CASE REPORT

Concomitant orbital cavernous haemangioma and schwannoma in a patient

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SUMMARY

A woman aged 39 years presented with right eye painless proptosis and decreased vision since 5 months. Right fundus examination revealed optic disc oedema. CT of the orbit revealed a well-defined homogeneous mass in the intraconal space in both orbits. Surgical excisional biopsy of the orbital lesions was performed. Histopathological examination of the right orbital lesion was suggestive of cavernous haemangioma and the left orbital lesion revealed schwannoma. The patient had an unremarkable postoperative course with improved visual acuity. This case illustrates benign orbital masses of 2 different tissues of origin in a single patient, which has not been described in the literature.

BACKGROUND

Primary space-occupying lesions of the orbit are mostly unilateral. In a study of 2480 consecutive patients with orbital space-occupying lesions, vascular lesions were most common (24%) with cavernous haemangioma being the most common vascular tumour (36%) with the mean age at presentation of 48 years.¹ Neural tumours constituted 12%, of which schwannomas were the third most common tumours (11%) with the mean age at presentation of 41 years.¹ In a histopathological study of 3476 orbital diseases, cavernous haemangioma was the most common lesion accounting for 15% of lesions and schwannomas accounted for 5% lesions.² In a study of 1264 consecutive American patients with orbital tumours and simulating lesions, the most common orbital tumour was of vascular origin accounting for 17% of lesions.³ Cavernous haemangioma was the most common vascular lesion (36%) with a mean age at presentation of 48 years.³ Peripheral nerve lesions accounted for 2% of the lesions, with schwannoma being the most common peripheral nerve lesion (61%) presenting at a mean age of 37 years.³ In a study of 1277 Asian Indian patients with primary space-occupying lesions of the orbit, the tumours were vascular in 29% cases and neural in 26% cases.⁴ Cavernous haemangioma constituted 25% of the vascular lesions, being the most common vascular lesion, and schwannoma constituted 13% of the neural tumours, being the third most common neural tumour.⁴

Bilateral primary benign tumours of similar tissue origin, cavernous haemangiomas or orbital schwannomas have been reported earlier.^{5–12} However, concomitant occurrence of two orbital tumours with different tissue origin, a vascular tumour and peripheral neural tumour, has not been

reported earlier. In this report, we discuss biopsy-proven concomitant presentation of cavernous haemangioma in the right orbit and schwannoma in the left orbit in a female patient aged 39 years.

CASE PRESENTATION

A woman aged 39 years presented with 5 months history of painless progressive proptosis and decreased vision in the right eye (figure 1A). There was no significant personal or familial medical history. On ocular examination, the best-corrected visual acuity was 20/800 in the right eye and 20/20 in the left eye. Decreased colour vision was noted in the right eye. Hertel's exophthalmometry revealed proptosis of 7 mm in the right eye. Anterior segment examination of both eyes was unremarkable. Fundus examination revealed optic disc oedema in the right eye. Systemic examination of the patient was unremarkable.

INVESTIGATIONS

CT of the orbit revealed a well-defined oval homogeneous soft tissue mass inferotemporally in the intraconal space in both the orbits (figure 1B). The mass in the right orbit was larger than one in the left orbital lesion and caused globe indentation posteriorly. The lesions in both the orbits were separate from the optic nerve, and the optic nerve was pushed medially in the right orbit.

DIFFERENTIAL DIAGNOSIS

A clinical diagnosis of bilateral cavernous haemangioma was made. A differential diagnosis of bilateral orbital schwannoma was also considered.

TREATMENT

Both the orbital masses were removed in toto by inferior transconjunctival orbitotomy approach (figure 1C, D). Removal of right orbital tumour was followed by left orbitotomy 1 week later.

OUTCOME AND FOLLOW-UP

On gross examination, the right orbital lesions showed a well-circumscribed oval purplish-red soft tissue mass measuring 25×25×12 mm. Histopathology showed interconnecting vascular channels lined by flattened endothelial cells consistent with cavernous haemangioma (figure 1E). There was no schwannoma area around the cavernous haemangioma. Gross examination of the left orbital lesion revealed an encapsulated greyish-white mass measuring 13×13×9 mm, with cut section showing grey white fibrous areas. Histopathological examination showed proliferation

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Figure 1 (A) Right eye proptosis with increased inferior scleral show. (B) CT of the orbit revealed well-defined homogeneous mass in the intraconal space in both orbits. (C) Well-defined purplish-red mass was removed from the right orbit. (D) Well-defined encapsulated yellowish-white mass was excised from the left orbit. (E) Histopathological examination (H&E stain, 40× magnification) of the right orbital lesion revealed blood-filled spaces separated by fibrous septae suggestive of cavernous haemangioma. (F) Histopathological examination (H&E stain, 40× magnification) of the left orbital lesion revealed Antoni A and pattern arrangement of cells suggestive of schwannoma. (G) The left orbital lesion was S100-positive confirming the neurogenic nature of the tumour. (H) Postoperatively, there was no proptosis in either eye.

of spindle cells in Antoni A and B areas with fascicles and verocay bodies suggestive of orbital schwannoma (figure 1F). S100 stain was positive confirming the neurogenic nature of the tumour (figure 1G). There was no cavernous haemangioma within the schwannoma. The patient had an unremarkable post-operative course (figure 1H) with improved visual acuity. The vision remained 20/20 in the left eye and the vision in the right eye improved to 20/20 and the optic disc oedema resolved in 6 weeks.

DISCUSSION

The majority of primary benign orbital tumours are unilateral. Simultaneous bilateral or multiple orbital tumours are more common with malignant tumours, especially tumours of

lymphoproliferative origin or those metastasising from a distant primary. Thus, most bilateral or multiple orbital tumours have a similar cell of origin.

Cavernous haemangioma of the orbit is the most common benign tumour accounting for 6% of all orbital tumours.³ They almost always occur unilaterally. Bilateral cavernous haemangiomas of the orbit are very rare, with only isolated case reports and small case series in the literature.^{5–9} Paonessa *et al*⁹ studied the role of MRI of the orbit in the diagnosis of cavernous haemangioma and noted that bilateral cavernous haemangiomas are more frequent than predicted in the literature. In their series of 14 patients, 3 (21%) cases had bilateral cavernous haemangioma, which were detected on careful interpretation of MRI of orbit.⁹ Bilateral cavernous haemangiomas can occur as a part of other systemic haemangiomatosis in Maffucci's syndrome and blue rubber bleb nevus syndrome.^{10 11} Rarely, unilateral multiple cavernous haemangioma of the orbit,¹² coexistence of cavernous haemangioma with other vascular malformations, including orbital varices, lymphangioma and low-flow arteriovenous malformation,¹³ and association of orbital multifocal cavernous haemangioma with bilateral arachnoid cysts of the middle cranial fossa¹⁴ have been reported. In our case, owing to well-defined lesions in both orbits and based on the literature on coexistence of bilateral cavernous haemangioma with no significant personal history, a preoperative clinical diagnosis of bilateral cavernous haemangioma was made. However, on histopathology, only the right orbital lesion had cavernous haemangioma.

Orbital schwannomas or neurilemmomas are benign, slow-growing tumours arising from Schwann cells.³ They usually have eccentric growth from sensory nerves. These tumours account for 1% of orbital tumours.³ Benign schwannomas are usually sporadic and solitary with one case of bilateral simultaneous involvement described in the literature.¹⁵ Heritable diseases are also associated with the development of multiple schwannomas, including neurofibromatosis type 2, schwannomatosis and Carney complex.¹⁶ In our case, a clinical diagnosis of bilateral orbital schwannoma was also considered. However, on histopathology, only the left orbital lesion was a schwannoma.

Conjoined association in which a cavernous haemangioma is located within a central or peripheral nerve tumour (schwannoma/ganglioglioma/oligodendroglioma) and discrete association in which cavernous haemangioma and a central or peripheral nerve tumour (oligodendroglioma/neurofibromatosis/meningioma/astrocytoma/ependymoma) are in separate locations are a rare occurrence.¹⁷ It is hypothesised that the association between cavernous haemangioma and tumours of neural origin may be related to a common genetic pathway in the majority of cases.¹⁷ In our case, cavernous haemangioma and schwannoma were discrete lesions, one in each orbit. None of the lesions displayed conjoined association.

Concomitant occurrence of cavernous haemangioma and schwannoma in the spine has been reported.¹⁸ Juxtaposition of extradural cavernous haemangioma and intradural schwannoma in the thoracic spine has been reported in a woman aged 47 years.¹⁸ Simultaneous occurrence of two tumours of different aetiologies could have been a coincidental finding. It was also speculated that the cavernous tumour in the extradural space could be related to altered venous drainage as a result of intradural tumour or may be related to mutation in a common precursor cell in the neural crest.¹⁸ In our case, the two tumours were not in juxtaposition and were rather located in either orbit.

To the best of our knowledge, this is the only reported case of benign orbital masses of two different tissues of origin, with cavernous haemangioma in one orbit and schwannoma in the other orbit in a single patient. This finding of two different aetiologies in our patient is most likely a coincidental finding with no evidence of any systemic association or intracranial lesion.

Learning points

- Bilateral orbital lesions are a rare presentation.
- Although bilateral orbital diseases commonly have same etiopathology, a possibility of varied pathogenesis should be considered.
- In the case of bilateral orbital masses, it is mandatory to rule out any systemic involvement or syndromic association.

Contributors SG is responsible for planning, conduct, reporting and data acquisition. SK is responsible for planning, conduct, reporting, conception and design, of the report. SwG is responsible for reporting and interpretation of data.

Competing interests None declared.

Patient consent Obtained.

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