

# Multiple Cavernous Hemangiomas of the Orbit

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## Abstract

**Purpose:** Cavernous hemangioma of the orbit is often unilateral and solitary. Multiple cavernous hemangiomas of the orbit are extremely rare. The authors reported four patients who had more than two cavernous hemangiomas in one orbit.

**Methods:** Case reports. The clinical features, imaging findings, differential diagnosis and surgical management were presented.

**Results:** We found 4 cases of multiple cavernous hemangiomas of the orbit out of 214 records of all patients with orbital cavernous hemangiomas who were referred to us from Jan 1, 1986 to Dec 31, 2000. Two patients were female and two were male. The mean age was 46.5 years, ranging from 37.0 to 57.0 years. The left orbits were affected in all patients, with two, three, eight and ten tumors, respectively. The most common complaint was a painless, gradually progressive proptosis. The characteristic CT imaging features were multiple well circumscribed, round or ovoid masses that were sharply delineated from the surrounding tissue. B-scan ultrasonography showed round-shaped orbital masses with high internal reflectivity. The therapeutic outcomes of lateral or anterior orbitotomy were excellent.

**Conclusion:** Although rare, cavernous hemangioma should be considered in the differential diagnosis of patients with multiple orbital mass lesions.

**Keywords:** Cavernous hemangioma; orbit; multiple; orbitotomy; diagnosis

## Introduction

Orbital cavernous hemangioma (OCH) is the most common benign primary intraconal tumor of the orbit in adults<sup>1-4</sup>. It is often unilateral and solitary. The

most common clinical sign of OCH is gradually progressive proptosis. Patients usually have decreased visual acuity when the mass compresses the optic nerve or the eyeball. Most OCHs should be treated by surgical excision. Multiple cavernous hemangiomas of the orbit are very rarely seen in clinical practice, which were reported in only a few case series<sup>1-7</sup>. We reviewed 214 records of all OCH patients referred to us from Jan 1, 1986 to Dec 31, 2000 and found 4 cases of multiple cavernous hemangiomas of the orbit which had been confirmed by surgeries and pathological examinations<sup>3</sup>. All of the 4 patients were Chinese. Here we summarized the clinical features and therapeutic outcomes of these cases.

## Case reports

**Case 1:** A 52-year-old woman was referred to us on Oct 18, 1999 because of gradually progressive proptosis together with upward displacement of the eye and slightly blurred vision in his left eye for ten years. General physical examination revealed no abnormalities. Visual acuity was 20/25 in the right eye and 20/20 in the left. Intraocular pressure was 14.6mmHg and 20.55mmHg in the right and left eyes, respectively. The lens cortex in both eyes was mildly opacified. Fundus examination showed radial choroidal striae at the posterior pole of the left eye. Hertel exophthalmometry readings were 15 mm in the right eye and 25 mm in the left eye at a base of 105 mm. On palpation there was a firm, nontender, well-defined mass over the medial aspect of the left upper eyelid with a size of 1.0×0.8 cm. The patient also had an exotropia of 15° and a hypertropia of 10° in the left eye (Figure 1). Extraocular movement examination showed slight abduction deficit in the left eye.

Color doppler ultrasonography displayed four well-defined homogenous mass measured from 0.5×

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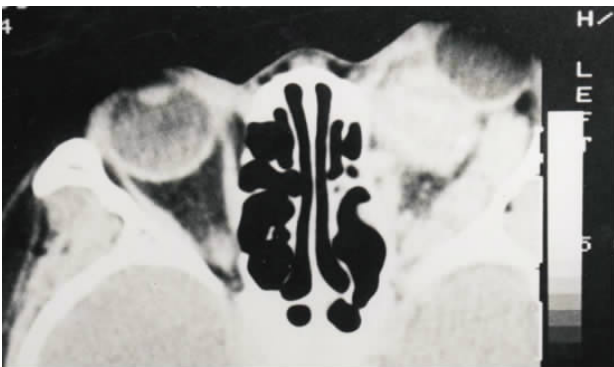
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**Fig.1** Proptosis and upward displacement of the left eye (Case 1).

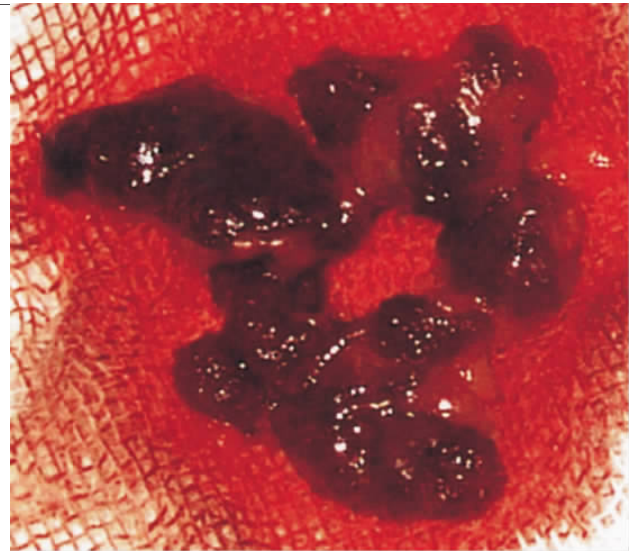
0.5 cm to 1.0×0.8 cm with no blood flow in the left orbit. Computed tomography (CT) scan of the left orbit demonstrated more than six round-shaped soft tissue masses, varying in diameter from 5 mm to 10 mm. Some of the lesions were intraconal, three masses lay along the lateral bony orbit. The left eyeball was pushed forward and laterally (Figure 2).



**Fig.2** Axial CT showing six soft tissue masses of various size in the left orbit (Case 1).

On Oct 20, 1999, a transconjunctival anterior orbitotomy of the left eye was performed to gain access to the tumors. Ten accessible, distinct, redish purple, round-shaped masses were excised from the left orbit with the tumors measuring from 0.5×0.5×0.6 cm to 1.0×1.0×1.0 cm (Figure 3). All tumors were readily removed. The histopathologic studies of these lesions revealed large, blood-filled spaces lined with endothelium and separated by fibrous septa of varying thickness that is consistent with the characteristics of cavernous hemangiomas of the orbit. Six months postoperatively, the patient's best corrected visual acuity of the left eye was 20/20, with a 9 mm reduction in proptosis. He had no abnormalities in extraocular movements after the operation.

Case 2: A 40-year-old woman experienced pro-



**Fig.3** Intraoperative tumor specimens (Case 1).

gressive left proptosis with decreased visual acuity for 1 year and consulted us on Aug 29, 2000. The past medical history was insignificant and the systemic examination revealed no positive signs. The best corrected visual acuity was 25/20 in the right and 20/20 in the left. Slit-lamp biomicroscopy, applanation tonometry, and fundus examination were normal in both eyes. Hertel exophthalmometry reading was 16 mm in the right eye and 17 mm in the left, with a base of 96 mm. No orbital mass was palpable. Ocular motility was normal in both eyes. CT revealed six well-demarcated soft tissue masses in the left orbit with remarkable enhancement. Incisional biopsy of anterior orbitotomy under local anesthesia was performed. During the surgery, 8 reddish purple soft masses were removed intact without complication. These masses were proved to be cavernous hemangiomas by histopathological examination. After the surgery, the best corrected visual acuity was 25/20 in the right eye and 25/20 in the left. Hertel exophthalmometry reading was 16 mm in both eyes, with a base of 96 mm. The patient had no abnormalities in extraocular movements after operation.

Case 3: A 57-year-old man presented with a progressive proptosis in the left eye for 1 year. The past medical history was insignificant. General physical examination showed normal results. The best corrected visual acuity was 20/70 in the right eye and 20/50 in the left. Intraocular pressure was 12.6 mm Hg in the right and 20.6 mm Hg in the left. The lens cortex was moderately opacified. Fundus was normal

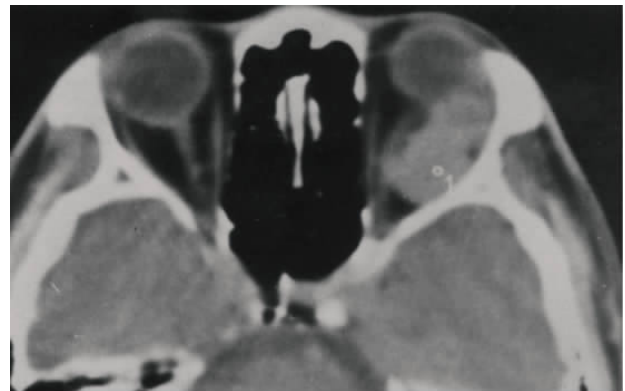
in both eyes. Hertel exophthalmometry reading was 9 mm in the right eye and 16 mm in the left, with a base of 110 mm. Extraocular movement was full in both eyes. No mass was palpable at the orbital rim. B-scan ultrasonography only showed one round-shaped, well-defined mass with moderate internal reflectivity. CT scan demonstrated two soft tissue masses in the left orbit. There were no osseous erosions and no infiltration into the eye or the extraocular muscles (Figure 4). A lateral orbitotomy was performed under general anesthesia on Jan 22, 1997. During the operation, we found two redish purple masses that adhered to the surrounding tissues and were located deep in the left orbit, with a size of 1.5×1.5×2.0 cm and 1.5×0.5×0.3 cm, respectively. We excised one mass completely and the other one incompletely. Histopathologically, the lesions were consistent with cavernous hemangiomas. After the surgery, the best corrected visual acuity was 20/70 in the right eye and 20/50 in the left. Hertel exophthalmometry reading was 9 mm in the right and 10mm in the left, with a base of 110 mm. He had no abnormalities in extraocular movements after the operation. The tumor did not recur one year after operation.



**Fig.4** Axial CT showing two soft tissue masses in the left orbit (Case 3).

Case 4: A 37-year-old man was referred to us on Mar 26, 1995 for the evaluation of a slowly increasing prominence in the left eye for 2 years. No redness, pain, diplopia, vision disturbance and headache were reported. The past medical history was insignificant and the general physical examination revealed no positive signs. Visual acuity was 25/20 in both eyes. Color vision was normal. Slit-lamp biomicroscopy

and fundus examination were negative. Hertel exophthalmometry reading at a base of 107 mm was 13 mm in the right eye and 16 mm in the left. No palpable mass was found at the orbital margin. The ocular motility was normal. B-scan ultrasonography displayed three round-shaped, well-defined and homogenous masses with high internal reflectivity in the left orbit. Computed tomography (CT) scan revealed three round-shaped soft tissue masses with no bone erosion (Figure 5). A lateral orbitotomy was performed under general anesthesia on Mar 29, 1995. Three round-shaped, smooth, redish purple, encapsulated masses with the size of 1.0×1.0×1.5 cm, 0.8×0.7×0.8 cm, 0.4×0.3×0.3 cm, respectively were removed intact without complication. The histopathological diagnosis was cavernous hemangiomas. Hertel exophthalmometry reading decreased to 13 mm in the left eye after the operation. According to the case record, her postoperative course was uneventful during the six months of postoperative follow-up.



**Fig.5** Axial CT showing three soft tissue masses in the left orbit (Case 4).

## Discussion

Orbital cavernous hemangioma is the most common orbital tumor in adults. It is usually unilateral and solitary. Multiple cavernous hemangiomas of the orbit are rarely seen. Only a few case series about this condition were reported. McNab et al reviewed 85 cases of orbital cavernous hemangioma and only 5 cases had multiple lesions<sup>4</sup>. Ruchmann et al found multiple masses in one of 13 patients<sup>5</sup>. Harris et al described 66 cases of orbital cavernous hemangioma and 2 were multiple<sup>2</sup>. Wolin et al reported one case

with two masses in the left orbit<sup>7</sup>. Sullivan et al<sup>6</sup> and Chang et al<sup>1</sup> described one case of bilateral multiple cavernous hemangioma in detail, respectively, and Chang's case belonged to the blue rubber bleb nevus syndrome.

Although rare, cavernous hemangioma should be considered in the differential diagnosis of cases with multiple orbital lesions. Apart from hemangioma, multiple orbital masses may possibly be pseudotumor, lymphoma, leukaemia, neurorilemoma, neurofibroma, Wegener's granulomatosis, other vasculitides and metastases. Due to the rarity, it was usually not until during the surgery was the diagnosis of multiple cavernous hemangiomas considered. The typical CT findings of orbital cavernous hemangioma are well-defined round-shaped intraconal masses with smooth contours which show minimal orbital bone expansion and enhance with intravenous contrast medium. These features allow differentiation from diffuse orbital processes, such as most of the pseudotumor, some of lymphoma, leukaemia, Wegener's granulomatosis vasculitides and metastases. In all our cases, the diagnosis was made until surgical excision was done.

In addition, bilateral or multiple hemangiomas may exist in the setting of underlying systemic hemangiomatosis (cutaneous or visceral lesions) and should alert the clinician to the possibility of a number of syndromes that may have life-threatening consequences.

Orbital cavernous hemangioma with multiple lesions usually need lateral orbitotomy, because all tu-

mors should be removed and anterior route could not provide us with a large surgical field. However, patients with anteriorly located tumors can be treated with anterior orbitotomy, as what we chose in case 1 and case 2. Just as the unilateral solitary lesion, multiple lesions can be easily removed during the surgery. In our series, all tumors were successfully removed except in case 3 where the second mass was incompletely removed. It was noteworthy that CT or ultrasound scan may miss some of the masses, so we should find out all masses in the orbit during the surgery. All patients acquired good surgical outcomes with no postoperative complication.

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