ORIGINAL RESEARCH PAPER

Pathology

KEY WORDS: Cavernous Haemangioma, Lymphovenous Malformation, Coexistence, Orbital

ORBITAL CAVERNOUS HEMANGIOMA WITH LYMPHOVENOUS MALFORMATION

Dr. Sumaira Siddiqui Dnb Pathology Student Regency Hospital , Kanpur 208005

Dr. Deepti Gupta* M.D., D.N.B Pathology Consultant Pathologist Regency Hospital , Kanpur 208005 *Corresponding Author

Dr. Shefali Agarwal Diploma Pathology Consultant Pathologist Regency Hospital , Kanpur 208005

Dr. Nupur Trivedi M.D. Pathology Consultant Pathologist Regency Hospital, Kanpur 208005

Dr Anjali Tewari M.D. Pathology Consultant Pathologist And Head Of Department Of Lab Sciences Regency Hospital, Kanpur 208005

Dr. Asha Agarwal M.D. Pathology Consultant Pathologist And Head Of Academics Regency Hospital, Kanpur 208005

ABSTRACT

INTRODUCTION: Cavernous hemangioma although not a true neoplasm, is the most common benign adult orbital tumour. The patient usually presents with painless slowly progressive bulging of the globe. An orbital venous-lymphatic malformation is a rare lesion. And a coexistence of orbital cavernous hemangioma with lymphovascular malformation is even more unusual with only a few case reports in the literature so far.

CASE REPORT: We hereby describe the clinical, radiological and histopathological features of a case of orbital cavernous hemangioma with coexisting lymphovenous malformation in an 11 year old girl child. The girl came to our ophthalmology department with a slowly increasing swelling on the medial aspect of her right eye ball since birth, which on histopathological examination revealed both cavernous haemangioma and lymphovenous malformation in coexistence.

CONCLUSION: This entity represents an association of different types of orbital malformations with probably same pathogenesis.

INTRODUCTION

Vascular lesions of the orbit have always been an enigma to the pathologist and ophthalmologist. They comprise around 7% of all orbital tumours. Visual loss and presentation may vary from chronic to acute and even catastrophic. Consequences include dense amblyopia, blindness, globe dystopia, orbital and orbitofacial deformity, pain and ulceration. Non recognition of incidental, non-contiguous clinical intracranial vascular anomalies may result in intracranial vascular incidents with residual severe morbidity [1]. Cavernous hemangiomas are the commonest vascular lesions of the orbit in adults, being commonly seen in middle age, with a female predilection [2]. Over 80% of orbital cavernous hemangiomas are located within the intraconal compartment, most commonly in the lateral aspect [3]. The lymphovenous malformations are vascular anomalies with inappropriate connections within the vasculature presented by vascular malformation. These malformations invade the normal tissues causing difficulties in the normal blood circulation [4]. Principal reasons for the wrong diagnosis and poor management of orbital vascular tumours include the lack of clear understanding of etiopathogenesis, their natural history, disparity between terminology and the underlying pathology.

CASE REPORT

An 11 year-old girl came to our ophthamology department with a history of a mass on the medial aspect of her right eye ball since birth. The size of the lump kept increasing with age. Over the past 10 months she developed right sided proptosis with intermittent swelling of the lower lid associated with blue discoloration of the overlying skin and inferior displacement of eye ball. The swelling did not alter with change in posture. She had no pain at any stage, and there were no associated visual symptoms but since it was abutting the nose, it created problems in reading.

Haematology laboratory tests - blood counts, urinalysis and electrolytes were within normal range. Chest x- ray was normal. Ophthalmic examination revealed a best corrected visual acuity of 6/9 in right and 6/6 in left eye. There was 3 mm of left axial proptosis and the right globe was displaced 3mm downwards. Intraocular pressure and fundus were unremarkable. Two small firm pea-sized masses were palpable, one above the eye ball and the other on medial aspect of eye ball. The rest of her ophthalmic examination was normal.

Contrast Enhanced Computed tomography (CECT) showed two round lesions, one approximately 14×13×8mm sized soft tissue density lesion in superior extraconal compartment and the other measuring 5×5×6mm in the medial compartment of right orbit. Extraocular muscles were normal. The rounded masses showed variable contrast enhancement. The superior ophthalmic veins were not adequately demonstrated, but the cavernous sinuses were normal. A probable diagnosis of Idiopathic inflammatory pseudotumour was made.

MRI showed a well-defined hypointense lesion on T1W1 and hyperintense lesion on T2W1 measuring 1.6×2×1.5 cm in size in subcutaneous plane in extraconal compartment of right orbit at supero-medial aspect. A probable diagnosis of Capillary hemangioma was made.

UM-B scan showed normal intraocular study with orbital study showing a hypoechoeic lesion at 1.30 – 2.00 clock with entire extent of lesion not clear. Intra-arterial digital subtraction angiography was carried out with injections into the left Common, internal, and external carotid arteries. They suggested lymphovenous malformation.

A presurgical diagnosis of lymphovenous malformation was made and it was decided to remove the anterior component of the vascular anomaly in the right orbit. During surgery, two discrete masses measuring 3×1.5×0.5 and 1.5×1.4×1cm respectively were excised. On gross examination, the masses were reddish brown in colour. Microscopic examination revealed ecstatic blood-filled (cavernous) venous spaces alongwith cystic spaces filled with proteinaceous material {lymph} and abortive vessels spreading along the normal structures. A histopathological diagnosis of cavernous hemangioma with co-existing lymphovenous malformation was made. (Figure1) The post operative period was unremarkable and no visual impairment was noted.

DISCUSSION

Cavernous hemangiomas are the commonest intraorbital tumors. Other common sites are skin, face, extremities, liver, gastrointestinal tract and even thymus. These grow as unilateral, painless and progressive lesions. Cavernous hemangiomas are believed to be present at birth and keep growing overtime without any manifestations. They are more prevalent in females showing the peak presentation during middle age with an enlagement during pregnancy [5]. The lymphovenous malformations are vascular anomalies with inappropriate connections within the vasculature presented by vascular malformation. These malformations invade the normal tissues causing difficulties in the normal blood circulation [4].

Historically, the vascular lesions had been labelled purely by their clinical presentation, histopathology and were purely descriptive entities [6]. In 2014 the International Society for the Study of Vascular Anomalies (ISSVA) classified all vascular anomalies into vasoproliferative tumours and vascular malformations based on clinical, radiological (including angiographic features), histopathological features and the natural history. According to ISSVA classification, firstly, the tumors are separated from the malformations. The malformations are then categorized into slow and high flow and then subcategorized into arterial, venous, lymphatic or mixed. [7,8] The ISSVA classifies cavernous hemangioma as the venous malformation in the slow-flow lesion category. The cavernous hemangioma presents with 80% of the angiomatous lesions. The main manifestations are progressive painless proptosis while, some patients may present with abrupt orbital enlargement and acute-onset proptosis, which are caused due to cytokine, or hormonal disturbances during puberty and pregnancy [5].

The pathogenesis is unclear, although two theories exist. One of the theories suggests that hemangioma arises during the gestation period due to the placental tissue disruption embedded in the fetal tissues. The markers of the hemangioma are found in the placental tissues and in the infants following birth. These markers are also evident in the infants in the cases of placenta previa, chorionic villus sampling and preeclampsia. According to another theory, the endothelial progenitor and stem cells were observed among the patients with hemangiomas. This theory can be supported by the occurrence of hemangiomas in animals occurring from the stem cells extracted from human specimens [4].

The lymphatic malformations are due to dilated lymphatic vessels with limited connection lined by endothelial cells and filled with lymphatic fluid. These vessels extend to the nearby structures and present as thin walled structures with fluid filled spaces of varied sizes. They exhibit a tendency of haemorrhage and may lead to compression of the optic nerve due to sudden proptosis and swelling, with decreased eye movement [9].

Diagnosis: Magnetic Resonance Imaging is the most preferred diagnostic modality for studying the components of the lymphatic malformations. The imaging results present with infiltrative, multilobulated mass with minimal encapsulation. CT scan shows T1 weighed image showing lymphatic or proteinaceous fluid while T2 weighted fat images showing the components of non-hemorragic fluid [9]. CT and MR angiography fail to identify the feeding vessels due to low caliber. The Color Doppler imaging shows extremely low venous flow. The differential diagnosis include venous varix, schwannoma, optic nerve sheath meningioma and lymphoma [5]. The tumors are round to oval in shape with well-defined borders and a specific honeycomb pattern of alternating weak and strong echoes corresponding to their structure, with flow on color Doppler [3].

Clinical features: The most common manifestation is a slowly progressive painless proptosis. Few patients exhibit abrupt orbital enlargement with proptosis due to hormonal stimulation [10]. The compression of the orbital apex may cause optic nerve neuropathy leading to monocular loss of vision. The other manifestations include pain, swelling of the eyelid, lump or diplopia [5].

Management: The management aims in reducing the symptoms, improvement of the functions and aesthetics. The treatment options include symptomatic management, sclerotherapy and surgical excision. These methods can be used according to the diagnosis and symptoms, either one method or in multiple combinations [10]. Medical management includes the use of corticosteroids, interferon and vincristine. These agents are used in case of multifocal diseases, visceral disease, lesions in the periorbital area and airway obstruction [4]. The compression of the optic nerve indicates surgical treatment. The asymptomatic cases must be managed according to the extent of the disease. As the progress of the disease is very slow and the chances of recurrences are very rare, it is advisable to avoid total excision especially in a complicated case [5]. The treatment is indicated in case of pain, functional imparment, or esthetic problems [8].

CONCLUSION

Cavernous hemangioma with lymphovenous malformation is a rare condition, which requires appropriate diagnosis and selection of proper treatment method. The treatment of such anomalies is critical and involves various therapeutic options.

CONFLICT OF INTEREST

There is no conflict of interest between the authors.

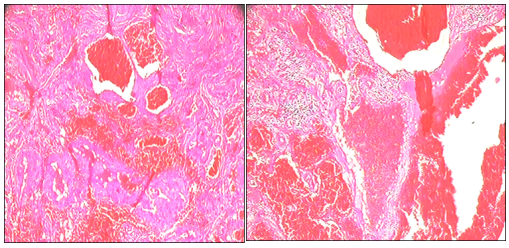

Figure 1 : H & E stained sections reveal ecstatic blood-filled (cavernous) venous spaces alongwith cystic spaces filled with proteinaceous material {lymph}.

REFERENCES

1. Katz SE, Rootman J, Vangveeravong S, Graeb D. Combined venous lymphatic malformations of the orbit (so-called lymphangiomas). Association with non-contiguous intracranial vascular anomalies. Ophthalmology 1998;105:176-84.
2. Ansari SA, Mafee MF. Orbital cavernous hemangioma: role of imaging. Neuroimaging Clin N Am. 2005;15(1):137-58.
3. Anand R, Deria K, Sharma P, Narula MK, Garg R. Extraconal cavernous hemangioma of orbit: A case report. Indian J Radiol Imaging 2008 Nov;18(4):310-2.
4. Richter GT, Friedman AB. Hemangiomas and Vascular Malformations: Current Theory and Management. http://dx.doi.org/10.1155/2012/645678.January17;2012.
5. 5. Kumar K. Cavernous Hemangioma of Orbit; Imaging Features. International

Journal of Recent Scientific Research. 2015 Sept 6;6(10):6941-6.
6. Rootman J, Kao SC, Graeb DA. Multidisciplinary approaches to complicated vascular lesions of the orbit. Ophthalmology 1992;99:1440-6.
7. 7. Available from: http://www.issva.org/classification.
8. Kim JH, Choi JG, Son Bc. Venous Malformation (Cavernous Hemangioma) of the Supraorbital Nerve. Asian J Neurosurg. 2018 April-June; 13(2): p. 499-502.
9. Strianese D, Russo C, Napoli M, Derrico A, Scotti N, Puoti G, et al. Coexistence of Cavernous Hemangioma and Other Vascular Malformations of the Orbit- A Report of Three Cases. The Neuroradiology Journal. 2014; 27: p. 223-231.
10. 10. Benotoin LA, Chan K, Steiner F, FitzJohn T, Tan ST. Management of orbital and periorbital venous malformation. Frontiers in surgery. 2017 May 29;4(27).