Intravascular Papillary Endothelial Hyperplasia as an unusual diagnosis for peri-orbital tumour - A case report

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Intravascular papillary endothelial hyperplasia (IPEH) is a rare cause of orbital mass which can affect healthy individuals of any age/sex/race/region. It usually requires a histopathological examination, characterized by the appearance of papillary proliferation of vascular endothelial cells within the lumen of the blood vessel, for the final diagnosis. One should be aware that this is a highly vascular lesion which can cause excessive intraoperative bleeding and incomplete removal can lead to recurrences. IPEH of the orbit/eyelid has been reported in few parts of the world as isolated case reports but none from India so far.

Key words: Intravascular papillary endothelial hyperplasia, Masson's hemangioma, orbital tumour

The clinical entity of intravascular papillary endothelial hyperplasia (IPEH) was first described in 1923 by Pierre Masson, within an ulcerated hemorrhoidal vein of a 68-year-old man.[1] He described it as a benign tumor occurring due to the papillary proliferation of vascular endothelial cells within the lumen of the blood vessel.[2] Masson called it "hemangioendotheliome intravasculaire" and was later described by many other names such as Masson's hemangioma, Masson's intravascular hemangioendothelioma. However, it was in 1976 that Clearkin and Enzinger proposed the name of IPEH.

IPEH has often been reported involving different parts of the body including head, neck, and extremities but rarely involving the eye or ocular adnexa. We report a case of IPEH arising in the superonasal quadrant of the orbit in an otherwise healthy male.

Case Report

A 58-year-old male, who was otherwise in good health, presented to us with a history of gradual onset of a slowly progressive painless swelling in the upper nasal quadrant of the orbit for 2 years. It was not associated with any pain or other ocular symptoms. The patient did not give history of trauma or any other systemic ailments or swellings elsewhere in the body.

Ocular examination revealed a best-corrected visual acuity of 20/20 in both eyes with normal intraocular pressure and posterior segment examination in both eyes. The left orbit showed a diffuse mass measuring 20 mm × 10 mm in the upper nasal quadrant [Fig. 1]. The eyelids were free from involvement and had complete movements. There was no proptosis and ocular movements were full and complete in both eyes. A plain computerized tomography scan of the brain and orbit was performed which showed the presence of a soft-tissue density mass lesion (HU 40–60) with few internal fat densities in the superonasal quadrant of orbit, measuring 12 mm × 8 mm in size. There was no evidence of internal calcification or scalloping/erosions of the underlying bone. Rest of the

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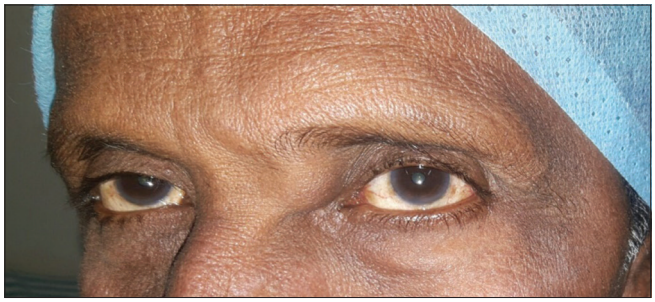

Figure 1: Showing a diffuse mass in the superonasal part of the periorbital area

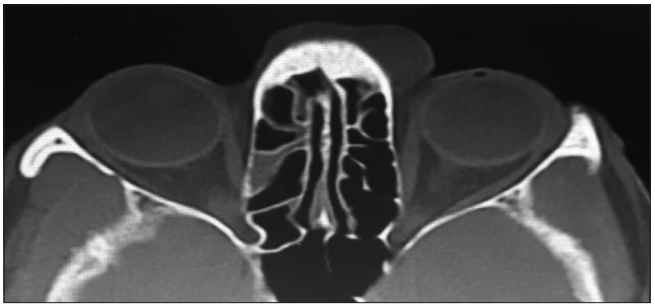

Figure 2: Plain CT scan showing a soft-tissue mass without involving/eroding the underlying bone

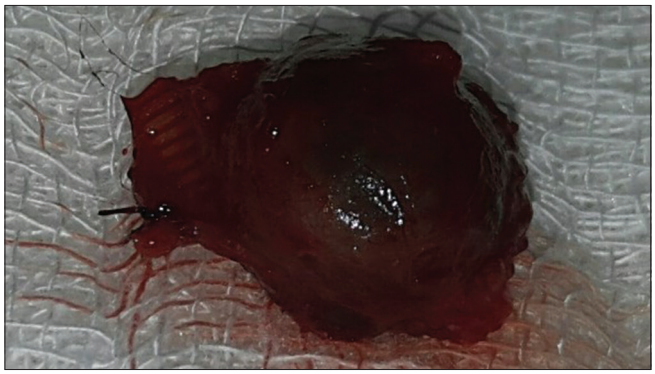

Figure 3: Excised lesion measuring 20 mm × 10 mm

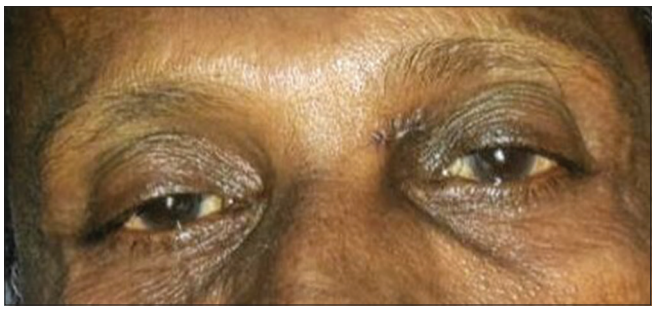

Figure 5: Postoperative picture

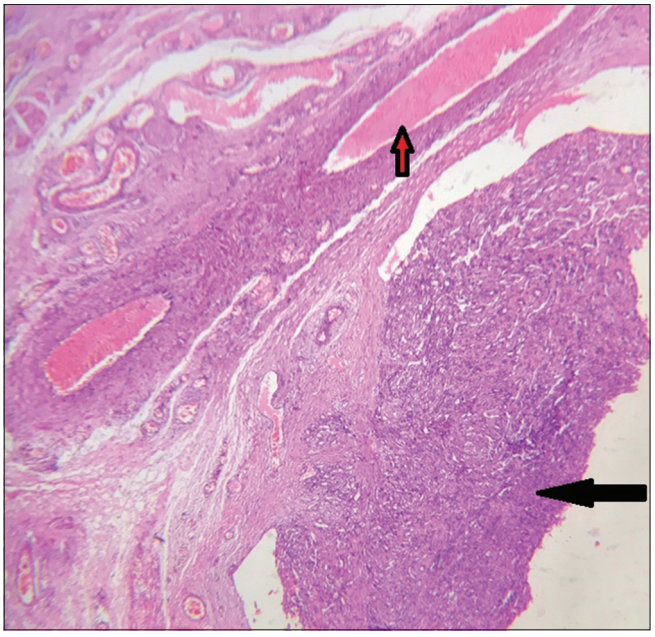

Figure 4: Histopathology showing small delicate papillae projecting into the lumen

structures of eye and brain were found to be normal [Fig. 2]. The patient underwent complete excision of the mass under local anesthesia. Profuse bleeding was noted during surgery and was attributed to the presence of a feeder blood vessel which was ligated before complete excision in toto [Fig. 3].

The excised intact mass was sent for histopathological examination which revealed a well-circumscribed tumor mass measuring 20 mm × 10 mm, composed of small delicate papillae projecting into the lumen [Fig. 4]. The papillae were composed of a single layer of endothelial cells surrounding a collagenized core. A diagnosis of IPEH was made. The patient was followed up for 20 weeks and was free from symptoms and showed no signs of recurrence [Fig. 5].

Discussion

Swelling in the orbital region seldom includes IPEH as differential diagnosis due to its rare occurrence. IPEH is a benign tumor which is usually not associated with any systemic diseases and is seen as an isolated mass involving medium-sized veins.[3]

There have been two theories which have been proposed to explain the pathogenesis of the lesion. The first one by Masson, in 1923, states that proliferation of endothelial cell is a primary phenomenon while thrombus arises secondary to it. However, in 1976, Clearkin and Enzinger proposed that the papillary projections appear after a preexisting thrombus organizes. At present, it is suggested that there is a reactive proliferation of endothelial cells following traumatic vascular stasis.[4] IPEH formation is said to be triggered by the release of fibroblast growth factor by macrophages which invade the site of trauma and causes proliferation of endothelial cells.[4]

Ocular involvement is relatively rare. Skin and subcutaneous soft tissue of head, neck, and extremities are most commonly involved in IPEH, although there have been reports of their locations in the liver, uterus, and gastrointestinal tract (GIT).[5,6] The first accepted case of eye lid IPEH was in 1974 by Wolter and Lewis and of IPEH in the orbit was reported by Weber and Babel in 1981.[7]

In 1997, Werner *et al*. reported a collection of 4 cases and a review of literature of IPEH.[1] In all their cases, there was the presence of thrombi in the specimen, strengthening

the theory that this tumor arose as the secondary process. The endothelial proliferation was the response to proximal thrombus inside the lumen of blood vessel rather than being the primary event.

There have been reports of presentation of orbital IPEH as eyelid mass,[1,3,8] unilateral or bilateral proptosis,[2,6,7] progressive diplopia,[5] mass in the supraorbital fissure,[1,5,9] ptosis/ectropion/deep lesion in the orbit when involving the ophthalmic or other orbital vessels.[10] Conditions which resemble the histopathological picture of IPEH are angiosarcoma, angiolymphoid hyperplasia with eosinophilia, and other vascular lesions. However, the intravascular nature of the papillary hyperplasia was the key point to differentiate this tumor from angiosarcoma, which invades tissues outside the lumen of the blood vessel.[2,6]

Age of patients ranged from 3 to 80 years and size of the lesion from 0.2 to 2 cm. IPEH can occur as solitary or multiple lesions.[6] Although reported as a painless lesion, patients may develop pain if the lesion is preceded by a thrombosed varix. There have been instances of recurrence following incomplete removal.[1,5] Gamma knife radiosurgery was proposed to be an effective alternative modality of treatment for residual tumor.[5]

Conclusion

The purpose of reporting this case is to bring this uncommon diagnosis of periorbital IPEH into the differential diagnosis of orbital/eyelid swellings because it shows no predilection toward age/sex/race/region/laterality/solitariness. To the best of our knowledge, this is the first reported case of periorbital IPEH from India.

Declaration of patient consent

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

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Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Werner MS, Hornblass A, Reifler DM, Dresner SC, Harrison W. Intravascular papillary endothelial hyperplasia: Collection of four cases and a review of the literature. Ophthal Plast Reconstr Surg 1997;13:48-56.
2. Fasina O, Adeoye A, Akang E. Orbital intravascular papillary endothelial hyperplasia in a Nigerian child: A case report and review of the literature. J Med Case Rep 2012;6:300.
3. Schulze S, Hoerle S, Koop G, Barth PJ, Huegens-Penzel M, Strempel I. Intravascular papillary endothelial hyperplasia - A rare finding in the orbital region. Ophthalmologica 2008;222:213-5.
4. Mahapatra QS, Sahai K, Malik A, Mani NS. Intravascular papillary endothelial hyperplasia: An unusual histopathological entity. Indian Dermatol Online J 2015;6:277-9.
5. Ohshima T, Ogura K, Nakayashiki N, Tachibana E. Intravascular papillary endothelial hyperplasia at the superior orbital fissure: Report of a case successfully treated with gamma knife radiosurgery. Surg Neurol 2005;64:266-9.
6. Shields JA, Shields CL, Eagle RC, Diniz W. Intravascular papillary endothelial hyperplasia with presumed bilateral orbital varices. Arch Ophthalmol 1999;117:1247-9.
7. Weber FL, Babel J. Intravascular papillary endothelial hyperplasia of the orbit. Br J Ophthalmol 1981;65:18-22.
8. Kim JY, Lee MK, Kim HY. A case of intravascular papillary endothelial hyperplasia of eyelid. J Korean Ophthalmol Soc 2016;57:1299-302.
9. Patt S, Kaden B, Stoltenburg-Didinger G. Intravascular papillary endothelial hyperplasia at the fissura orbitalis superior: A case report. Clin Neuropathol 1992;11:128-30.
10. Dutton JJ, Gayre GS, Proia AD. In: Diagnostic Atlas of Common Eyelid Diseases. 1st ed. New York: CRC Press; 2007. p. 171-2.