



Multiple Orbital Hemangiomas: A Rare Case Presentation

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Intraorbital cavernous hemangiomas are the most known benign vascular tumors of the orbit in adults, however, multiple lesions diagnosed as cavernous hemangiomas in the orbits are very rarely reported. The most common presenting complaints are slowly progressive painless proptosis. They rarely present with vision impairment. Multiple cavernous hemangiomas, although rare, systemic evaluation of the patients should be done in these cases to rule out multi-centric lesions. Complete excision of the tumors should be done surgically, and the patients need to be followed on a long-term to monitor for recurrence. Here, we report a case of multiple unilateral cavernous hemangiomas in a 44-year-old male patient, who was treated as a case of neurofibroma clinically based on radiological investigations.

Key words: Multiple cavernous hemangiomas, neurofibroma, benign tumors, orbital tumors

INTRODUCTION

The most frequent benign vascular tumor in individuals in their fourth and fifth decades is cavernous hemangioma of the orbit (CHO). Between 4.5% and 7.4% of primary and secondary orbital cancers are of this kind.¹ Slowly progressing painless proptosis is the most frequent complaint. When the lesions are deeper or have injured the surrounding tissues, such as the optic nerve, they present with visual impairment to total blindness.² The diagnosis of cavernous hemangiomas involves using radiological tests including magnetic resonance imaging (MRI) and computed tomography (CT). Numerous or bilateral lesions within the orbit are seldom recorded, and most tumors are unilateral or solitary.³ Here, we report a case of multiple unilateral cavernous hemangiomas, which was provisionally diagnosed as neurofibroma.

CASE REPORT

A 44-year-old man who had a considerable decline in visual acuity for 6 months and painless progressive proptosis of the left eye for over 10 years presented to our institute.

The best-corrected visual acuity (BCVA) was 5/10 in the left eye and 10/10 in the right eye. The left eye's lateral eye movement was slightly limited. In the right eye, ocular motility and the direct and consensual photo-motor reflex were both still present. Luedde's had an 18 mm proptosis. The remainder of the ophthalmologic examination was within normal limits. Exophthalmos was not present, and the ocular auscultation was normal. Both eyes had normal intraocular pressure. The diagnosis of neurofibroma or schwannoma was finalized by orbital imaging with the help of, B-scan, A-scan, CT [Figure 1], and MRI [Figure 2]. In this case, a left eye posterior orbitotomy with mass excision was performed and two lesions were removed and sent for further histopathological examination. One of the lesions could not be resected because of its proximity to the orbital nerve and was left behind.

We received two brown, soft tissue pieces. Grossly, the larger piece measured 2.5 cm × 1.5 cm × 1 cm and the smaller measured 0.9 cm × 0.3 cm × 0.2 cm. The external surface of both the tissue pieces was congested, gray, and brown on the cut surface. All the tissue was submitted for processing. On microscopic examination, variably sized vascular spaces

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lined by endothelial cells and filled with red blood cells were seen. No neural elements were seen in the sections studied. These microscopic features were suggestive of cavernous hemangioma [Figure 3].

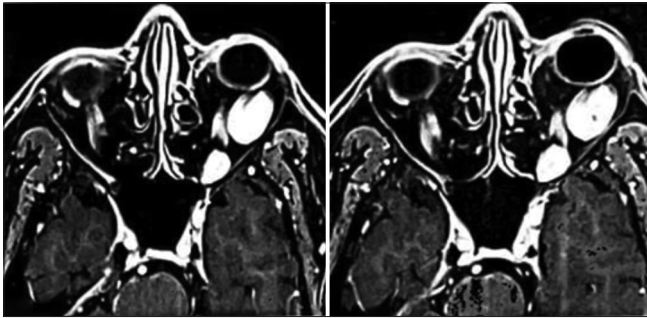


Figure 1: Magnetic resonance imaging – T2 weighted images showing left intraorbital, intraconal hyperintense. Lesions between optic nerve and lateral rectus



Figure 2: Computed tomography postcontrast image showing left intraorbital, intraconal lesions, showing intense enhancement

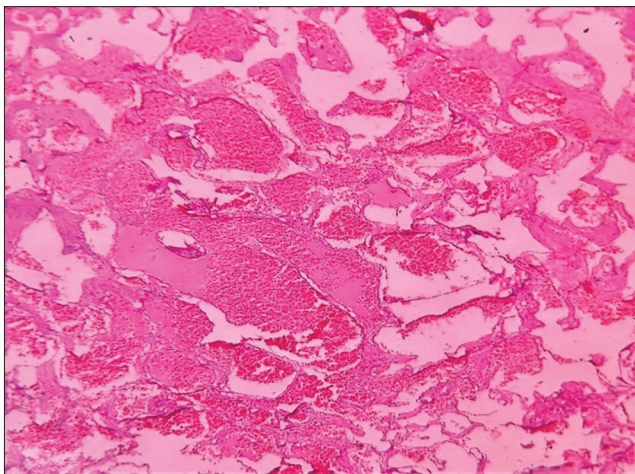


Figure 3: Photomicrograph showing variably sized vascular spaces lined by endothelial cells and filled with red blood cells (H and E, $\times 100$)

DISCUSSION

Majority of the cases of cavernous hemangiomas, which are benign orbital tumors, are made up of dilated blood vessels divided by fibrous tissue. They are usually discovered in middle-aged women.⁴ Multiple cavernous hemangiomas in the orbit have been recorded relatively infrequently.⁵ The intraconal compartment contains more than 80% of orbital cavernous hemangiomas, most frequently in the lateral aspect.⁶ About 60% of CHO cases are in women, and current research appears to indicate that female sex hormones may have an impact on the clinical course of CHO.

CHO should be included in the category of low-flow nondistensible venous malformations, according to the International Society for the Study of Vascular Anomalies. It is made up of a septate and thrombosed venous convolution that is encased in a tight, robust fibrous capsule.⁷ A well-circumscribed, round to oval, solid orbital mass may be caused by peripheral nerve sheath tumors (schwannoma and neurofibroma), hemangiopericytoma, fibrous histiocytoma, solitary fibrous tumor, or melanoma.⁸

Imaging of isolated neurofibromas reveals ovoid lesions with a smooth border that may or may not be lobulated. On a CT scan, they look isodense or hypodense to the extraocular muscles and exhibit varied contrast enhancement, according to some accounts. Low-moderate T1 signal intensity and moderate-high T2 signal intensity are shown on an MRI. The heterogeneous histology and vascularity of the tumors may be reflected in the variability of signal intensity inside the lesion. Like CT, MRI contrast enhancement varies.⁹

When cavernous hemangiomas are small on a CT scan, they appear as well-circumscribed, rounded, or oval soft-tissue density masses; as they enlarge, however, their soft nature causes them to become deformed. They are also somewhat hypoattenuating in comparison to the muscle, which gradually and incompletely fills in after the administration of contrast.

The appearance on MRI is morphologically the same as on CT, isointense on T1 relative to muscle, and if there are any sites of thrombosis, hyperintense regions may be seen. On T2, the hyperintense tissue may show low-intensity septation and low-intensity pseudo capsule relative to muscle.¹⁰⁻¹²

Most lesions of CHO are round to ovoid, and larger lesions have somewhat lobulated margins which can distort the surrounding structures and can lead to visual complications. Hence, it is important to surgically remove these lesions as soon as possible to prevent further visual compromise.³ Along with radiological investigations, histopathology plays an important role to make the final diagnosis of cavernous hemangiomas. Surgery is the treatment of choice in cases of cavernous hemangiomas. The treatment for localized neurofibroma is decided based on the severity of symptoms

and clinical signs. Postsurgical visual improvement is reported in most cases.

In this case, the clinical diagnosis made was neurofibroma of the left eye based on CT and MRI findings. The postoperative histopathological examination led to the final diagnosis of cavernous hemangiomas. On follow-up, the postsurgical recovery was uneventful. On postoperative physical examination, the BCVA was improved to 8/10 in the left eye.

CONCLUSION

When there are several orbital mass lesions, cavernous hemangiomas should be ruled out as a possibility. Although uncommon, multiple cavernous hemangiomas should be ruled out in these circumstances by doing a systematic assessment of the patients. Surgery should be undertaken to completely remove the tumors, and patients should be monitored for a long time to check for recurrence.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

Data availability statement

The data that support the findings of this study are available from the corresponding author, PL Nibe, upon reasonable request.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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