Original Article

Management of lipodermoid in hemifacial mircrosomia of OAV dysplasia rare case report

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Abstract

Congenital hemifacial microsomia of OAV dysplasia along with birth mark on cheek and for head is rarely occur. In this case right side of race is more affected than left side. Ocular lipodermoid is benign slow growing Tumor presented in lateral can thus of right eye, creates serious complications disfigurement of eye which is managed by cosmetic surgical approach.

Key Words: OAV dysplasia, preauricular fistula.

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INTRODUCTION

A 9 year female child presented mainly for slow growing ocular tumor in right eye parents notice increasing swelling at the age of 3 years thought to seek opinion fot it. Swelling is progressive, painless involving inferotmporal 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 medical one third of eye brow in oblige line 4.2cm long(Fig.1)

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 color swelling present in infer temporal quadrant 5 mm away from the limbos, conjuctive over the swelling is yellowish brown color, this fleshy mass having Flat drone shaped appearance, on palpation it is soft mass conjunctiva moves over it.(Fig.2) Consistency no compressible on reducible non pulsatile. Tumor mass extending up to lateral orbital margin, medical, border rounded smooth, lower border of conjunctive in fornix superiorly margin of tumor is merged in the epibulbar conjunctiva. Clinically diagnosis confirmed as epibulbar dermoid of large size disfiguring the ocular surface. Apart from this child had era 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, complete deafness present. Face on right side show facial asymmetry, under developed muscles hemifacial, microsomia. on cheek birth mark 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 epibulbardermoid, preauricular appendages with horizontal birth mark on cheek open mouth, hypoplasia of face muscles, himfacial microsomia micromandible and vertebral anomalies this combination constiture golden har syndrome. Oculo auriculo vertebral

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dysplasia its other name of constiture goldenhar. 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 vertebra fused posteriorly. No other deformity. CT-Scan of right eye orbit shows ill defined fat attenuation lesions(average attenuation -50 to -60 HU) lesions of approximate size 1.5 X 1.1X0.4 cm. Involving temporal side of conjunctival surface of right eye globe surrounding by moderately enhancing hyperdense soft tissue attenuation rim e/o mucosal thickening seen in right frontale and left maxillary sinus without any collection. Right maxillary sinus left forntal sinus and both ethmoid and sphenoid sinuses are normal no e/o mass or polyps left eye globe extraocular muscles, optic nerve on both side are normal visualized shull vault is normal opinion revales 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 nonneoplastic non inflammatory epibulbar cysts orbital dermoid cysts, foreign body granulomas, staphylomas.

Management

Depends upon severity of lesions cosmetics apporch of surgical excisison of growth to relive symptoms created by tumor mass and for confirmation of diagnosis maximum Tumor mass is excised without spilling its content care is taken to proctect 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 is covered by lose adjacent conjunctiva with sutures.

Histopathological Report

of excide speciman lesion on nouter can thus of right eye. grossly single globular solid tissue 2.5 X 1.5cm with prinkish cut surface (fig.3) Section shows stratified squamous lining with undertaking skin appendages placed haphazardly, hair follicies and sweat and glands with dilated ducts seen. Another component consist of mature adipose tissue divided in to lobules by hyalinised fibrous tissue impression is lipodermoid (Fig 4,5,6) Post operative recovery was uneventful. The child improved cosmetically with correction of lateral movement of eye, for this ocular exercise given. Paitent is under regular follow up no e/o recurrence observed.

RESULT

Epibulbar lipodermoid are benign slow tumor rarely appear on temporal side in lateral can thus of eye. To hide unacceptable defect of lipodermoid in femal child surgical excision is best option to correct cosmetically disfigured eye for her future better life. Prognosis is good.

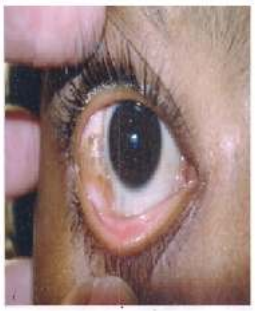

Figure 1

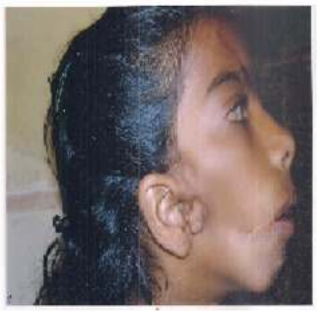

Figure 2

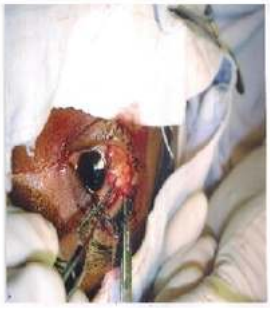

Figure 3

Figure 4

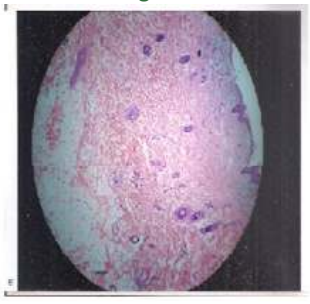

Figure 5

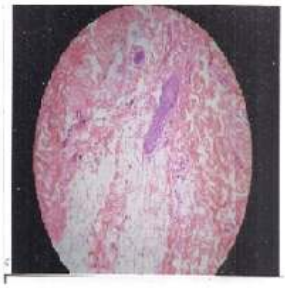

Figure 6

DISCUSSION

Incidence of OAV dysplasia is varying from 1 in 3500 to 1 in 5600 babies live birth. In present case OAV hyperplasia on right side along with pretragal fistula opening and accessory auricular appendages in the external auditory opening is very rarely occur. 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 is very important and reporting rare finding for the first time in this case Goldenhar syndrome is complex entity though it is rare congenital anomalie cause of it is unknown. Cosmetically unacceptable defects occur needs to be managed by team of craniofacial ophthalmic ent orthopedic surgeons.

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