Lacrimal gland myxoma

Drushti Parikh, Bipasha Mukherjee

Myxomas are rare neoplasms of mesenchymal origin. Cases of conjunctival, corneal, and orbital myxomas have been reported in the literature; however, to the best of our knowledge, there is no report of a lacrimal gland myxoma. We report a case of an orbital myxoma involving the lacrimal gland and its management.

Key words: Lacrimal gland, myxoma, orbit

Myxomas are true neoplasms of mesenchymal origin most commonly encountered in the heart, but may also arise from the bone, skin, skeletal muscle, genitourinary tract, and nasal sinuses. Fuch was the first to report an orbital myxoma in 1914, in a 40-year-old female patient.[1] Orbital myxomas are rare, benign tumors with few reported cases in the literature. They show no particular sex and racial predilection or genetic predisposition. We report a case of myxoma involving orbital portion of the lacrimal gland and its management.

Case Report

A 65-year-old Asian-Indian male presented to the Department of Orbit and Oculoplasty with complaints of painless protrusion of his right eye for the past 1 year [Fig. 1]. He gave history of significant progression over the past 3 months. His past medical history was unremarkable and he had no other systemic complaints. On examination, his best-corrected visual acuity was 6/6; N6 (Snellen) in both the eyes. Exophthalmometry revealed a 4 mm axial, nonpulsatile proptosis in the right eye with significant resistance to retropulsion. Extraocular motility was full in both the eyes. Orbital rims on both sides were normal though there was difficulty in insinuation in the superolateral area. Valsalva manoeuver revealed no change in the degree of proptosis. Computerized tomography (CT) scan of the orbits was requested, which revealed a well-circumscribed, oval, isodense soft tissue lesion in the right lacrimal gland area, with displacement of the surrounding extraocular muscles, suggestive of a lacrimal gland lesion, possibly a pleomorphic adenoma. The optic nerve appeared normal. No bony erosion was noted [Fig. 2].

The patient underwent excision biopsy of right orbital mass, through an upper eyelid skin crease incision. A 17 mm × 10 mm × 5 mm mass was excised from the superolateral region of the right orbit. It was cystic in appearance with gelatinous contents [Fig. 3]. The lacrimal gland was not identified separately. Histopathologic examination showed a capsulated tumor with myxoid hypocellular material with fibrocytes, scattered blood vessels, and few inflammatory cells. Study of deeper sections of the mass revealed few scattered spindle cells and blood vessels in myxoid matrix. There was no atypia or abnormal mitotic activity [Fig. 4]. These histopathologic features were consistent with the diagnosis of a myxoma. On the last follow-up, the patient's visual acuity was maintained at 6/6, and there was no recurrence of mass or proptosis.

Discussion

Orbital myxoma is an uncommon benign soft tissue tumor derived from connective tissue of mesenchymal origin.[1-6] Patients usually present with slowly progressive painless proptosis. Sudden increase in growth has been reported presumably due to myxoid degeneration of the contents as seen in this patient.[7] Isolated myxomas of the orbit are very rare and only a few cases have been reported so far. Other ocular sites of reported myxomas are in the conjunctiva, cornea, and eyelid. Orbital myxomas are also found in association with Carney complex, Mazabraud syndrome, and McCune–Albright syndrome.[1,4,6,8]

Carney complex is a syndrome comprising myxoma, spotty pigmentation, and endocrine overactivity. Myxomas in such cases are commonly encountered in the heart, breast, skin and other sites including the orbit. While examining a patient with an orbital mass, the presence of skin hyperpigmentation and a history of cardiac disease should prompt one to consider the possibility of an orbital myxoma in association with Carney complex. It is prudent to get a cardiac evaluation in

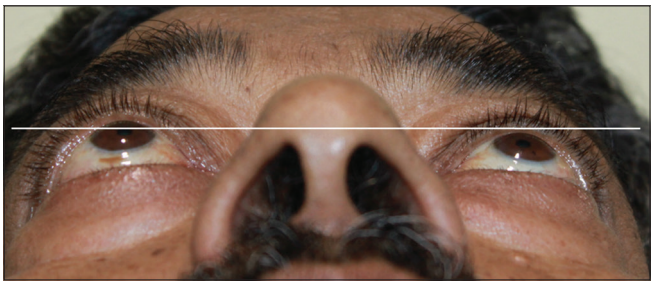

Figure 1: Clinical picture of patient showing right eye proptosis (Worm's hole view)

Access this article online

Quick Response Code:

Website: www.ijo.in

DOI: 10.4103/ijo.IJO_226_17

Department of Orbit, Oculoplasty, Reconstructive and Aesthetics, Sankara Nethralaya, Medical Research Foundation, Chennai, Tamil Nadu, India

Correspondence to: Dr. Bipasha Mukherjee, Medical Research Foundation, 18, College Road, Chennai - 600 006, Tamil Nadu, India. E-mail: beas003@yahoo.co.uk

Manuscript received: 25.03.17; Revision accepted: 28.05.17

Cite this article as: Parikh D, Mukherjee B. Lacrimal gland myxoma. Indian J Ophthalmol 2017;65:887-9.

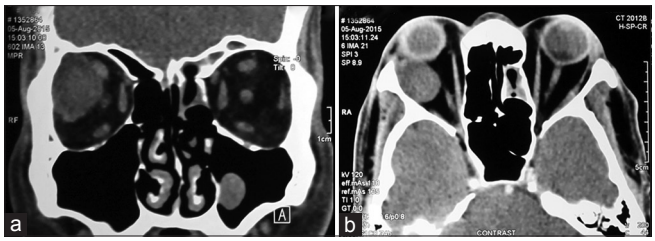

Figure 2: Coronal (a) and axial (b) sections of computerized tomography scans showing a well-circumscribed, oval, isodense soft tissue lesion in the right lacrimal gland area

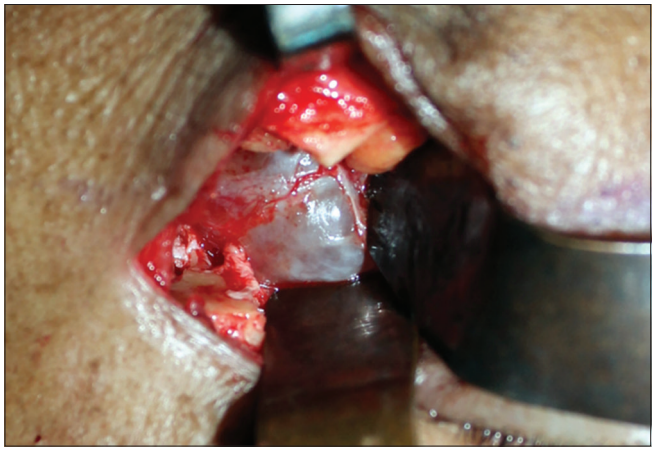

Figure 3: Intraoperative appearance of the lesion

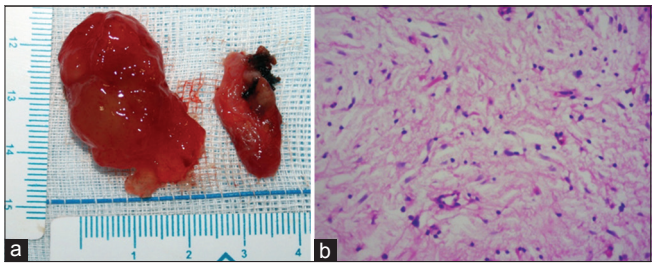

Figure 4: (a) Gross appearance of the tumour after excision. (b) Scattered spindle cells and blood vessels in myxoid matrix with fibrocytes and few inflammatory cells (H and E, ×100)

such patients. If any syndromic associations are suspected, additional diagnostic workup may be carried out, which include thyroid-stimulating hormone, adrenocorticotropic hormone, growth hormone level, and PRKAR1A gene analysis.[4] Kennedy *et al*. reported a case of orbital and eyelid myxomas, with truncal lentigines and a history of excision of benign testicular tumor and multiple excisions of nodular tumors from the eyelids. This patient had a positive family history of similar features and on cardiac examination revealed asymptomatic cardiac myxoma.[9]

Histopathologically, myxomas are characterized as hypocellular and hypovascular lesions with abundant myxoid matrix that is rich in hyaluronic acid.[3] They are characterized by presence of abundant mucoid material with sparse number of spindle- and stellate-shaped cells. Positive staining with alcian blue is a useful adjunct to establish the diagnosis of myxomas.[5,10]

Another entity which is histopathologically similar to myxomas is angiomyxoma. Unlike myxomas, they are characterized by presence of blood vessels interspersed within the stroma. They are more aggressive and infiltrative and tend to infiltrate the bony walls of the orbit and paranasal sinuses. Recurrence of a myxoma is comparatively rarer than angiomyxomas. Hidayat *et al*. studied six cases, three each of myxoma and angiomyxoma. They noted recurrence in one of the three cases of myxoma and two of the three cases of angiomyxomas. All the three were noted to have bony extension with or without paranasal sinus spread. Thus, they concluded that the vascularity of the tumor and the bony extension is a prognostic factor for recurrence.[3]

Myxomas should be differentiated from other neoplasms with similar histopathologic appearance. These include schwannomas, neurofibromas, and malignant tumors such as myxoid chondrosarcoma and malignant fibrous histiocytoma. The malignant tumors will typically show increased mitotic activity, pleomorphism, increased cellularity, and more number of blood vessels, unlike myxomas.[1]

Radiation therapy is not helpful for these tumors as they originate from fibroblastic cells and display very low mitotic activity.[3] Complete surgical excision is the preferred treatment modality.[3,4,10] Patient's needs should be closely monitored after surgery to look for recurrences. If complete surgical excision is ensured, the prognosis is excellent.

Conclusion

We report a unique case of lacrimal gland myxoma.

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.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Rambhatla S, Subramanian N, Gangadhara Sundar JK, Krishnakumar S, Biswas J. Myxoma of the orbit. Indian J Ophthalmol 2003;51:85-7.
2. Ting SL, Koay CL, Ngo CT, Chieng LL, Chua CN. Orbital myxoma – An unusual cause of inferoorbital mass. Med J Malaysia 2010;65:224-6.
3. Hidayat AA, Flint A, Marentette L, Torczynski E, Al-Qahtani JM, Ahl NC, *et al*. Myxomas and angiomyxomas of the orbit: A clinicopathologic study of 6 cases. Ophthalmology 2007;114:1012-9.
4. Demirci H, Shields CL, Eagle RC Jr., Shields JA. Report of a conjunctival myxoma case and review of the literature. Arch Ophthalmol 2006;124:735-8.
5. Mottow-Lippa L, Tso MO, Sugar J. Conjunctival myxoma. A clinicopathologic study. Ophthalmology 1983;90:1452-8.

6. Bielory BP, Turbin RE, Mirani NM, Barry MT. Radiographic and histological findings in an atypical orbital myxoma. Ophthalmol Eye Dis 2011;3:55.
7. Batsakis JG. Myxomas of soft tissues and the facial skeleton. Ann Otol Rhinol Laryngol 1987;96:618-9.
8. Kini Rao AC, Nayal B. Conjunctival myxoma-a case report. Malays J Med Sci 2013;20:92-4.
9. Kennedy RH, Flanagan JC, Eagle RC Jr., Carney JA. The Carney complex with ocular signs suggestive of cardiac myxoma. Am J Ophthalmol 1991;111:699-702.
10. Xiong MJ, Dim DC. Conjunctival myxoma: A synopsis of a rare ocular tumor. Arch Pathol Lab Med 2015;139:693-7.