

Limbal Dermoid with Melanosis and Hypertrichosis (Unusual Combination)

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Abstract

Seven year old male child presented with complaints of decreased vision and swelling in left eye present since birth with pigmentation of conjunctiva and skin with excessive hair growth on the lids (Hypertrichosis).

Introduction

Dermoids at limbus are congenital benign tumours; most frequent site is Infero-temporal.¹ They contain variety of histological aberrant tissues including epidermal appendages, connective tissue, skin, fat, sweat glands, lacrimal gland, cartilage, bone, teeth, vascular and neurological tissue usually the tumour is superficial straddles limbus. Second type involves only the cornea sparing Descemet's membrane and limbus. Third type involves anterior segment including iris, ciliary body and lens along with cornea melanosis is usually associated with naevus of Ota and Sturge Weber's syndrome, usually hereditary transmission. Hypertrichosis is dominant hereditary transmission.⁸ Hypertrichosis is dominant hereditary condition and may occur alone or with Cornelia de Lange syndrome.

Case Report

Seven year old male child presented with complaints of swelling in left eye present since birth. Pigmentation of conjunctiva and skin (Melanosis) and excessive hair growth (Hypertrichosis) of lids. The skin lesions were present since birth and not increasing with age. The swelling in left eye, was present since birth and increasing with age with reduction in vision. There was no family history of

Hypertrichosis or dermoid, nor hyperpigmentation (Melanosis). Birth history and mile stones were normal. No maternal illness or medication during pregnancy was reported. 3 older siblings were normal. On examination the skin of both eye lids and eye brows showed hypertrichosis with bluish black hyperpigmented plaques (Melanocytic naevus) visual activity in right eye was 6/6 and left eye was finger counting 1 meter. External and slit lamp examination showed melanosis of the conjunctiva in both eyes, with dermoid at limbus from 3.50 to 5.30 O'clock inferotemporally 3 mm x 4 mm in size superotemporal lipodermoid about 3 mm x 4 mm was seen at 12.30 to 2.30 O'clock position along with melanosis. Anterior chamber and fundus examination were within normal limits Extra ocular movements were normal in right eye but abducting was minimally restricted in left eye intraocular pressure was digitally normal. Systemic examination did not reveal any abnormality. Audiogram and ENT check up were normal. Paediatric opinion was also taken to confirm absence of features of Goldenhar syndrome, mandibulofacial dysostosis, hemifacial microsomia, Cornelia de Lange syndrome. Dental opinion was taken to confirm normal dentition. Blood investigations showed Hb 9.6 mg% total WBC count 8200/cm. Polymorphs 56% Lymphocytes 44% ESR was 22 mm/1st hour. BUN 11mg%, NA140, K.3.6, Cl. 82, Creatinine 0.5 mg%, Random blood sugar 174 mg%, X-ray spine did not show any abnormality, CT scan of skull was normal, B scan confirmed that dermoid was superficial and not extending to iris of ciliary body and angle of anterior chamber. Excision biopsy of the lesions confirmed diagnosis of dermoid.

Pathophysiology

Ocular melanosis noted alone or with Sturge Weber syndrome,² Naevus of Ota⁸ is seen since birth,

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Fig. 1 : Patient having right eye Hypertrichosis and left eye melanosis of conjunctiva with lipodermoid at limbus.



Fig. 2 : Right eye showing lower lids Hypertrichosis

unilateral involving skin supplied by 1st and 2nd division of 5th (Trigminal) cranial nerve, rarely 3^d division. It may also involve conjunctiva, iris, choroid, sclera and optic nerve but no hair growth was present with it.

Hypertrichosis, may show persistence of foetal hair all over body as dominant hereditary condition. Failure of dentition may be associated with it. Duplication supercilli shows two rows of eyebrows separated by a clear area. Sometimes it may be only temporal it may be associated with Cornelia de Lange syndrome showing dwarfism and multiple anomaly, Polytrichia, 2 rows (districhiasis); 3 rows (tristichiasis); 4 rows (Tetrachiasis). Accessory rows of lashes run along inner part of intermarginal strip. Kahnt considered it as heterotypical developmental anomaly. Von Szily suggested atavistic implication, phenomena



Fig. 3 : Right eye showing lower lids Hypertrichosis



Fig. 4 : Left eye showing lipodermoid at about 3 mm x 4 mm at 12 :30 to 2:30 O'clock position. Dermoid at limbus from 3:50 to 5:30 O'clock

common in animals. Meibobion glands are modified sebaceous glands which are probably associated with lashes in lower animals but have disappeared in man. Ectopic cilia where cluster of lashes grow outward from skin of upper lid.⁸

Goldenhar Syndrome is recognized in 1st and 2^d decade because though dermoid is present at birth, it enlarges as child grows. Early developmental error resulting in metaplastic transformation of the mesoblast between rim of the optic nerve and surface ectoderm. Another theory is sequestration of the pluripotent cells during embryonic development. Hemifacial microsomia and mandibulofacial dysostosis result with first and second brachial arch involvement.⁴

Patient with limbal dermoid presents with complaints of cosmetic disfigurement, decreased vision and foreign body sensation. 85% dermoids are located at inferotemporal limbus.⁴ They have a dome shaped keratinized surface with visible hairs, it appears fleshy with fine superficial vascularisation. May be associated

with coloboma of eye lids, aniridia, microphthalmia, Staphyloma, lacrimal anomaly and Duane's retraction syndrome. Associated systemic anomaly include extra auricular appendages, pretagal blind ended fistula, vertebral column anomalies like spina bifida, hemivertebrae, butterfly vertebrae etc. This was collected and classified as Goldenhar syndrome in 1952. Georlin in 1963 Introduced term oculo auriculovertebral (OAV) syndrome, Mandibulofacial dysostosis may present with malar hypoplasia involving, both jaws (Maxillary and mandibular bones) wide mouth, cleft lip and cleft palate. Absence or malformations of ear (Anotia or microtia) middle or inner ear anomaly resulting in sensorineural hearing loss, dermoid, coloboma of lids, microphthalmia, blepharophimosis [narrow palpebral fissure] strabismus may occur. Cardiac, pulmonary renal or gastrointestinal abnormalities may be present. 15% may be mentally retarded.⁷

Histology

Limbal dermoids contain choristomatous tissue, including epidermal appendages like hairs, smooth and striated muscle, cartilage, bone, teeth, brain, adipose tissue, lacrimal gland, lymphoid nodule and vascular elements. Lesion may be cystic or solid.

Treatment

Surgical excision of tumour by superficial sclerokeratectomy cutting flush with surface of globe, is preferable; if lesion is involving deeper structure.⁵ Complete removal is likely to cause perforation of globe and affect vision. Risk of surgical complication should be compared with likelihood of improving vision or cosmetic appearance. Prognosis is good.⁶

Medical pitfalls of limbal dermoid are risk of developing amblyopia. It must be explained to patient and the relatives and documented in the medical records.

Discussion

The child had limbal dermoid without any features of Goldenhar syndrome, mandibulofacial dysostosis. Child had melanosis but no features of Sturge Weber

syndrome. Child had bilateral melanosis of the conjunctiva of pigment along 1st, 2nd and rarely, 3rd division of trigeminal nerve, as seen in naevus of Ota, child had hypertrichosis resembling duplication supercillii (2 Rows of eye brows) Failure of dentition is usually seen with hypertrichosis but in this child dentition was normal. There were no features of Cornelia de Lange syndrome which is also associated with hypertrichosis. Thus the child presented with unique features of 3 different conditions, without any associated features which are seen with the 3 conditions and hence we thought it will be interesting to report this case.

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