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ISSN No: 2319-5886

International Journal of Medical Research & Health Sciences, 2017, 6(4): 97-101

Management of Lipodermoid in Hemifacial Microsomia of OAV Dysplasia: Rare Case Report

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ABSTRACT

Congenital hemifacial microsomia of OAV (oculo-auriculo-vertebral) dysplasia along with birth mark on cheek and forehead is a rare occurrence. In this case, right side of face is more affected than left side. Ocular lipodermoid is benign, slow growing tumour presented in lateral canthus of right eye, creates serious complications disfigurement of eye which is managed by cosmetic surgical approach.

Keywords: OAV dysplasia, preauricular fistula

INTRODUCTION

A 9-year old female child presented mainly for slow growing ocular tumour in right eye. Parents notice increased swelling at the age of 3 years and thought to seek opinion for it. Swelling was progressive, painless involving inferotemporal ocular surface, which gives discomfort and restricted lateral movement of the eye. No other family members found to be affected like her.

On examination, her head position tilted slightly forward and upward. On the forehead, there is birth mark running from lateral side towards medial one third of eye brow in oblige line 4.2 cm long (Figure 1).

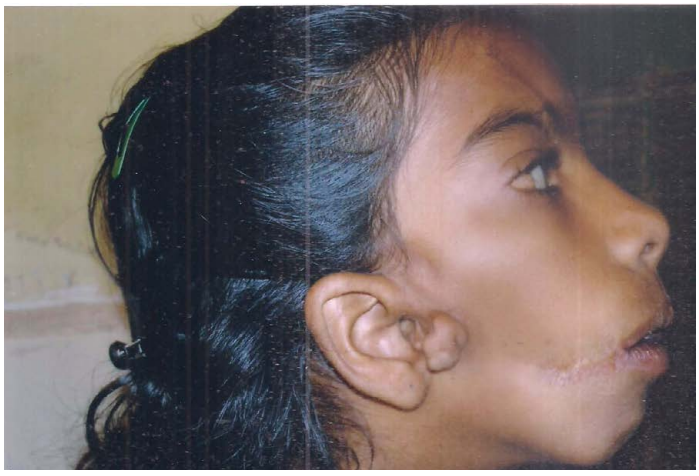

Figure 1 Initial examination of head

Ocular examination

Except ocular swelling temporally her visual acuity in both eye-within normal limits, fundus examination unremarkable, anterior segment of both eye-within normal limits.

White yellow colour swelling present in infer temporal quadrant 5 mm away from the limbos, conjunctive over the swelling is yellowish brown colour, this fleshy mass having flat drone shaped appearance, on palpation it is soft mass and conjunctiva moves over it (Figure 2).

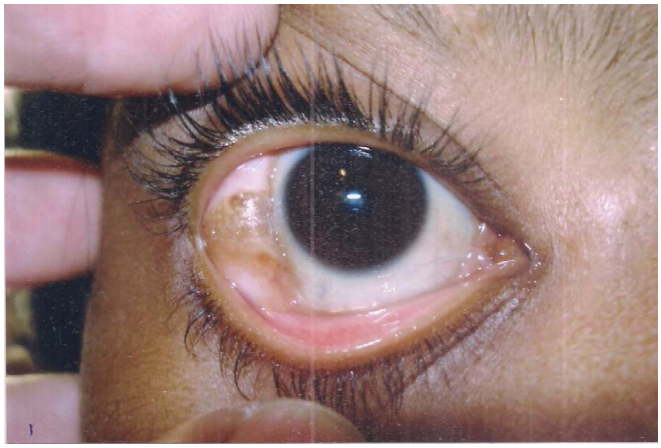

Figure 2 Initial examination of eye

Consistency no compressible on reducible non-pulsatile. tumour mass extending up to lateral orbital margin, medical, border rounded smooth, lower border of conjunctive in fornix superiorly margin of tumour is merged in the epibulbar conjunctiva.

Clinically diagnosis confirmed as epibulbar dermoid of large size disfiguring the ocular surface. Apart from this, child had ear deformity, externally atresia of external ear, preauricular big appendages along with pretragal fistulous opening, atresia of external auditory opening with presence of preauricular appendages in the auditory canal with complete deafness.

Face on right side show facial asymmetry, under developed muscles hemifacial, microstomia. On cheek, a birth mark is present from angle of mouth to middle portion of cheek horizontal curved line 7 cm long parallel to lower border of mandible.

Mouth remains open, complete closer not possible to her angle of mouth slightly pulled on right side towards, direction right ear, micromandible present.

Open mouth, horizontal birth mark from angle of mouth to middle of cheek and preauricular appendages are present in straight line is rarest finding observed.

Neck is short, no difficulty in movement of neck. Cervical vertebral anomalies present otherwise no other vertebral anomalies observed.

The child presented with epibulbar dermoid, preauricular appendages with horizontal birth mark on cheek open mouth, hypoplasia of face muscles, hemifacial microsomia micromandible and vertebral anomalies this combination constitute Goldenhar syndrome. No other systemic abnormalities found in this child.

Lab investigations

Clinical examination of ocular mass clinically gives confirmation of diagnosis of epibulbar dermoid. Blood haemogram along with kidney function test are within normal limits. X-ray shows two upper cervical vertebrae fused posteriorly. No other deformity.

CT-Scan of right eye orbit shows ill-defined fat attenuation lesions (average attenuation -50 to -60 HU) of approximate size 1.5 × 1.1 × 0.4 cm involving temporal side of conjunctival surface of right eye globe surrounded by moderately enhancing hyperdense soft tissue attenuation rim e/o mucosal thickening seen in right frontal and left maxillary sinus without any collection. Right maxillary sinus, left frontal sinus and both ethmoid and sphenoid sinuses are normal, no e/o mass or polyps on left eye globe extraocular muscles, optic nerve on both side are normal visualized skull vault is normal. Opinion reveals ill-defined fat attenuation lesion involving the temporal side of conjunctival surface of right eye globe surrounded by hyperdense soft tissue attenuation rim imaging features favour the diagnosis of conjunctival lipodermoid.

MRI shows extension of lipodermoid into conjunctival fornix, e/o connective tissues and orbital fat, no attachment to extraocular muscles.

Radiological CT-scan, MRI of lipodermoid are valuable in confirmation of diagnosis and also helps to guide intraorbital extension and in further management after all investigations possibility of benign lesion of lipodermoid diagnosed. which is differentiated from other non-neoplastic, non-inflammatory epibulbar cysts orbital dermoid cysts, foreign body granulomas, staphylomas.

Management depends upon severity of lesions. Cosmetics approach of surgical excision of growth to relieve symptoms created by tumour mass and for confirmation of diagnosis maximum. Tumour mass is excised without spilling its content and care was taken to protect the involvement of extra ocular muscles. In this case to the exposed sclera, mitomycin C 0.02% was applied for one minute to prevent recurrence of normal tissue and the exposed sclera was covered by lose adjacent conjunctiva with sutures.

Histopathological report of excised specimen lesion on outer canthus of right eye shows grossly single globular solid tissue 2.5 × 1.5 cm with pinkish cut surface (Figure 3).

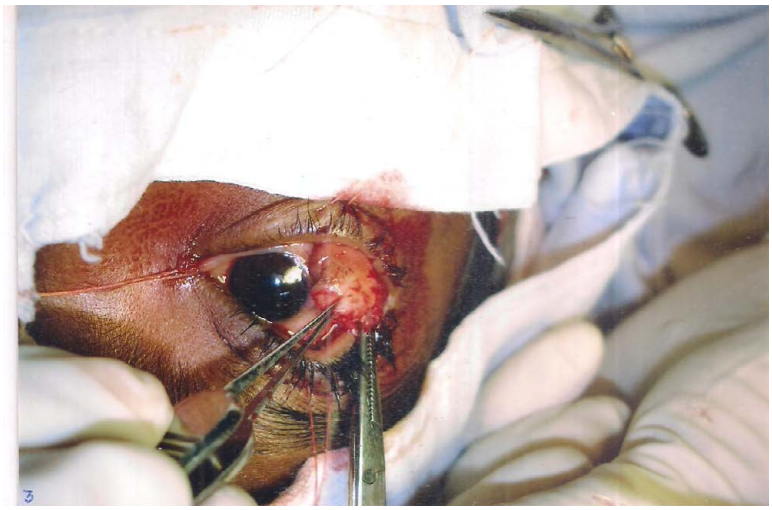

Figure 3 Surgical removal of the adipose tissue

Section shows stratified squamous lining with undertaking skin appendages placed haphazardly, hair follicles and sweat and glands with dilated ducts seen. Another component consists of mature adipose tissue divided into lobules by hyalinised fibrous tissue impression is lipodermoid (Figures 4, 5, and 6).

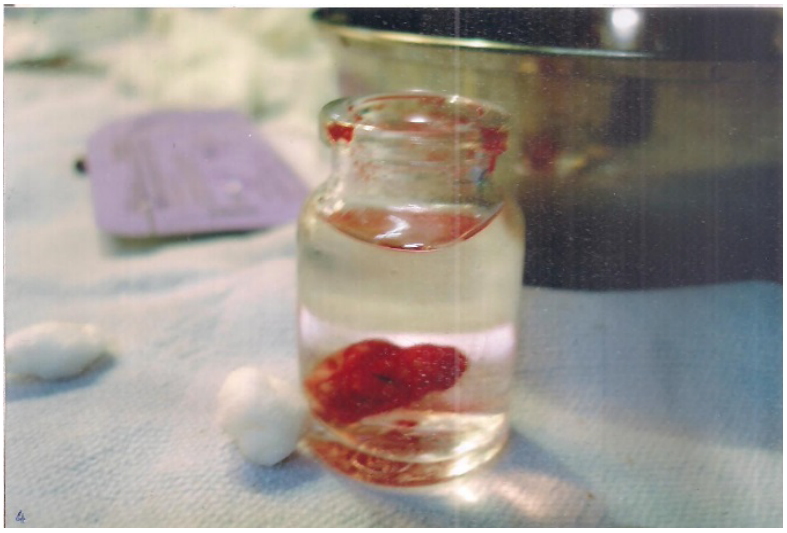

Figure 4 Adipose tissue after surgical removal

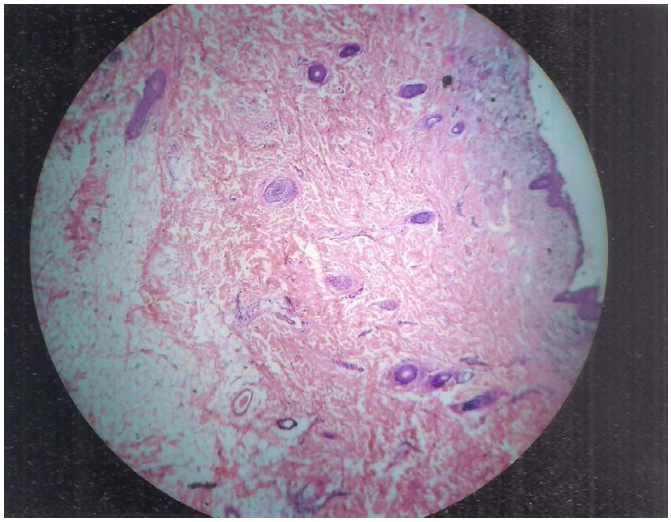

Figure 5 Microscopic examination of the excised adipose tissue

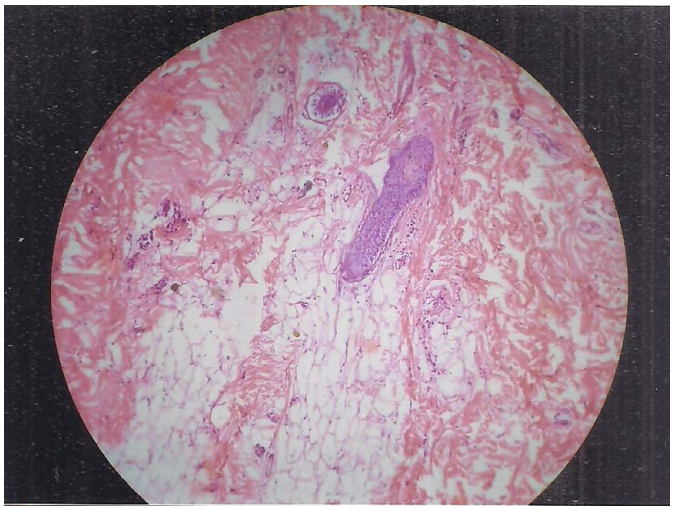

Figure 6 Microscopic examination of the excised adipose tissue

Post-operative recovery was uneventful. The child improved cosmetically with correction of lateral movement of eye, for this ocular exercise given. Patient is under regular follow up no e/o recurrence observed.

DISCUSSION

Incidence of OAV dysplasia is varying from 1 in 3500 to 1 in 5600 babies live births [1,2]. In present case, OAV hyperplasia on right side along with pretragal fistula opening and accessory auricular appendages in the external auditory opening very rarely occurs [3].

Preauricular tags open mouth pulled angle of mouth on right side along with horizontal birth mark angle of mouth to middle of cheek are in single straight line. This is very important and such rare case is reported for the first time. Goldenhar syndrome is complex entity [4]. Though it is rare congenital anomaly, the cause of it is unknown. Cosmetically unacceptable defects occur and need to be managed by team of craniofacial ophthalmic ENT orthopaedic surgeons [5,6].

CONCLUSION

Epibulbar lipodermoid is a benign, slow tumour that rarely appear on temporal side in lateral canthus of eye. To hide

unacceptable defect of lipodermoid in female child, surgical excision is best option to correct cosmetically disfigured eye for her better future. It is very necessary to make an accurate prognosis.

CONFLICT OF INTEREST

There are no conflicts of interest.

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