

# Primary Orbital Hemangiopericytoma: An Unusual Cause of Unilateral Proptosis

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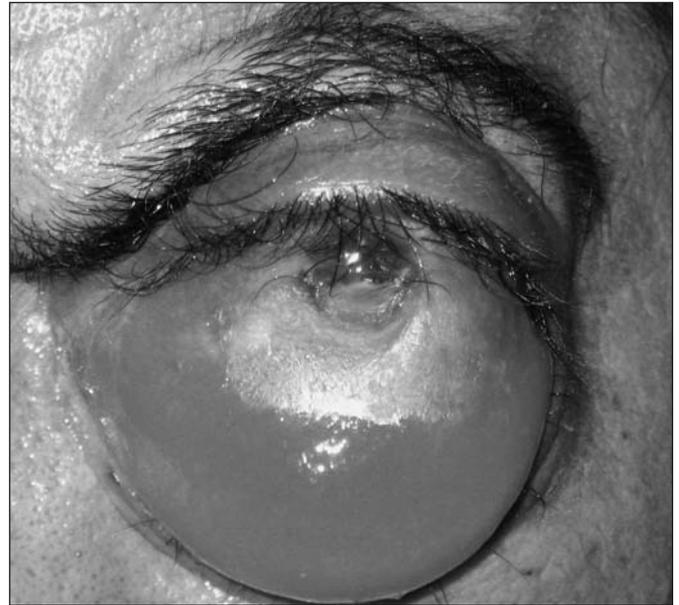
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Painless unilateral proptosis is a frequent manifestation of numerous orbital neoplastic and non neoplastic processes. Various mesenchymal tumours of both fibrohistiocytic and vascular origin are well-described causes. Hemangiopericytomas (HPC) are rare vascular tumours which can infrequently involve the orbit and their incidence is estimated to be 0.8% to 3% of primary orbital tumours<sup>1,2</sup>. We herein report a new case of orbital HPC revealed by unilateral proptosis in a 38-year-old man. Our aim was to highlight the clinicopathological and radiological features of this rare neoplasm with review of the current literature.

## CASE REPORT

A 38-year-old previously healthy man, presented with a one-month history of rapid expanding intra-orbital mass resulting in painless proptosis of the left eye. On admission, ophthalmologic examination revealed severe left eye proptosis (grade III). A palpable mass in the left orbit was protruding out of the palpebral fissure (Figure 1). It was irreducible, non-tender and non-pulsatile. The conjunctiva was chemotic and there was marked ulcerative keratitis. The visual acuity was 0 / 10 in the affected eye and ocular movements were restricted in all directions. The contralateral eye was within normal limits. Computed tomography (CT) scan of the orbital region (Figure 2) revealed a 87 x 54 x 32 mm, well-circumscribed mass within the intraconal space of the left orbit extending along the optic nerve and homogeneously enhanced after intravenous administration of contrast medium. Magnetic resonance imaging (Figure 3) showed a soft tissue mass iso-intense on T1-weighted images and hypo-intense on T2-weighted images. Laboratory tests and chest radiograph were within normal limits. An incisional biopsy of the intra-orbital mass was performed but was difficult to realize due to excessive haemorrhage. Histological examination of the tiny biopsy specimen obtained, established an initial diagnosis of an undifferentiated sarcoma. Orbital exenteration was carried out due to the infiltrative and highly vascular nature of the tumour.

Macroscopically, the exenteration specimen measured 130 x 120 x 105 mm. A small atrophic globe at the anterior aspect of the specimen did not appear invaded by the tumour. The cut section of the mass showed a firm greyish white tumour. The eyelids were not infiltrated by the tumour. Histological examination of the surgical specimen showed a hypercellular neoplasm predominantly composed of randomly oriented polygonal and rounded cells with ill-defined cytoplasm (Figures 4A & 4B). Large and small blood vessels lined by a single layer



**Figure 1:** Preoperative case photograph showing severe proptosis of the left eye and marked conjunctival chemosis.

of endothelium with a branching or “staghorn” appearance were readily apparent (Figures 4A & 4B). Mitotic figures were scarce and did not exceed 1 mitosis per 10 High power fields (HPF). Necrosis, psammoma bodies, intranuclear pseudo-inclusions and whorl formations were not identified. Immunohistochemically, tumour cells stained for vimentin and factor XIIIa but were negative for CD34, glial fibrillary acid protein, S-100 protein, epithelial membrane antigen, cytokeratin, neurofilament protein, desmin, muscle-specific actin, smooth muscle actin and bcl-2. The MIB-1 (Ki-67) labelling index was approximately 1 %.

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**Figure 2:** Preoperative axial CT scan showing a well-circumscribed spontaneously dense intraconal mass extending along the optic nerve.



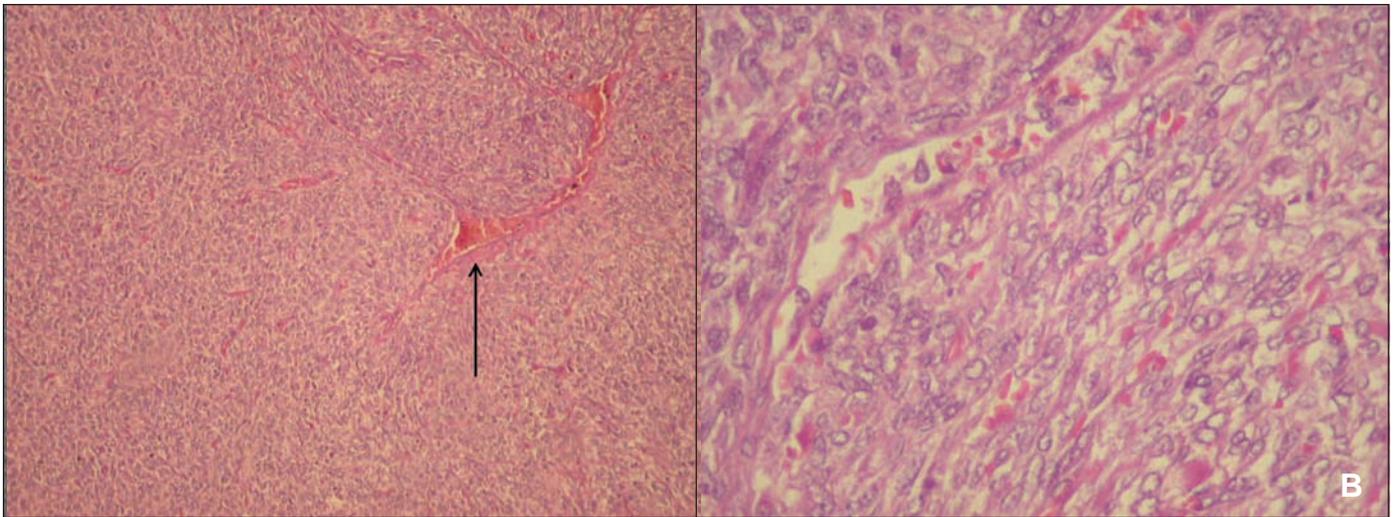
**Figure 3:** Coronal magnetic resonance imaging showing an iso-intense intra-orbital mass.

final pathologic diagnosis was hemangiopericytoma. No radiation therapy was administered post-operatively. During the six-month follow-up period, the patient did show evidence of recurrence.

#### DISCUSSION

Hemangiopericytoma (HPC) was first documented as a distinct vascular tumour of soft tissue by Stout and Murray in 1942<sup>3</sup>. It is proposed to derive from a pluripotential perivascular mesenchymal cell and can therefore occur anywhere in the body where there are capillaries. The localization of HPC in the orbit is particularly rare and mostly isolated cases or small series have been reported in literature. Orbital HPC generally affects adults with no gender predilection<sup>4,5</sup>. It is a slow-growing tumour with a symptomatic duration ranging from one month to several years<sup>4,6,7</sup>. In our patient, the symptomatic duration was one month. Clinically, orbital HPC often presents with painless proptosis and downward displacement of the globe. Intermittent upper lid swelling with blue or red discoloration of the adnexa oculi is occasionally noted. The chief complaint in our patient was perception of a rapid expanding intra-orbital mass resulting in painless proptosis of the left eye. Many previous reports described tumour growth as causing relatively little disturbance of visual function, which was not the case in our patient who lost vision in his affected eye<sup>4,6-8</sup>. The tumour tends to be non-tender and non-pulsatile as it was the case in our patient. Orbital CT

often reveals a superiorly located, well-circumscribed, mass occupying either the extraconal or intraconal space, which is intensely enhanced after contrast medium injection<sup>8-10</sup>. In the presented case, CT scan revealed a well-circumscribed mass within the intraconal space of the left orbit extending along the optic nerve and intensely enhanced after contrast medium injection. Magnetic resonance imaging may reveal a mass of isointensity (to grey matter) in T1-weighted images and a mass of isointensity to slightly high intensity in the T2-weighted and proton-density images<sup>8-10</sup>. In view of the aggressive and potentially malignant nature of the tumour, complete and intact removal is the ideal treatment of orbital HPC<sup>5</sup>. In case of incomplete excision, adjunct radiotherapy may be beneficial in reducing recurrence. The local recurrence rates of primary orbital HPC vary from 13% to 40 %<sup>5,11,12</sup>. Adjunct radiotherapy and chemotherapy can be used in cases of local recurrence<sup>13</sup>. Exenteration has been suggested by Sullivan et al because of the infiltrative pattern rather than circumscription in cases of local recurrence<sup>14</sup>. By far, the definite diagnosis of HPC must depend on pathological examination of the surgical specimen. Grossly, the majority of the tumours are fairly circumscribed and some are encapsulated. On microscopic examination, HPC are monomorphous tumours composed of randomly oriented tumour cells with little intervening fibrosis. Cytoplasm is scant and cell borders are indistinct. Nuclei are round to oval with moderately dense chromatin and inconspicuous nucleoli. Nuclear atypia and



**Figure 4:** A) Highly cellular tumour with dilated, staghorn-type vessels (arrow). (Haematoxylin and eosin staining; original magnification, x 100). B) Spindle to ovoid-shaped cells with uniform nuclei and ill-defined cytoplasm. Some irregular vascular channels are present. No mitotic figures are identified. (Haematoxylin and eosin staining; original magnification, x 400).

mitotic activity vary. A rich network of reticulin fibres, typically investing individual cells is one of the most characteristic but not invariable features of this neoplasm<sup>15</sup>. Hemangiopericytoma is highly vascular, with numerous slit-like vascular channels lined by flattened endothelial cells and frequent gaping thin-walled and branching vascular spaces, so-called “staghorn sinusoids”. Immunohistochemically, neoplastic cells are diffusely immunoreactive for vimentin (85%), factor XIIIa (80-100%) in individual scattered cells, Leu-7 (70%) and in 33-100% of cases, for CD34. The latter is usually patchy, in contrast to the diffuse pattern typical of solitary fibrous tumour<sup>15</sup>. The main histologic differential diagnoses of orbital HPC include meningioma, solitary fibrous tumour and fibrous histiocytoma. Prediction of aggressive behaviour of HPC may be difficult. However, it has been reported that malignant clinical course is associated with large size of the tumour (>5 cm), increased mitotic rate (4 or more mitotic figures per 10 HPF), a high degree of cellularity, immature and pleomorphic neoplastic cells, foci of hemorrhage and necrosis<sup>16-18</sup>. In our case, the main factors associated with prognosis were large tumour size and high degree of cellularity. Hemangiopericytoma has evoked controversy since its initial description. Recent articles have questioned whether it is a “histopathologic pattern” or a “clinicopathologic entity”, a “diagnosis of exclusion” a “wastebasket diagnosis” or a “dying breed”. The dubiousness attached to HPC stems from a number of features. First, the histological, immunohistochemical and ultrastructural features of HPC are not specific. Second, the marked variation in the clinical course and histological grading do not necessarily correlate with behaviour. Third, there is a lack of understanding about the nature of pericyte<sup>19-22</sup>.

In approximately 12 to 45% of cases orbital HPC metastasises<sup>5,23</sup>. The most common sites for metastases are: lung, bones, mediastinum and liver. The long interval between the

onset of symptoms and the first manifestations of metastasis strongly suggests that long follow-up periods are mandatory for more accurate evaluation of the biological potential of these tumours.

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