Medico Research Chronicles

ISSN No. 2394-3971

Case Report

ATYPICAL PRESENTATION OF A DIFFUSE LIMBAL DERMOID

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Submitted on: November 2014
Accepted on: November 2014
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Abstract
Limbal dermoids are benign congenital tumours that contain choristomatous tissue. They remain localized mostly to the superficial layers of the cornea and sclera. Here we report a case of a 20 year old female patient with a deep, diffuse limbal dermoid. However the patient had no squint, diplopia, or proptosis, or amblyopia. Patient had no other systemic association. This peculiar and atypical presentation of a limbal dermoid is rare.
Key words: Dermoid, limbal, orbit, choristoma.

Introduction

Dermoid is described as a solid congenital tumor consisting of mesoblastic tissue covered by ectoderm and invaded by ectodermal derivatives. Limbal dermoids are benign congenital tumors that contain choristomatous tissue. They remain localized mostly to the superficial layers of the cornea and sclera.[1] They appear most frequently at the inferior temporal quadrant of corneal limbus.[2] Its systemic association with colobomas, Goldenhar syndrome or epidermal nevus syndrome is known. Here we report a case of a 20 year old female patient with a deep, diffuse limbal dermoid encroaching on the cornea at one end, and lateral and superior fornix on the other. However the patient had no squint, diplopia, or proptosis, or amblyopia. Patient had no other systemic association. This peculiar clinical entity was investigated and managed with good results.

Case Report

A 20 year old Indian female reported to our OPD in April 2014 with a huge pale mass and swelling on the outer corner of left eye since childhood [Figure 1]. The swelling was painless, gradually progressive, not associated with redness, discharge, diminution of vision, double vision, protrusion or deviation of eye. There was no history of surgery, trauma, or use of medication for the same. On examination, the best corrected visual acuity was 20/20 with projection of rays accurate in left eye. Intraocular pressure was 16 mm Hg and on syringing the nasolacrimal duct was patent. The extraocular movements were full in all gazes with no strabismus [Figure 2, 3].

A diffuse white mass in the bulbar conjunctiva starting from the temporal

limbus reaching the superior fornix, laterally, and extending to the inferior fornix partly was noted. The mass encroached the cornea up to 4mm making a temporal leucomatous corneal opacity, sparing the pupil. The mass was **10mm x12mm x 10mm**, firm in consistency, non tender, immobile, with overlying conjunctival changes. Conjunctiva appeared skin-like with pigmentation, thickening and change of colour. Hair shafts sprouted from the mass at the limbus with incorporation of lid like fragments. Fundus findings were normal. The right eye had a visual acuity of 20/20 with the rest of the anterior and fundus findings unremarkable.

On investigation the hemogram was within normal limits. MRI scan did not show any pressure effects or bony defects [Figure 4a 4b 4c]. We considered dermoid, benign lacrimal gland tumor, dermolipoma as the clinical differential diagnoses. Excision biopsy under local anesthesia was planned [Figure 5, 6]. Debulking surgery was performed and a section was sent for histopathology. Microscopic examination showed stratified squamous epithelium with dense collagen, few sebaceous glands, and multiple pilosebaceous units in the substantia propria [Figure 7]. Patient is on follow up and no recurrence has been noted so far.

Discussion

Dermoid is described as a solid congenital tumor consisting of mesoblastic tissue covered by ectoderm and invaded by ectodermal derivatives. They remain localized mostly to the superficial layers of the cornea and sclera.[1]Limbal dermoids are benign congenital tumors that contain choristomatous tissue. Choristomas are congenital proliferations and occasionally familial. Choristomas are defined as mature tissue elements not normally present at the site of occurrence. Types include limbal dermoid, dermolipoma, ectopic lacrimal gland, and episcleral osseous choristoma. [3] The dermolipoma features dense collagen bundles, abundant adipose tissue.[4] Dermoids appear most frequently at the inferior temporal quadrant of corneal limbus.[2,5] Deep dermoids present in old age with proptosis, or visual or oculomotor disturbance.[6] Various studies have reported morphogical changes in bone in presence of a dermoid.[7,8,9]They suggest that the majority of dermoids in and around the lateral part of the orbit extend into the bone.[10] In our case the lesion was deep diffuse skin-like, arising from lateral corneal limbus and extending temporally but inferior fornix was relatively clear. Also despite being thick and immobile, there was hardly any muscle involvement or distortion of the globe. The peculiar feature was that no bony changes were noted on radiological imaging.

Conclusion

Such dermoids can be a diagnostic dilemma to both clinician and pathologist. It emphasizes the role of preoperative evaluation by both the specialties so as to arrive to an appropriate entity.

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Legends
Figure 1: Mass in the left eye.
Figure 2, 3: Extent of lesion and NO restriction of movement.
Figure 4(a,b,c): MRI shows NO intracranial extension, or bony defects.
Figure 5, 6: Intraoperative excision with debulking.
Figure 7: Histopathology of lesion.

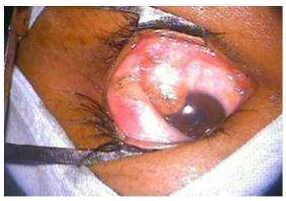
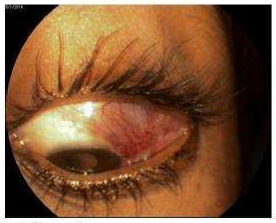


Figure 1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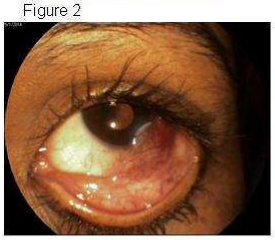
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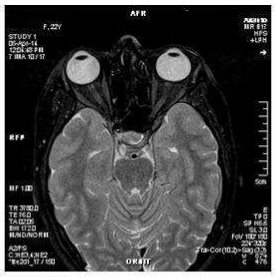
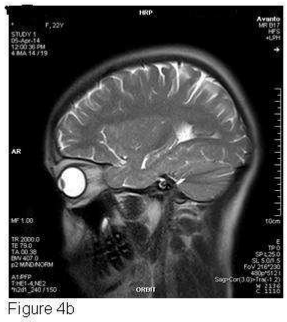


Figure 4a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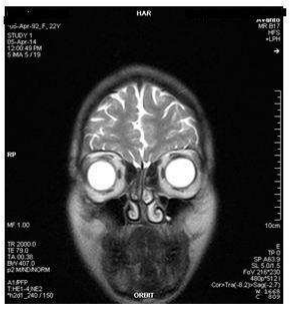
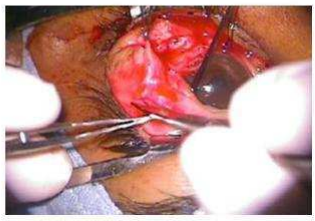


Figure 4c


Figure 5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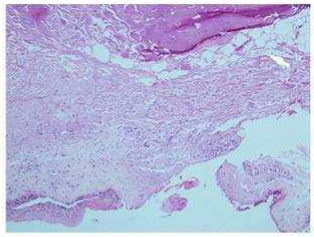
Figure 7