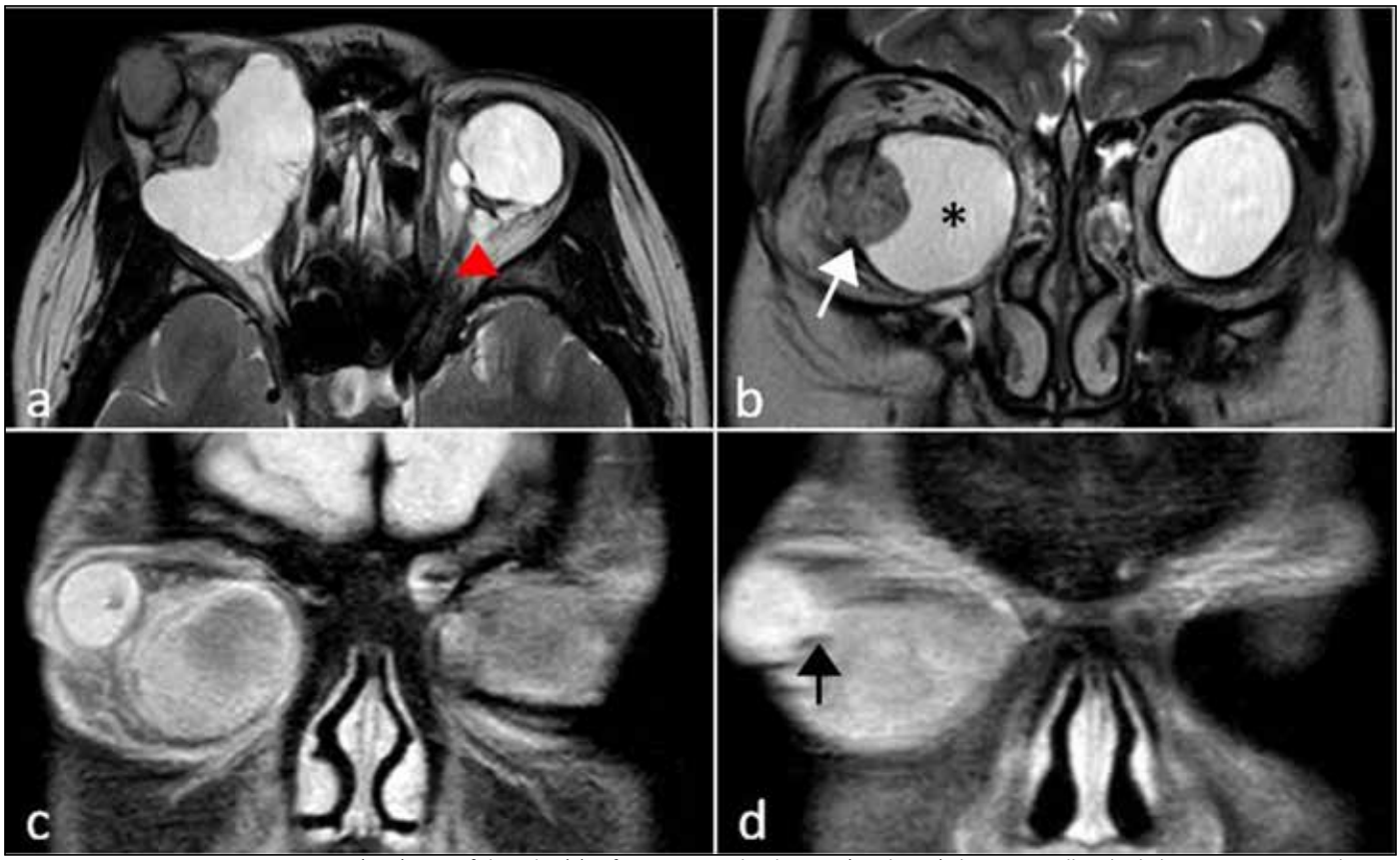

Figure 3: *Magnetic Resonance Imaging (MRI) scan of the orbit (a) Left eye, T2 weighted image (axial view) showing small multiple hyper-intense cystic lesions behind the posterior sclera (red arrow head). (b) Right eye, T2 weighted image (coronal view) showing a hyper-intense cystic lesion (asterisk) with a hypo-intense solid component (white arrow). (c) Right eye, post contrast scan showing enhancement of the lesion as well as the eyeball. (d) Right eye (coronal view) showing anterior attachment of the cystic lesion to the globe (black arrow).*

RE cystic mass was aspirated and sclerosing agent was injected elsewhere. However, it recurred with no further improvement in symptoms. RE enucleation with cyst removal into was performed. Gross specimen showed a globe-like structure measuring 2 x 1.8 x 1.7cm filled with soft greyish-white solid material with no evidence of crystalline lens. Another encapsulated solid-cystic mass of 3 x 2.5 x 2cm was found adherent to the globe-like structure, infero-nasally. Optic nerve was not seen separately from this lesion but was found attached at some points. On light microscopy, sections of gross specimen of both components consisted of closely packed fusiform cells with eosinophilic cytoplasm suggestive of Antoni A pattern. The nuclei were arranged in a typical palisade pattern known as a verocay body (Figure 4a)(Figure 5). In some areas, fewer cells with oval nuclei arranged haphazardly within loose myxomatous tissue descriptive of Antoni B pattern, with hyalinised blood vessels were seen (Figure 4b)(Figure 5). Cystic spaces with extensive degeneration were a characteristic feature noted on histopathological examination (Figure 4c)(Figure 5). No signs of mitosis, necrosis and nuclear atypia were noted. With the confirmatory histopathology findings, a diagnosis of Orbital Schwannoma with cystic degeneration and intra-ocular extension was made.

Discussion

Orbital schwannomas are seen commonly in females occurring in second to fifth decade.[2] In the orbit, specific origin of the tumor cannot be identified owing to complexity

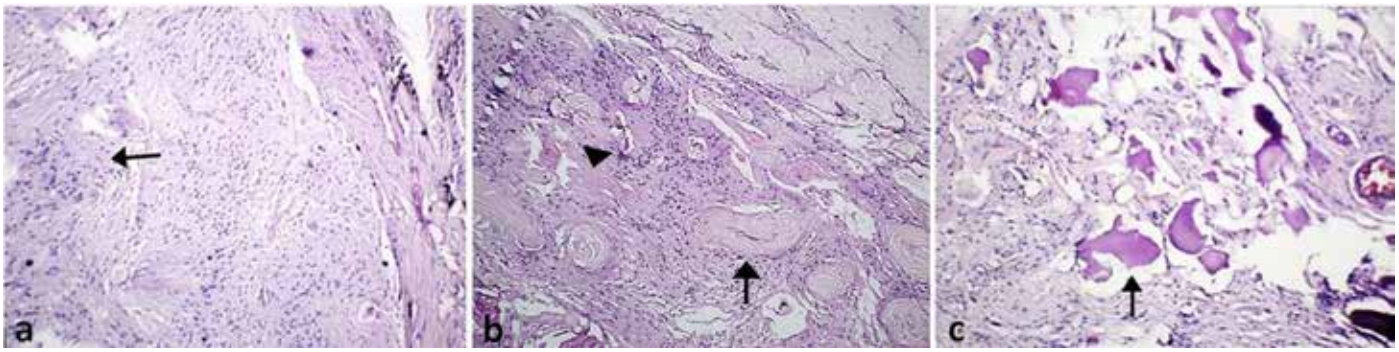

Figure 4: *(Hematoxylin-eosin stain, 10x). Photomicrograph of intra-ocular mass showing (a) Antoni A pattern with typical verocay bodies (black arrow) (b) Antoni B pattern (arrow heads) with hyalinised blood vessels (black arrow) and (c) Areas of extensive cystic degeneration (black arrow).*

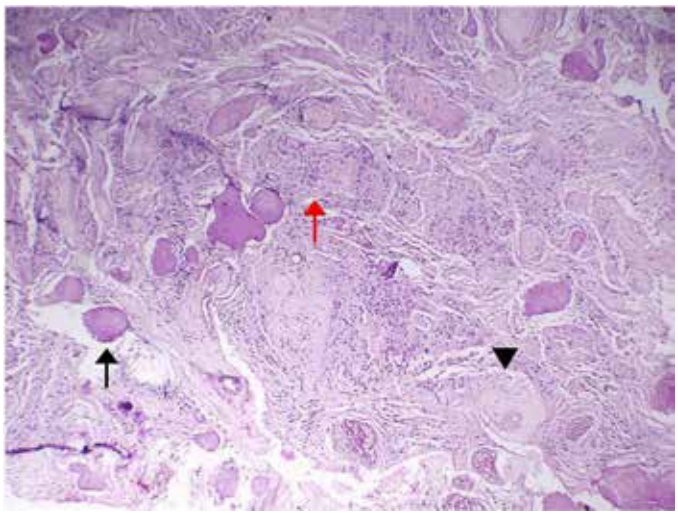

Figure 5: *(Hematoxylin-eosin stain, 10x).Photomicrograph of orbital mass showing typical features of a cystic Schwannoma. Cystic degeneration (black arrow); Antoni A pattern with verocay bodies (red arrow); Antoni B areas with hyalinised blood vessels (arrow heads)*

of nerves traversing it. However, tumor often arises from the supratrochlear or supraorbital branches of trigeminal nerve.[2] They present with proptosis, decreased vision, diplopia, pain or numbness along the nerve of origin.[2] Schwannomas are usually unilateral.[2] Although, in our patient, MRI showed retro-orbital cystic lesions in the LE, nature of which cannot be determined without biopsy.

Long standing tumours can undergo cystic degeneration which has been reported in a few cases of orbital schwannomas,[3] but none have demonstrated its intra-ocular presence. Cystic areas are secondary to coalescence of microcysts within the Antoni B tissue. It is hypothesized that alteration in the vascular supply resulting in necrosis and haemorrhage within the tumor or hyaline degeneration causes cyst formation.[1] Diagnostic difficulty in our case stemmed from the cystic nature of the lesion seen bilaterally on MRI, long standing duration and past history of cyst aspiration which misled us to a diagnosis of a right sided congenital cystic eye, which is known to occur either in isolation or with malformations such as contralateral microphthalmos with cyst.[4]

MRI is the imaging of choice in suspected cases of orbital schwannoma, due to its high sensitivity especially with the use of contrast agents.[5] On MRI, they appear hyper-intense similar to cerebrospinal fluid on T2 weighted images and hypo-intense on T1 weighted images with homogenous or heterogeneous enhancement on contrast. This heterogeneity is seen in cases of cystic degeneration. Although, imaging is helpful in diagnosing orbital schwannomas, there are no pathognomonic diagnostic features and histopathological confirmation is needed. Gross appearance of schwannoma is characteristic with a smooth homogenous yellow-grey well-encapsulated mass, the capsule derived from perineurium of the nerve of origin. Microscopically, biphasic cellular arrangements are seen: a densely cellular Antoni A pattern with picket-fence-like palisaded nuclei (verocay bodies) and less cellular Antoni B pattern distributed in myxoid tissue. Rarely, calcifications can be seen within the schwannoma.

Our patient showed characteristic features of a cystic schwannoma with intraocular extension probably via optic nerve, explaining locally aggressive nature of the tumor.[6] Similar cystic lesions were seen in LE but since the patient was asymptomatic, we decided against any intervention. Presence of these retro-orbital cystic lesions speculates the possibility of a bilateral schwannoma, which is not reported in literature till date and can be confirmed only on histopathology. These varied presentations of schwannoma make it challenging for the physician to come to a conclusive diagnosis. Thus, schwannoma must be borne in mind as a differential for an orbital cystic lesion.

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Address for correspondence

Shruthi Tara, DO
Deparment of Orbit and Oculoplasty,
Sankara Eye Hospital,Coimbatore,
Tamil nadu, India.
E-mail: shruthitarav@gmail.com

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