Case report

Intraorbital hemangiopericytoma: a rare case report

1Vaanika Kaira, 2Shruti Semwal, 3Asha Agarwal, 4Pankaj Kaira

1Senior Resident, Department of Pathology, Pt B.D. Sharma PGIMS, Rohtak, Haryana
2Senior resident, Department of Pathology, AIIMS, Bhopal
3Professor, Department of Pathology, GSVM Medical College, Kanpur
4General practitioner, Riyadh, Saudi Arabia

Corresponding author: Dr Shruti Semwal

Abstract

Introrbital hemangiopericytomas are rare tumors of fibroblastic origin, with an incidence of 0.8-3% and only a few cases have been described in literature. We present a case of a 34 year old woman with complaint of progressive diminution of vision in left eye presenting with sudden loss of vision and severe proptosis. CT scan revealed a large mass in the left orbit. Surgical excision of lesion was done and on histopathological examination diagnosis of intraorbital hemangiopericytoma was made, which was confirmed on immunohistochemistry using CD 34. We are presenting this case because of its rarity and its probable confusion with epithelioid hemangioendothelioma on histopathological examination alone, because the later behaves in benign fashion, whereas intraorbital hemangiopericytoma may have potentially malignant behavior with high local recurrence rate in case of incomplete excision.

Keywords: intraorbital, hemangiopericytoma, CD34, rare

Introduction

Hemangiopericytomas are rare tumors derived from pericytes-myofibroblast like cells that are normally arranged around capillaries and venules.1 They were originally described by Stout and Murray in 1942, and account for approximately 1% of vascular tumors. Intraorbital hemangiopericytomas are even less common, with an incidence of 0.8–3%.2 They occur as slowly enlarging, painless masses at any anatomic site but are most common on the lower extremities especially the thigh and in the retroperitoneum.1 They may be found in the lungs, bones, skull, but orbital hemangiopericytoma is a rare presentation. Mainly affects adults between the third and sixth decade of life. The tumor has an unpredictable prognosis and when it affects the orbital region, may have an aggressive behavior, with high incidence of recurrence.2

Case report:

A 34 year old woman with complaint of progressive diminution of vision in left eye presented with sudden loss of vision and severe proptosis. CT scan revealed a large mass in left orbit. Surgical excision of lesion was done and specimen was submitted for histopathological examination.

On gross examination the mass was well encapsulated, globular, greyish brown in colour measuring 7.5x6.5x4.0 cms. External surface revealed some congested blood vessels. Cut surface was largely solid, greyish brown with areas of hemorrhage, necrosis and few cystic areas. Specimen grossed, processed for blocking and
haematoxylin and eosin (H/E) stained slides were examined.

On microscopic examination H/E stained sections revealed a cellular tumor comprising of oval to spindle shaped cells. The tumor cells were arranged around a sinusoidal vascular component forming staghorn channels. Large areas of necrosis and chronic inflammatory infiltrate was evident at places. The number of mitotic figures were >5/10 high power field. [Fig 1,2]

Based on these findings the tumor was diagnosed as Hemangiopericytoma which was confirmed on immunohistochemistry using CD34, which revealed staghorn pattern of vasculature. [Fig 3]

**Discussion**

Hemangiopericytoma are rare tumors mainly found in deep soft tissues of lower extremities and retroperitonium. Orbital hemangiopericytoma is generally a solid, slow growing tumor, without apparent predilection for race and gender. Although it preferentially affects adults between the third and sixth decades of life, it can also occur in children. Symptoms usually include slowly progressive unilateral proptosis, which may be accompanied by mild pain and decreased visual acuity. It can be found anywhere in the orbit but has a predilection for the upper orbit. Most are small, but they occasionally achieve a diameter of 8 cms. Orbital hemangiopericytomas should be considered in the differential diagnosis of well defined orbital tumors along with epidermoid cyst, fibrous histiocytoma, lipoma, schwannoma, neurofibroma and vascular malformation.

Microscopically the most close mimic is hemangioendothelioma, chiefly the epithelioid variant, that have numerous capillary channels which are surrounded and enclosed within nests and masses of rounded endothelial cells. Epithelioid hemangioendotheliomas behaves in a benign fashion and rate of recurrence after complete surgical recurrence is very low. Imaging studies are not pathognomonic in the diagnosis of orbital hemangiopericytoma but most of the tumors appear as well defined lesion with or without contrast enhancement. The diagnosis is confirmed by histopathologic examination and sometimes complemented by immunochemistry. The characteristic histopathology is cell proliferation with mild to moderate pleomorphism, arranged in bundles interspersed with numerous angulated vessels. On immunohistochemical study, the immunoreactivity of CD34 and factor VIII favors the diagnosis. Special stains (Reticulin) confirm that these cells are outside the endothelial cell basement membrane and are therefore pericytes.

In histologically diagnosed case it is very difficult to predict the behaviour of the tumor, whether malignant or benign. Tumor diameter more than 6.5 cm, mitotic figures >4/10 hpf, hemorrhage, necrosis, cellular anaplasia and increased cellularity associated with thrombosis are the criteria of malignancy, proposed by Enzinger and Smit. MacMaster, et al stated that even mild degree of anaplasia with one mitotic figure /10 hpf or moderate degree of cellular anaplasia with one mitotic figure/10 hpf are sufficient evidence to predict malignant potential of tumor.

With regard to treatment, there is unanimity on the need for complete excision with wide margins. Both radiotherapy and chemotherapy have shown controversial results, and are usually used only in cases of relapse. Various surgical routes can be used to expose orbital lesion. Incomplete removal of orbital hemangiopericytomas is apparently a factor contributing to eventual recurrence. The tumors may recur after excision, and roughly half will metastasize, usually hematogenously to brain, lungs, bone or liver.
Conclusion
In view of potentially malignant behaviour of orbital hemangiopericytoma, complete and intact surgical resection should be attempted at first surgery. In case of histologically aggressive tumors long term follow up may be needed to detect an early recurrence to improve overall survival rate and outcome.
Fig 1. Photomicrograph of tumor specimen showing closely packed small cells with hyperchromatic nuclei between numerous thin walled vessels and capillaries.(H & E, 100X)

Fig 2. Photomicrograph of tumor showing areas of necrosis and infiltration by mixed inflammatory infiltrate.(H & E, 100X)

Fig 3. Photomicrograph showing CD34 positivity and revealing characteristic staghorn vasculature.(H & E, 100X)

Fig 4. Photomicrograph showing thin walled vessels, many capillaries and tumor cells marked by reticulin sheaths.(Silver reticulin stain, 400X)

References