Bilateral orbital cavernous haemangiomas

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SUMMARY Simultaneous bilateral orbital lesions are rare. The differential diagnosis includes orbital pseudotumour, metastasis, leukaemia, lymphoma, Wegener's granulomatosis, and neurofibromatosis. We report what we believe to be the first case of bilateral orbital cavernous haemangiomas.

Cavernous haemangioma is the most common primary adult benign orbital tumour.^{1,2} It usually presents in middle aged adult females with slow, painless onset of unilateral proptosis. Multiple unilateral cavernous haemangiomas have been reported.² Cavernous haemangiomas have been diagnosed several years apart, as growing during pregnancy, or in association with haemangiomas elsewhere.^{3,6} To our knowledge this is the first reported case of bilateral orbital cavernous haemangiomas.

Case report

A 48-year-old white male developed bilateral swelling of the eyes. Four to five years previously he noted a slowly increasing prominence of both eyes, the right somewhat greater than the left. Two years previously he observed that his eyelashes touched the inner surface of his spectacle lenses and four months prior to referral began noting bilateral epiphora. No redness, pain, diplopia, decreased vision, or headache was reported. The past medical history and results of systemic examination were negative.

The best corrected visual acuity was 20/25 OD and 20/30 OS. The pupils were 3 mm in size, reactive to light, and were without an afferent defect. Examination of ocular motility revealed mild limitations in all fields of gaze without diplopia. Colour vision was normal. Hertel exophthalmometry at a base of 96 measured 23 mm OD and 20 mm OS. External examination revealed 2 mm of inferior scleral show on the right and 1.5 mm on the left. Prominent insertions of the horizontal rectus muscles were

noted in both eyes. No orbital mass was palpable. A subtle lid lag was present. Slit-lamp biomicroscopy, applanation tonometry, automated visual fields, and fundus examination were normal.

1.5 mm axial computed tomography views of both orbits, with coronal reconstructions, were obtained with and without contrast. Bilateral, moderately enhancing, homogeneous, smooth-contoured masses were seen. The right lesion was ovoid, displaced the globe anteriorly, displaced the optic nerve nasally, and showed mild bony orbital wall contour changes. The mass was approximately 30×15×12 mm. Because of the unusual feature of bilateral, simultaneous orbital masses the patient was referred to the UCSF Ocular Oncology Unit for examination and therapy.

Initial examination of the patient gave findings similar to those already presented. A magnetic resonance imaging (MRI) scan was performed on a 1.5 Tesla superconducting magnet (General Electric) with orbital surface coils (General Electric). Sections of 3 mm at 4.5 mm intervals in transverse, sagittal, and coronal planes were obtained. Spin-echo technique with routine T1-weighted examination was performed with a repetition time (TR) of 600 ms and an echo time (TE) of 20 ms. The T2-weighted examination had a TR of 2000 ms and a TE of 70 ms. This scan revealed similar information as was gathered on CT. A bilobed, lateral, intraconal, homogeneous mass was localised in the right orbit. The single tumour in the left orbit was both intraconal and extraconal at the inferior temporal aspect of the posterior orbit. The lesions were isointense with brain and sharply contrasted against orbital fat on both T₁ and T₂ weighted images (Figs. 1A, B).

Our clinical and radiological impression was of cavernous haemangioma of the orbit, but owing to

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Fig. 1A Left: T_1 -weighted axial MR image (TR 600:TE 25) of orbital cavernous haemangiomas.

Fig. 1B Right: T_2 -weighted coronal MR image (TR 2000:TE 70) outlining bilateral orbital cavernous haemangiomas.

the bilaterality doubt remained. At right orbitotomy a large, bilobed, smooth, encapsulated tumour was removed intact without complication (Fig. 2). Histologically this proved to be a cavernous haemangioma. Ocular movements and proptosis returned to normal postoperatively. Owing to a paucity of symptoms OS, no operation has been performed.

Discussion

Bilateral proptosis is most commonly caused by endocrine ophthalmopathy. Other less common causes include pseudotumour, Wegener's granulomatosis, other vasculitides, metastases, leukaemia, lymphoma, cavernous sinus thrombosis, and neuro-

Fig. 2 Histology of specimen removed from the right orbit. Large blood filled spaces separated by fibrous septa are typical of orbital cavernous haemangioma. H and E $\times 6$.

fibromatosis.^{1 7-10} Computed tomography and magnetic resonance imaging will usually distinguish diffuse from solitary orbital processes. The typical CT findings of cavernous haemangioma of the orbit are its homogeneous, ovoid, smooth-margined appearance. Usually it is intraconal, shows minimal orbital bone expansion, enhances with contrast, and spares the orbital apex.¹¹ These features allow differentiation from diffuse orbital processes. However, more localised entities such as neurofibroma, neurolemmoma, benign mesenchymal tumour, or haemangiopericytoma may show a well circumscribed, oval mass.^{11 12} In our case the diagnosis was suspected but not confirmed until surgical resection, because no previous cases of bilateral cavernous haemangioma of the orbit have been reported.

The pathogenesis of orbital cavernous haemangioma is not clear. Probably they arise as acquired lesions due to localised, progressively ectatic, capillary-type proliferation resulting in large vascular spaces.² Witschel and Font, in their study of haemangioma of the choroid, proposed that localised haemodynamic disturbances may be a nidus to initiate the development of these lesions.¹³ Hood maintained that growth is not due to proliferation of vascular elements but rather to canalisation and blood flow into pre-existing vascular malformations secondary to changing systemic haemodynamics.¹⁴ None of these theories suggest a rationale other than random chance to explain the bilaterality of the cavernous haemangiomas observed in our case.

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