

Limbal Dermoid on Clinical Presentation, but on Histology was Epidermal Cyst

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Abstract

An 8 yr old female child presented with complaints of swelling in the left eye noticed by her parents before 6 months. On examination, best corrected visual acuities in both eyes were 6/6, right eye normal, left eye, a swelling 3 mm x 4 mm, pinkish red in colour in the inferior temporal area involving the limbus. Excision biopsy was done. Histopathology report was suggestive of epidermal cyst.

Introduction

Limbal dermoids are benign congenital tumours that contain choristomatous tissue (tissue not found normally at that site). They appear most frequently at the inferior temporal quadrant of the corneal limbus. However, they occasionally may present entirely within the cornea or may be confined to the conjunctiva. They may contain a variety of histological aberrant tissues, including epidermal appendages, connective tissue, skin, fat, sweat gland, lacrimal gland, muscle, teeth, cartilage, bone, vascular structures, and neurological tissue including the brain. Malignant degeneration is extremely rare.

The most common system for classifying dermoids is based on the location of the lesion and separates the lesions into 3 broad categories. The most common dermoid is the limbal dermoid, in which the tumour straddles the limbus. These are usually superficial lesions but may involve deeper ocular structures. The second type involves only the superficial cornea, sparing the limbus, the Descemet membrane, and the endothelium. The third type of dermoid involves the entire

anterior segment, replacing the cornea with a dermolipoma that may involve the iris, the ciliary body, and the lens.

Case Report

An 8 yr old female child, came with history of swelling in the left eye since 6 months. There was no history of injury or diminution of vision. No history of any systemic illness. No history of any similar complaints in other siblings. No history of any similar complaints in the past. Birth history was normal.

On examination, best corrected visual acuities in both eyes were 6/6, right eye was normal. Left eye lids were normal. A solitary pinkish red mass measuring 3 mm x 4 mm with well defined margins occupying the inferotemporal limbus was seen, having an irregular surface, with multiple hair follicles on its surface. Mass was firm in consistency. Cornea was clear and IOP and extraocular movements were normal. Excision biopsy was done. Histopathological report confirmed epidermal cyst. Complete blood investigations showed Hb- 13 gm%, CBC- 8500 cu/mm, ESR- 18 mm/hr and urine routine was normal. X-ray spine, CT scan and USG Bscan were normal and did not show any anomalies. Ultrabiomicroscopy did not show invasion into the deeper tissues. ENT check up was normal. Systemic examination was within normal limits (Fig. 1).

Pathophysiology

Several theories have been proposed to explain the development of limbal dermoids. One theory suggests an early developmental error resulting in metaplastic transformation

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of the mesoblast between the rim of the optic nerve and the surface ectoderm. Another proposed mechanism is sequestration of the pluripotent cells during embryonic development of the surrounding ocular structures. The exact pathogenesis probably varies from case to case.

Limbal dermoids generally are not inherited, although some exceptions have been reported. Familial presentation of limbal dermoids in association with systemic disorders, such as Goldenhar syndrome, is well recognized and follows a multifactorial pattern of inheritance. Two rare forms of epibulbar dermoid (the annular limbal form and corneal dystrophy form) presenting in multiple family members also have been reported.

Frequency

Internationally: The estimated worldwide incidence of limbal dermoids is 1 per 10,000 to 3 per 10,000. In a study at the Armed Forces Institute of Pathology, 7.5% of epibulbar lesions examined were choristomas. They found that 52% of the epibulbar choristomas were located in the bulbar conjunctiva, 29% at the limbus, 6% on the cornea, 4% at the caruncle, and 2.5% in the conjunctival fornix or the palpebral conjunctiva. In a study of epibulbar choristomas, in patients with Goldenhar's syndrome, 14% were nasal, 86% temporal, 16% superior, and 84% inferior.

Mortality/Morbidity

Visual morbidity may result from encroachment of the lesion into the visual axis, development of astigmatism, or formation of a lipid infiltration of the cornea, which obstructs the visual axis.

Large limbal dermoids can be cosmetically disfiguring.

Staphyloma formation adjacent to

dermoids has been reported and may be associated with spontaneous perforation of the cornea or sclera.

Race

No racial predisposition exists.

Sex

Limbal dermoids occur with equal frequency in males and in females.

Age

Limbal dermoids are present at birth but may not be recognized until the first or second decade of life. They may appear to enlarge as the body matures.

In a study conducted at the Armed Forces Institute of Pathology, 36% of epibulbar lesions removed in the first decade of life were choristomas. They constituted about 23% of the epibulbar lesions removed in the second decade of life, 7.2% of the lesions removed in the third decade, and 0.9% in the fourth decade.

In another study conducted at the Wilmer Ophthalmologic Institute, epibulbar choristomas constituted 33% of epibulbar lesions removed before age 16 years and 2.2% of the epibulbar lesions removed after age 16 years.

History

Patients present with decreased vision or poor vision, foreign body sensation, cosmetic disfigurement, or an enlarging ocular mass.

Most patients present before age 16 years.

Physical

Mostly 85% epibulbar dermoids are located at the inferior temporal limbus.

Rarely, they may only affect the cornea or the bulbar conjunctiva.

Epibulbar dermoids have a dome shape, and the surface may appear keratinized.

Hair follicles and cilia may be visible.

The dermoid appears fleshy and may have fine superficial vascularization.

Associated ocular abnormalities include colobomata of the eyelids, Duane retraction syndrome and other ocular motility disorders, lacrimal anomalies, scleral and corneal staphylomata, aniridia, and microphthalmia.

Associated systemic abnormalities include preauricular appendages and auricular fistulae (in combination with limbal dermoids constituting the Goldenhar syndrome). Other abnormalities include hemifacial microsomia, microtia, and vertebral anomalies.

Causes

Most cases of limbal dermoids are sporadic and not related to any known toxic exposure or mechanical irritant.

Instances are reported of epibulbar dermoids being related to maternal ingestion of teratogenic agents during the first trimester of development.

Differential Diagnosis

- Epidermal Cyst
- Juvenile Xanthogranuloma
- Peters' Anomaly
- Foreign body granuloma
- Sclerocornea
- Staphyloma
- Corneal scar (from trauma or infection)
- Haemangioma
- Atypical pterygium

An estimated 30% of patients with dermoid choristomas have associated systemic abnormalities, including Goldenhar syndrome, nevus flammeus, and neurofibromatosis.

Other systemic disorders reported include cardiovascular abnormalities, vertebral

anomalies, and auricular defects.

Work up

Lab Studies

The diagnosis of a limbal dermoid requires a direct clinical examination. Specific laboratory studies are generally not necessary.

Imaging Studies

MRI

- Some dermoids may appear to extend into the conjunctival fornix or lateral canthus. These lesions may contain connective tissue that entangles with the orbital fat and muscle tissue belonging to the extraocular muscles.
- Radiologic imaging with an MRI can be useful in identifying such lesions, especially if surgical management is being considered.

UBM

To determine the depth of the corneal tissue involvement.

Procedures

Biopsy is not necessary except in rare instances when the diagnosis is doubtful.

Histologic Findings

Limbal dermoids contain choristomatous tissue, including epidermal appendages, adipose and lacrimal gland tissue, smooth and striated muscle, cartilage, brain, teeth, and bone. Lymphoid nodules and vascular elements also have been reported. The surface of the dermoids consists of corneal or conjunctival epithelium. The lesion may be cystic or solid.

Treatment

Surgical Care

Treatment of limbal dermoids may consist

of periodic removal of irritating cilia, topical lubrication to prevent foreign body sensation, or excision of the lesion if it is causing significant cosmetic disfigurement or interfering with vision.

Surgical treatment should be instituted only when the risk of subsequent scar formation or surgical complications are outweighed by the likelihood of improving the patient's vision or cosmetic appearance.

A superficial sclerokeratectomy, cutting flush with the surface of the globe, is the procedure of choice for removal of the dermoid. Excised tissue always should be sent to the pathologist for examination.

Attempts at complete removal are unnecessary. The lesion may extend into the deeper structures of the eye and the risk of perforation increases if attempts are made to remove the lesion completely.

The exposed sclera should be covered by relaxing the adjacent conjunctiva and sewing it into the scleral defect. If a deep excision is necessary, then a lamellar keratoplasty can be performed to reinforce the site of excision.

Consultations

Obtaining a thorough family and medical history helps determine whether further consultation is necessary.

In some cases, referral to a paediatrician with specialization in genetics is appropriate.

Prognosis

Prognosis is generally favourable.

Medical/Legal Pitfalls

A young patient presenting with a limbal dermoid may be at a risk of developing amblyopia. Likewise, a patient presenting with a limbal dermoid already may have developed amblyopia. This issue must be discussed clearly with the patient and/or the patient's family. A plan for treating the amblyopia and an explanation for the possible reduction in vision should be given to the family and documented in the medical record.

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