CASE REPORT

Orbital cystic schwannoma masquerading as hydatid cyst

Shabeer Basheer, Amar Pujari, Mandeep S Bajaj, Hameed Obedulla

Ophthalmology, Dr. Rajendra Prasad Centre for Ophthalmic Sciences, AIIMS, New Delhi, India

Correspondence to
Dr Amar Pujari,
dramarpujari@gmail.com

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SUMMARY

Gradually increasing abaxial proptosis in a middle-aged female patient revealed a large well-defined cystic lesion behind the globe extending till the orbital apex. B scan ultrasound and CT findings were favouring a diagnosis of orbital hydatid cyst. Thus as a diagnostic/curative surgical protocol, the cystic lesion was removed in total. However, intraoperative gross specimen examination was not classical of hydatid cyst, thus a further histopathological evaluation was carried out, which confirmed the diagnosis of orbital schwannoma with extensive cystic degeneration. Postoperatively the clinical course was uneventful at the end of 1 year.

BACKGROUND

Orbital schwannomas are the great masquerades among the orbital tumours.¹ These are benign, slow-growing, painless tumours arising from the Schwann cells of the peripheral nerve endings. Schwannomas with extensive cystic degeneration have been infrequently noted in the literature.¹⁻⁶ Clinical evaluation and radiological findings may not be accurate to label the final diagnosis. A strong suspicion of this tumour beforehand with appropriate surgical intervention and final analysis of the histopathology is of paramount importance. Here, we discuss the clinical features, evaluation and management of a case of orbital cystic schwannoma which was masquerading as hydatid cyst.

CASE PRESENTATION**Case description**

A 35-year-old female patient presented with painless progressive proptosis in the right eye for the past two years. On examination visual acuity was no perception of light; the proptosis was non tender, without any raise in local temperature; however, the extraocular motilities were severely restricted in all gazes. The proptosis was not associated with any pulsation; bruit or any change in the size following Valsalva manoeuvre. Measurement revealed an axial proptosis of >12 mm, with an inferior globe dystopia of 8 mm and temporal dystopia of 10 mm (figure 1A). Cornea was exposed with a leucomatous scar, and the surrounding sclera showed severe congestion and mucopurulent discharge. To look for the nature of mass lesion, a B scan ultrasonography of the orbit was performed which showed a large solitary cystic lesion in the absence of any appreciable contents. Two separate spikes were made out corresponding with the ocular

coat and the capsule of the cyst; these findings in endemic countries hinted the possible diagnosis of hydatid cyst which is classically seen in these cases, thus favouring a diagnosis of orbital hydatid cyst (figure 1B).

A contrast-enhanced CT of the orbit confirmed a large, well-defined, solitary cystic lesion along the superior orbital compartment extending from the posterior aspect of the globe till orbital apex with significant globe indentation (figure 1C). The cystic mass did not reveal any significant heterogeneity but based on the clinical and radiological findings a probable diagnosis of orbital hydatid cyst was made.

Under general anaesthesia, the patient was planned for superior transconjunctival orbitotomy. Intraoperatively after dissecting all the adhesions (a thinned inferiorly displaced superior rectus muscle was noted; however, the lacrimal gland appeared normal), a cryo-assisted extraction was performed to remove the lesion in total. Gross specimen appeared yellowish in colour with a large cystic cavity having gelatinous material within it (figure 2A). Histopathology showed a spindle cell lesion consisting of Antoni A and Antoni B areas with numerous cystic spaces devoid of lining epithelium suggestive of cystic schwannoma (figure 2B). Postoperatively, there was complete resolution of proptosis and the patient remained symptom free for a year. For the corneal opacity, rehabilitation was advised using cosmetic contact lens.

TREATMENT

Under general anaesthesia, the patient underwent superior transconjunctival orbitotomy.

OUTCOME AND FOLLOW-UP

At the end of 1 year, the clinical course was uneventful.

DISCUSSION

Cystic schwannoma may involve the intraconal as well as an extraconal compartment of the orbit to masquerade as orbital dermoid, frontal mucocele, venolymphatic malformation and others.¹⁻⁶ This clinical variant appears to involve females slightly more than male patients among the reported cases and case series.¹⁻⁶ Orbital schwannomas usually arise from the first division of the trigeminal nerve along the superior and medial compartment of the orbit. Clinically these tumours may cause significant proptosis, exposure keratopathy, globe indentation and optic nerve compression leading to visual deterioration. Complete surgical excision in these cases is possible

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Figure 1 (A) Clinical profile revealing an abaxial proptosis with conjunctival chemosis and corneal scar. (B) B scan orbital ultrasound revealing a well-defined cystic with an indentation over the globe giving the appearance of two layers to confound as double-wall sign (red arrows) of hydatid cyst. (C) Sagittal views of the CT showing a well-defined oblong cystic mass extending till the orbital apex without appreciable internal contents.

Figure 2 (A) Gross specimen appeared yellowish in colour with a large central cystic area. (B) Histopathology revealed Antoni A (more cellular area) and Antoni B (less cellular area) areas along with numerous cystic spaces suggestive of extensive cystic degeneration of the schwannoma.

in almost all cases because of their well-defined nature. Following complete surgical excision, grossly these tumours may appear greyish in colour and on further dissection they may reveal solid

and cystic areas depending on the extent of degeneration. Microscopically two cellular areas can be identified: Antoni A and Antoni B, Antoni A areas are solid and Antoni B areas are due to aggregation of mucinous and microcystic areas.

In the literature, Kashyap *et al* have studied a total of 39 cases of orbital schwannoma, of which 19 (41%) showed cystic degeneration. In their case series, the age varied from 20 to 65 years and the cystic degeneration was seen in 11 female patients. Duration of symptoms also varied from a few months to years. The preoperative diagnostic dilemma was evident in their series too; however, all cases were completely removed surgically to achieve good visual and cosmetic results.¹ Similarly, Subramanian *et al* described four cases of cystic orbital schwannoma involving the intraconal compartment.² Rest are isolated case reports describing the varied manifestation of the tumour and final conclusion of the definite diagnosis after the histopathological confirmation.^{3–6}

Orbital hydatid cyst is endemic in the Middle East, India, Africa and other southern American and European countries.⁷ In general, orbital hydatid cyst accounts for <1% of all hydatid cysts but it may contribute up to 20% of all the orbital cystic masses in endemic countries.⁸ Thus orbital proptosis due to cystic mass in endemic countries needs a tailored approach to prevent inadvertent complications.

To conclude, the present case under discussion was diagnosed as orbital hydatid cyst preoperatively based on the clinical profile in the endemic countries and imaging findings. A standard surgical protocol was followed to achieve a complete excision of the cystic mass. Postoperatively careful histopathological examination by an experienced ocular pathologist revealed it to be a case of schwannoma with cystic degeneration. Thus cases of cystic orbital lesions in endemic countries need special attention throughout the clinical course for a definite diagnosis.

Learning points

- ▶ All cases of proptosis secondary to definite orbital mass need careful preoperative, intraoperative and postoperative evaluation.
- ▶ In endemic countries, long-standing orbital mass may have an infectious aetiology like hydatid cyst and cysticercosis or a benign orbital pathology.
- ▶ Orbital schwannomas, if not intervened in their early clinical course, may undergo extensive degenerative changes to masquerade as cystic lesions.

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