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ANKYLOBLEPHARON FILIFORM ADNATUM AND ITS RARE ASSOCIATED SYNDROMES-A UNIQUE CASE SERIES

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ABSTRACT This paper presents a case series of two neonates reporting with difficulty in opening the eyes on the first day of life. On ocular examination multiple adhesional bands are found between lids in both the eyes. On systemic evaluation a smooth philtrum, low set ears suggesting Edward's syndrome was revealed in baby A and baby B had posterior cleft palate, dermal erosions along with mandibular dysplasia revealing ectodermal dysplasia cleft palate syndrome. Transection under local anesthesia was done on both the babies. Examination post transection revealed normal eyelid function. Ankyloblepharon Filiform Adnatum (AFA) in a neonate should alert the ophthalmologist because it can be rarely associated with life threatening multi-systemic defects. This case series aims to highlight such rare presentations and exemplifies the need for multidisciplinary approach. Early surgical intervention performed can reduce the risk of occlusion induced amblyopia and also helps in the detailed ophthalmologic examination later in life.

KEYWORDS :

INTRODUCTION

Ankyloblepharon filiforme adnatum (AFA) is a congenital anomaly where complete or partial fusion of the ciliary edges of eyelids is seen due to formation of single or multiple thin bands between the margins of the eyelids anterior to the tarsal gland orifices and posterior to the cilia thus causing reduction in the palpebral fissure height.[1] It is usually benign but rarely be associated with multiple systemic disorders, which can cause remarkable complications later in life. We report here a case series of two such new-borns presented with syndromic Ankyloblepharon Filiform Adnatum (AFA) at birth, successfully treated in the initial days of life.

CASE REPORT:

Case 1- Baby A

A 1-day-old full term female baby delivered by spontaneous vaginal delivery weighing 2.5kg during birth was referred from the department of neonatology, Sri Ramachandra Institute of Higher Education and Research, Chennai, India with difficulty in opening her eyes at birth. The antenatal, intranatal, and postnatal period were uneventful. The mother reports a history of no drug intake other than prescribed supplements, no history of radiation exposure, no history of any congenital anomalies or consanguinity in the family. On ocular examination the baby had bilateral partial adhesion of left upper and lower eyelids with two thin stretchy bands measuring 2mm and 1 mm of length and width respectively. Three similar bands were noticed in the right eye. Detailed systemic evaluation revealed smooth philtrum, low set ears, precordial bulge, supra sternal retraction and prominent occiput. Cranial Ultrasound was normal. Echocardiogram showed presence of Large Ventricular septal defect (VSD), Atrial septal defect (ASD), Patent ductus arteriosus (PDA) and pulmonary hypertension. These features suggestive of Edward's syndrome was confirmed by fluorescence in situ hybridization (FISH) karyotyping.

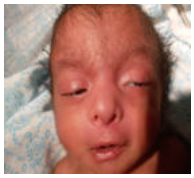

Figure 1 – Shows Fused Eyelids And Smooth Philtrum In Baby A.

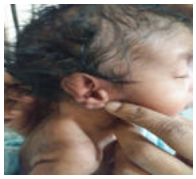

Figure 2- Shows Low Set Ears In Baby A.

Case 2 -Baby B

A 3 day-old, full term, male neonate with birth weight of 2.31kg was delivered by spontaneous vaginal delivery, was referred from the department of neonatology, Sri Ramachandra Institute of Higher Education and Research, Chennai, India for difficulty in opening both the eyes. The antenatal, intranatal, and postnatal periods were uneventful. Elder sibling was healthy with no systemic illness. There is no history of congenital anomalies or consanguinity in family. Maternal history revealed no habit of smoking, alcohol consumption and drug intake.

Detailed ophthalmic examination revealed two fine extensible bands of skin in both the eyes each measuring about 1 mm in breadth and 2 mm in length. On physical examination, baby had features of cleft palate, mandibular hypoplasia, Dysplastic nails, Broad nasal bridge, Mongolian spot on the trunk along with dermal erosions suggestive of Ankyloblepharon ectodermal dysplasia cleft lip/ cleft palate syndrome. Renal ultrasound showed right hydroureteronephrosis. Echocardiogram revealed mild Left ventricular hypertrophy.

Figure 3- Shows Fused Eyelids In Baby B

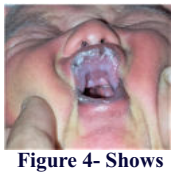

Figure 4- Shows Cleft Lip And Cleft Palate In Baby B.

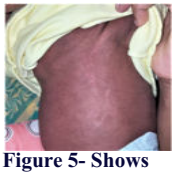

Figure 5- Shows Mongolian Spots In Baby B.

Both the babies underwent transection following aseptic precautions. The elastic strands were transected with No. 15 blade under topical anaesthesia. No sedation was required. Post procedure minimal bleeding was noted at the transection site. Eyelid function, anterior segment, fundus examination and ocular movements were found to be normal on post transection examination.

Figure 6- Eyelids Post Transection In Both Babies (a And B).

DISCUSSION

The incidence of Ankyloblepharon Filiform Adnatum (AFA) in India is 4.4 in 1 million births[2]. Ankyloblepharon Filiform Adnatum (AFA) being a rare birth defect can usually be missed at birth due to periocular oedema during early examination. The fusion of eyelids is seen until the 5th month of gestational period but is not present after birth[3]. Ankyloblepharon filiforme adnatum (AFA) was first expressed by Von Hasner in the year 1881. These bands of tissue between the lids usually consist of stratified squamous epithelium surrounding a vascularised central core.[4]

Ankyloblepharon Filiform Adnatum (AFA) can be classified broadly into 4 groups according to Rosenman et al[5]. Groups 1, 2, 3 and 4 were described as Ankyloblepharon Filiform Adnatum (AFA) without other associated anomalies, Ankyloblepharon Filiform Adnatum (AFA) with association of cardiac or central nervous system anomalies, Ankyloblepharon Filiform Adnatum (AFA) with associated ectodermal syndromes and Ankyloblepharon Filiform Adnatum (AFA) associated with cleft lip and/or palate syndromes respectively. Also groups 1 and 2 were described as sporadic dominant and group 3 and 4 as autosomal dominant with variable expressivity. An addition of a fifth group: Ankyloblepharon Filiform Adnatum (AFA) in association with chromosomal abnormalities was suggested by Bacal et al.[6]

Initially Ankyloblepharon Filiform Adnatum (AFA) was thought to be a partial failure of the eyelids to separate. Later, several studies reported that Ankyloblepharon Filiform Adnatum (AFA) could be a result of a pathological skin growth which is of inflammatory origin or connective tissue growth due to a defect in the epithelium of the foetus. Most common cause was reported as trauma during fetal life (e.g. trauma due to fingernail)[7]

Generally, the temporary fusion of eyelids in-utero is composed solely of epithelium and is completed by the eleventh week of gestational age. Thus it is clear that the presence of mesenchymal bands is due to abnormality in the fusion process and not in separation. The currently accepted theory is that of pure aberrance of development due to interplay of temporary epithelial arrest and abnormally rapid proliferation of mesenchymal tissue, allows fusion of the lids at certain points without interposition of epithelium.[8]

Ankyloblepharon Filiform Adnatum (AFA) was rarely found to be present with ophthalmic condition like developmental anomaly of iris and angle (iridio-gonio dysgenesis) with juvenile glaucoma.[9] Other syndromic conditions [Edwards′ syndrome , Hay-Wells syndrome[10], Curly hair-ankyloblepharon-nail dysplasia syndrome(CHANDS)[11], popliteal pterygium syndrome[12]] and systemic conditions (meningocele, hydrocephalus, bilateral syndactyly, imperforate anus and cardiac problems like patent ductus arteriosus and ventricular septal defects) were reported to be in association with Ankyloblepharon Filiform Adnatum (AFA)[13].Detailed systemic assessment by an experienced pediatrician is the main forte in the management of Ankyloblepharon Filiform Adnatum (AFA).

Eyelids examination of every neonate should be an integral part of neonatal physical evaluation before discharging the new-born, to detect this condition at early stage itself . Due to association of eyelids malformations with multiple systemic diseases, a thorough Systemic evaluation is necessary in the presence of an anatomic anomaly such as Ankyloblepharon Filiform Adnatum (AFA). The practical importance of this case series is that prompt and early surgical intervention can be performed to reduce the risk of occlusion induced amblyopia and for detailed ophthalmologic examination.

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