Conjunctival Dermolipoma

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INTRODUCTION

Choristomas are congenital lesions.(1) Heterotopia or choristomas represent mature and normal tissue in abnormal sites. They include pancreatic tissue in the wall of the stomach or small intestine; adrenal cells found in the kidney, lungs, ovaries or elsewhere.(2) Ocular choristomas include limbal dermoid, lipodermoid, ectopic lacrimal gland, and episcleral osseous choristoma of which the former two are the most common orbital and epibulbar tumors in children. They commonly affect the cornea, limbus, and bulbar conjunctiva. Lipodermoids are common near the superior temporal quadrant of the globe and do not tend to affect the peripheral cornea. Histologically, a lipodermoid is adipose tissue covered by connective tissue while a dermoid is comprised of collagen connective tissue covered by epidermal epithelium.(1)

Congenital epibulbar dermolipoma are composed of adipose tissue covered by connective tissue. They are usually located superotemporally and generally do not involve the peripheral cornea. If the dermoid or dermolipoma is accompanied by other systemic conditions or ocular anomalies in young children, Goldenhar syndrome needs to be ruled out. This paper reports a unilateral conjunctival dermolipoma in a healthy male child.

PRESENTATION OF CASE

A 9 years old child presented with a painless swelling in the lateral aspect of the left eye since birth. He had no visual disturbances. He had no documented history of infection, trauma, or surgery to the orbit or adjacent skin. On examination, the non-mobile, non-tender lesion was located in the temporal aspect of the left eye with fine hair on its surface. (Clinical photograph 1 a, b) It could not be repositioned into the orbit. There was no associated feature of surrounding inflammation. The other eye did not show any finding. The patient did not present with any other congenital abnormalities. There were no pre-auricular appendages, facial or palatal clefts, or postural abnormalities. The patient was free of any systemic symptoms.

The child had undergone CT scan and MRI for the orbital lesion. On CT scan, there was a soft tissue density lesion in the supero-temporal quadrant of the left eye with fat (Fig. 1a) and focal calcification, not continuous with the epibulbar fat. (Fig 1b). The child was further taken up for Gadolinium enhanced MRI, which showed a non-enhancing fat intensity lesion in the outer canthus of the left eye continuous with the conjunctiva. (Fig. 2).

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DIFFERENTIAL DIAGNOSES

1. Episcleral osseous choristoma.
2. Subconjunctival fat prolapse.
3. Limbal dermoid.

DISCUSSION OF MANAGEMENT

The child was taken up for surgical resection of the lesion and histopathology revealed squamous lining of skin with hair follicles along with collagenous and fatty tissue along with sebaceous glands, thus confirming the radiological diagnosis of conjunctival dermo-lipoma. There was no pathological documentation of mature compact bone, as typical for osseous choristoma.

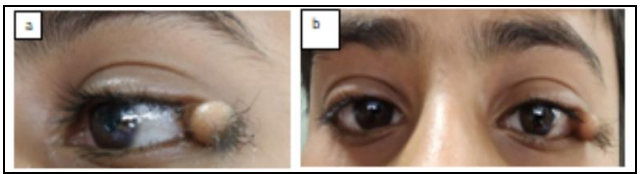

Figure 1a, b. A Swelling with Superficial Fine Hair Located in the Supero-Temporal aspect of the Left Eye without Surrounding Inflammation

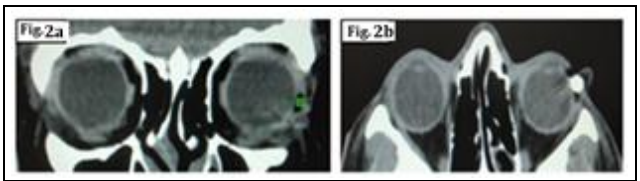

Figure 2a. Coronal Reformatted CT Scan of the Orbits Reveals a Soft Tissue Density Lesion with Fat in the Supero-Temporal Quadrant of Left Eye. Figure 2b. Axial CT Scan Shows a Focal Calcification in the Lesion which is not Continuous with the Epibulbar Fat

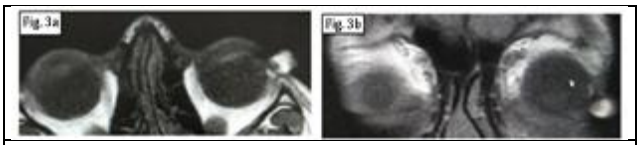

Figure 3a, b. Axial and Coronal T1 WI Images of the Orbits Reveals a Hyperintense Lesion with a Hypointense Focus (Calcification in the Outer Canthus of Left Eye)

DISCUSSION

Conjunctival dermolipoma usually occur in the lateral canthus beneath the temporal or superotemporal bulbar conjunctiva. Dermolipoma is a congenital lesion occurring early in life with a female preponderance. It usually presents as a unilateral soft or firm pinkish-white or yellow mass with a straight or slightly concave anterior margin. Fine hairs are often seen on the surface. It is not freely movable, cannot be repositioned into the orbit and is not affected by retropulsion of the globe. Dermolipoma is a congenital solid choristoma that is derived from an ectopic sequestration of ectoderm to the conjunctiva.[3] Dermoid or a lipodermoid may be associated with Goldenhar syndrome that appears as oculoauriculovertebral dysplasia. A diagnosis of GS includes the following clinical manifestations: auricular agenesia, hemifacial microsomia, oblique facial cleft, epibulbar dermoids and/or upper eyelid colobomas, and vertebral anomalies.[4]

Histologically, dermolipoma is characterized by the stratified squamous epithelium and the subepithelial stroma contains various amounts of collagenous connective tissue and adipose tissue. The stroma may also contain cartilage and glandular acini.[3]

Surgical resection of dermolipoma should be done with limited debulking of the tumor because attempts at a wide or complete surgical excision may result in complications such as ptosis, diplopia, and lacrimal gland injury, which are unacceptable for an essentially cosmetic problem.[3] In addition, proximity of the dermolipoma to the lacrimal ductules, the lateral rectus and upper lid structures makes extensive surgical excision dangerous.[5]

Osseous choristomas are solid nodules, composed of mature, compact bone together with pilosebaceous units and hair follicles. They are the rarest forms of ocular choristomas and are found at the superior temporal region of the episclera. They are generally adherent to extraocular muscle sheath, overlying conjunctiva or underlying sclera.[2]

Subconjunctival fat prolapse occurs mainly in elderly obese men. It is due to herniation of intraconal fat due to an age-related weakening of the Tenon capsule (fascia bulbi) and the intermuscular septum and manifests typically as a unilateral or bilateral soft yellowish mass. It has a convex anterior margin and superficial fine blood vessels. It can be repositioned into the orbit and becomes more prominent by retropulsion of the globe.[3]

Limbal dermoid is is usually seen as a well-circumscribed firm solitary mass most commonly located inferotemporally but may arise also in the nasal or superior portions. Histologically, it is a simple choristoma, consisting of fibrous tissue lined by conjunctival epithelium along with hair follicles and sebaceous glands.[3]

The CT and MR imaging findings of subconjunctival fat prolapse and dermolipoma are quite different. The most important imaging feature distinguishing these 2 conditions is the continuity of the fatty mass with the intraconal fat. Although both lesions were located at the temporal or superotemporal aspect of the epibulbar region, they are invariably continuous with the intraconal fat in case of subconjunctival fat prolapse which is not so in case of dermolipoma.[3] Except for the different predilection sites of occurrence, it may be difficult to differentiate dermolipoma and limbal dermoid radiologically. Distinction between osseous choristoma, limbal dermoid and conjunctival dermolipoma essentially rests on the histological findings.

CONCLUSIONS

Imaging plays a pivotal role in distinguishing epibulbar masses like dermolipoma from other pathologies of the conjunctiva and eyelids, such as subconjunctival fat prolapse, dermoids, osseous episcleral choristomas, prolapsed lacrimal gland or lacrimal gland neoplasms, lymphomas, and lipomatous tumors, though histopathologic confirmation may ultimately be required to make a diagnosis. Radiologically, fat-containing

epibulbar masses that should be included in the differential diagnosis are limbal dermoid and lipoma.

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