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Case Report:
Corneal Dermoid

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Abstract:
A 20 years old boy presented with left corneal mass. The mass involved entire cornea extending to the sclera. The mass had a skin like surface and protruded outside the palpebral aperture. The eye with the mass was excised .The histopathology report confirmed the diagnosis of corneal dermoid. This late presentation of huge corneal dermoid extending to sclera is first such report in the literature.
Key Words: Cornel dermoid; Keratoplasty; Cosmetic

Introduction:

Dermoids which are localized on the limbus and cornea are very unusual congenital tumors from the group of choristomas.[1] Choristomas are congenital masses of tissues not normally present in the location in which they are found. These lesions are present at birth and they occasionally enlarge, especially at puberty. Most likely, tissue destined to become skin is displaced to the surface of the eye during fetal development.- Dermoids can occur over the cornea, limbus or conjunctiva.[1] They are round or ovoid, yellowish white, solid vascularized and dome like. They can involve the central or entire cornea or form a ring around the circumference of the limbus.[1]

We discuss an interesting case of late presentation of huge central corneal dermoid.

Case Report:

A 20 years old boy presented with a painless left corneal mass. The mass involved the entire cornea with a skin like surface and protruded outside the palpebral fissure. Patient gave history of swelling since birth that had gradually progressed to the present size.

On examination patient had visual acuity of no perception of light in the left eye and 6/6 in the right eye. Examination with diffuse illumination showed huge central corneal mass which was highly vascularized, measuring around 11×15mm in size involving whole of cornea and abutting the sclera. Mass was non tender, soft, cystic, fixed to the cornea. (Figures 1,2,3,4)

Ultrasound B-scan showed intact globe and hypoechoeic (cystic) nature of the swelling (Figure 5,6). UBM showed intact cornea with a well formed anterior chamber (Figure7). CT scan was not done for this patient. Systemic examination done, revealed no positive findings.

Surgical excision was planned. Entire corneal mass with the eye was excised and sent for histopathology. The histopathology report confirmed the diagnosis of corneal lipodermoid. (Figure 8,9)

Discussion:

Dermoids are solid benign congenital tumors that frequently arise at the inferotemporal corneoscleral junction.[2] They are classified as choristomas because they contain cellular elements not normally present in the location, ectodermal derivatives such as hair follicles, as well as sebaceous and sweat glands embedded in connective tissue and covered by squamous epithelium.[2]

They can also contain smooth and skeletal muscle, nerves, blood vessels, bone, cartilage and teeth. In the eye they most often present as yellowish white, solid, vascularized, elevated nodules straddling the corneal limbus. They vary greatly in size ranging from 2-15mm in diameter.

Corneal dermoids occur more commonly as single lesions but may be multiple and they may be unilateral or bilateral, the former being the most common. Dermoids can be central and often appear to have satellite lesions.

Approximately 30% of individuals with Goldenhar's syndrome have epibulbar dermoids. They most commonly occur unilaterally and at the inferotemporal limbus. Other systemic syndromes associated with epibulbar dermoids include Treacher Collins syndrome, incontinentia pigmenti, encephalocraniocutaneous lipomatosis, linear sebaceous nevus syndrome and cri du chat syndrome.[1] In our case we did not find any systemic associations. The different types of dermoid choristomas have been classified according to the extent of involvement. (Table1)[2]

Figures1,2,3,4: Clinical photographs of the patient with huge central corneal dermoid in the left eye

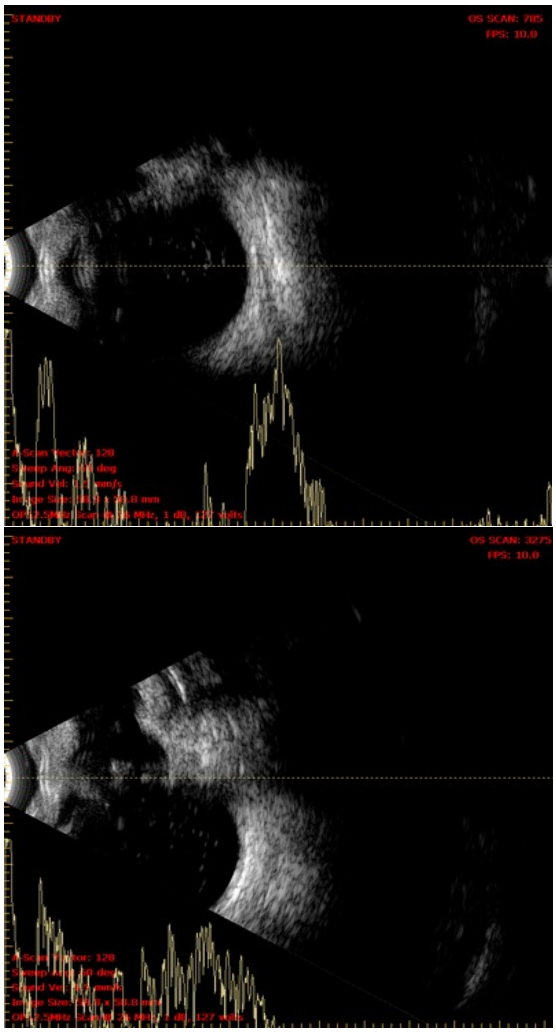

Figure 5,6: Ultrasound B scan showing intact globe with hypoechoeic lesion on the cornea

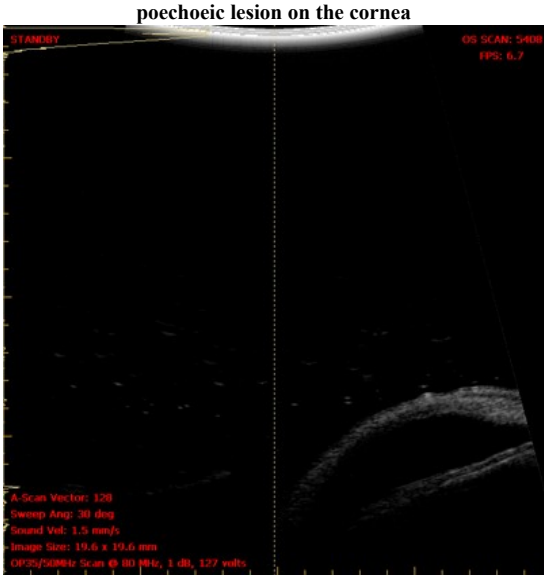

Figure7: UBM image showing a well formed anterior chamber in the left eye

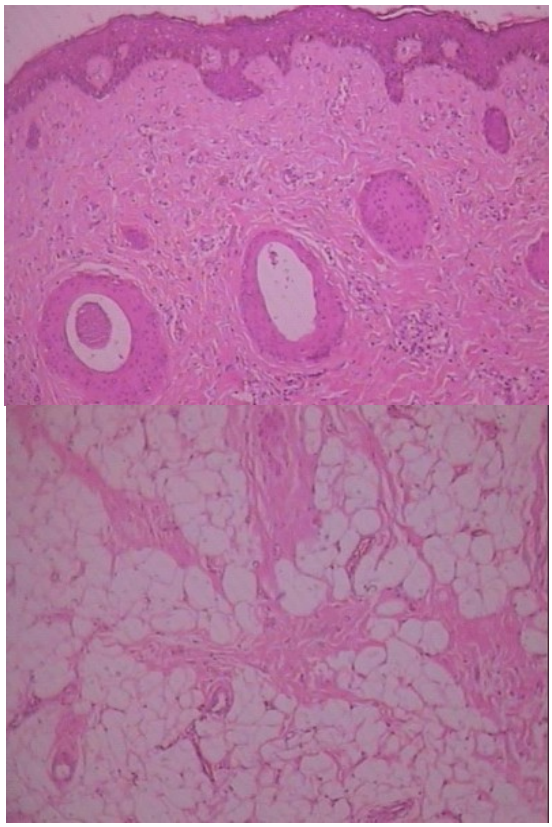

Figure 8,9: Histopathology showing hair follicles, squamous epithelium, adipose tissue (H & E stain)

Table 1: Grading Dermoids		
Grade1 (Limbal or epibulbar)	**Grade2**	**Grade3**
• Most frequent type • Small(5mm in diameter) • Single • Inferotemporal limbus • It may enlarge at puberty • Superficial • One third cases associated with Goldenhars's syndrome	• Much larger • Covers part or entire central corneal surface • Variable depth of stromal extension • Does not involve descemets membrane or the corneal endothelium	• Most severe • Very rare • Entire anterior segment is involved • Associated abnormalities such as microphthalmos, posterior segment abnormalities

In our case dermoid was of grade 2 variety which is rare unlike the grade1 variety. Dermoids that involve or distort the central cornea can decrease the quality of visual image and create amblyopia. Various earlier reports on corneal dermoids are summarized in Table 2.

Late presentation of the corneal dermoid(20 years) as in our case is probably the first such report in literature. Stergiopoulos P and co-workers studied 46 patients with different types of dermoid. The surgeries done for corneal dermoids included lamellar sclerokeratectomy, lamellar keratoplasty, corneoscleroplasty, and lamellar removal with autologous episcleral transplant.[3]

Penetrating keratoplasty can be performed for central dermoids if they are 7mm or less in diameter. Large central dermoids require a two staged procedure. First the tumor is excised and a-

Table 2: Case Reports			
Author	**Age of presentation**	**Description of dermoid**	**Treatment**
Leung AT et al[4]	14 days	Pedunculated corneal dermoid	Excision of dermoid, awaiting rotational autokeratoplasty
Mohammad AE et al[5]	25 days	Huge corneal dermoid extending to sclera	Eye with mass was excised
Golubovic S et al[6]	2 year	Large corneal dermoid	Penetrating keratoplasty
Shields JA,et al[7]	1 year	Central corneal dermoid	Twice penetrating keratoplasty was done
Zaidman GW et al[8]	1 month	Protruberant corneal dermoid	Lamellar keratectomy followed by penetrating keratoplasty

.large lamellar graft is placed in the bed, once that is healed a smaller central keratoplasty is done. A good but not perfect cosmetic result can be achieved with a lamellar graft. Operating for cosmetic reasons can be unsuccessful if unsightly scarring occurs.[2] We concentrated more at improving the cosmetic outlook of the patient.

Though penetrating keratoplasty was thought of in our case, it could not be done due to huge size of the dermoid extending to the sclera. Enucleation was done in our case, followed by fitting prosthesis giving a very good cosmetic result (Figure 10, 11)

Figure 10,11: Clinical photograph of the patient before and after fitting custom made prosthesis in the left eye

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