A rare case of benign fibrous histiocytoma of the cornea

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Fibrous histiocytoma (FH) commonly occurs in the superficial layers of the skin. Orbit and limbus are documented ophthalmic sites of involvement but isolated corneal FH has never been reported in literature. We present the first case of FH exclusively involving the cornea where a 10-year-old male child presented with a 3-month history of a painless growth on the superior cornea of the right eye with deterioration of vision. Tumor excision with therapeutic penetrating keratoplasty was done and the histopathological examination confirmed the diagnosis. There was no recurrence and the corneal graft was clear at 1 year.

Key words: Benign fibrous histiocytoma, cornea, Fibrous histiocytoma, malignant fibrous histiocytoma

Fibrous histiocytoma (FH) encompasses a heterogeneous group of soft tissue tumors that are composed of cells that resemble fibroblasts and histiocytes, arranged in a cartwheel or storiform pattern.[1] This tumor most commonly occurs in the dermis and superficial subcutis, with rare deeper involvement. Ocular involvement of FH has been well documented in literature with orbit being the most common site.[1] Rarely, FH in both benign and malignant forms have been reported at the corneoscleral limbus. However, the tumor within the confines of the cornea has never been reported and we present a case of benign FH exclusively in the cornea. We further discuss the clinical and histopathological findings as well as the treatment options.

Case Report

A 10-year-old male child presented to the outpatient department of our hospital, with a 3 month history of a painless growth on the superior cornea of the right eye [Fig. 1a and b]. There was an associated vision deterioration over the past one month. He had no medical history of any systemic disorder or trauma and no associated cutaneous lesions.

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On examination, his visual acuity (VA) was 6/18 (unaided) in the right eye, which improved to 6/9 with correction. VA was 6/6 (unaided) in the left eye. The growth was present on the superior aspect of the cornea and was approximately 7 * 5.5 mm in diameter. It was extending from 10 to 2'o clock quadrant, vertically reaching up to the pupillary border. The lesion was vascularized with yellowish appearance. The Ultrasound Bio Microscopy (UBM) of the lesion revealed an elevated mass of 1.54 centimeters solely in the cornea without any posterior extension. [Fig. 2] Initially, the differentials were either foreign body granuloma, corneal abscess or a neoplasm.

The lesion was excised keeping 2 mm clear margin superiorly by taking a corneal button of 9.5 mm and therapeutic penetrating keratoplasty (TPK) was done. [Fig. 3] There was no post-operative complications.

Histological sections of the lesion showed a typical picture of benign FH with stromal hypercellular nodule consisting of spindle cells arranged in storiform pattern. The cells were admixed in numerous lymphocytes, histiocytes and also a few multinucleate giant cells. There were no mitotic figures or nuclear atypia. [Fig. 4a] There was cytoplasmic positivity for vimentin, CD 68 and factor XIIIA in the tumor cells [Figs. 4b-d].

The patient at one year had VA of 6/24 improving to 6/9 with correction in the right eye. No signs of recurrence were seen [Fig. 5].

Discussion

FH are soft tissue mesenchymal tumors that can be both benign and malignant, with the former being the more common.[1] The benign form is seen in the orbit, eyelid and episclera.[2] In the anterior segment of the eye, the most common site is the corneoscleral limbus and the conjunctiva. As per our literature search, there have been 36 such reported cases of FH out of which 22 were benign and the remaining 14 lesions were malignant. 26 cases showed presence of limbal FH, 8 conjunctival FH while 2 had caruncular FH.[3-5] However, a lesion confined to the cornea has not been reported earlier and ours is the first such case ever documented. The symptoms of the benign FH may include decreased vision, pain, restricted eye movements, diplopia, and disc swelling. The average age for benign tumors is between 25-35 years with a male preponderance.[3] The typical clinical picture is of a well-defined yellowish- white mass, at the limbus with adjacent vascularization. The malignant variety appears beyond 50 years of age.[3] The present case was peculiar as the patient was only 10-years-old.

Conjunctival FH have shown an association with trauma, leukemia, chemotherapy, prednisone, radiation, rheumatoid arthritis and xeroderma pigmentosum,[3] while SLE and

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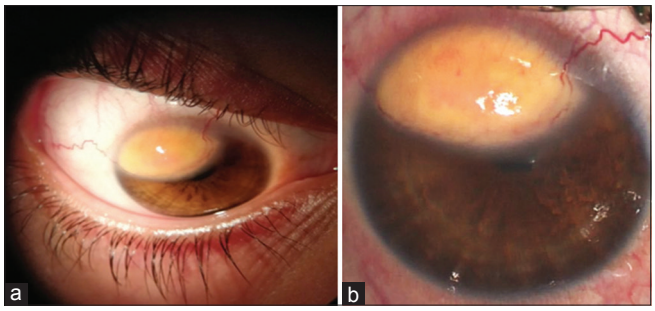

Figure 1: (a) Yellowish orange mass approximately 7*5.5 mm in diameter, extending from 10 to 2'0 clock and vertically reaching the pupillary border. (b) Yellowish appearance with vasculature

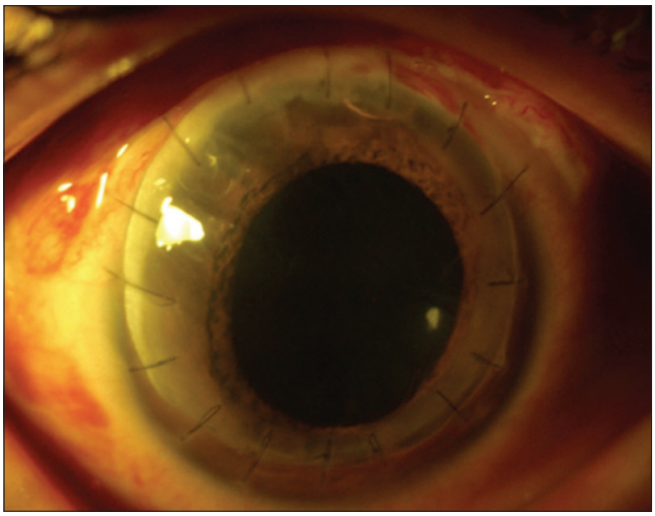

Figure 3: Penetrating Keratoplasty with 16 interrupted nylon sutures

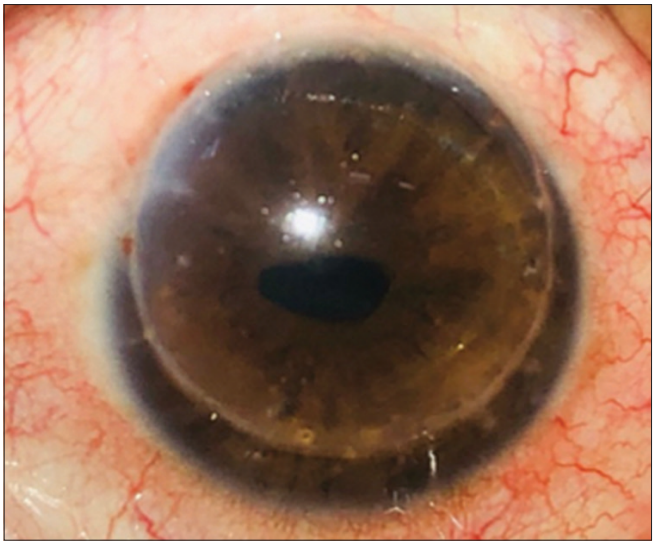

Figure 5: At post-operative 6-month follow-up

multiple dermatofibromas have been shown to be associated with extraocular FH.[4]

The mainstay of diagnosis of FH is histopathology; benign FH typically shows histiocytic and fibroblastic cells in a cartwheel or storiform arrangement.[5] Foam cells,

Figure 2: The Ultrasound Bio microscopy (UBM) of the lesion revealed an elevated mass of 1.54 cm with corneal involvement and no intraocular extension

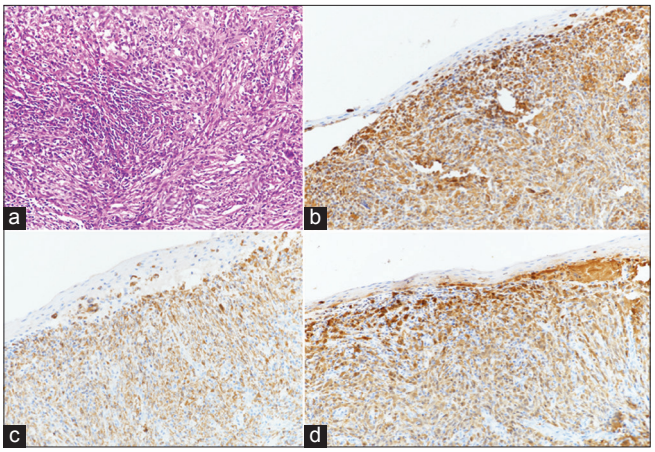

Figure 4: (a) Higher magnification showing fibrohistiocytic cell population with scattered lymphocytes in the background (b) positivity for vimentin in tumor cells (c) Cytoplasmic positivity for CD68 in tumor cells (d) Cytoplasmic positivity for factor XIIIA in tumor cells

macrophages and siderophages may also be seen. Malignant FH shows nuclear atypia, mitotic figures and significant pleomorphism.[3]

CD68, vimentin, smooth muscle actin and focal immunoreactivity for factor XIIIA are some of the available immunostains for FH. They may show mild immunoreactivity for CD34 but lacks it for cytokeratin, S-100, and the melanocytic marker HMB-45.[6] Surgical excisions with tumor free margins are routine for FH, along with corneoscleral/lamellar patch graft.

Few important clinical differential diagnoses of BFH include malignant FH, amelanotic melanoma, lymphoma and juvenile xanthogranuloma.[7] Juvenile xanthogranuloma remains the closest differential even on histopathology. However, presence of inflammatory infiltrate containing eosinophils and S100 positivity as seen in histiocytes of Langerhans cells clinche the diagnosis of xanthogranuloma.[7]

Recurrence is <5% for cutaneous benign FH after local excision. However, malignant FH are prone for local

aggressiveness and recurrences with/without orbital extension and might require enucleation or orbital exenteration.[3]

Role of therapeutic topical steroids has been assessed for the treatment of benign FH with conflicting outcomes and their efficacy remains controversial.[8]

Overall, FH is an uncommon tumor of the eye. Limbus is the commonest site of FH within the anterior segment, while isolated involvement of cornea has never been reported. FH requires prompt diagnosis and skillful excision to prevent catastrophic recurrences. Enucleation or exenteration are final options especially in cases of malignant FH which have shown recurrences with orbital involvement.

Conclusion

FH of the conjunctiva or corneoscleral limbus is a rare tumor. Ours is the first case with an exclusive corneal involvement. Management of these tumors involve complete resection with clean surgical margins to minimize recurrences that may require a more aggressive treatment.

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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Conflicts of interest

There are no conflicts of interest.

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