Extensive isolated bilateral Ankyloblepharon Filiforme Adnatum: a case report

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Abstract

We report a case of a rare congenital eyelid abnormality, ankyloblepharon filiforme adnatum, in an otherwise healthy male neonate, born to healthy parents out of non-consanguineous marriage. This newborn presented with extensive band of tissue connecting upper and lower eyelid of both eyes, resulting in extremely limited palpebral aperture opening. Antenatal history was unremarkable. The parents came to us at the age of one month. The baby was operated on that day at admission under local anesthesia. The eyelid band of both eyes were excised at the level of each lid margin resulting in normal eye opening. Dilated fundoscopic examination was done along wit,h anterior segment examination. No underlying ocular abnormalities were noted. A detail systemic examination by pediatrician failed to reveal any other congenital anomaly. Echocardiography and cerebral echography (both normal) were performed to exclude congenital heart disease and cerebral malformations. The baby was then discharged without complications. Ankyloblepharon filiforme adnatum may be isolated or may present with other congenital anomalies. Therefore, the presence of eyelid bands should alert for complete systemic examination. Prompt treatment is required for proper opening of the eye and to decrease the risk of amblyopia.

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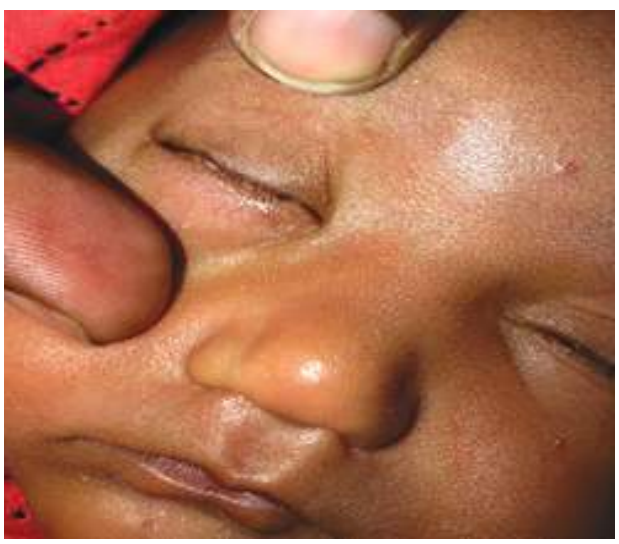
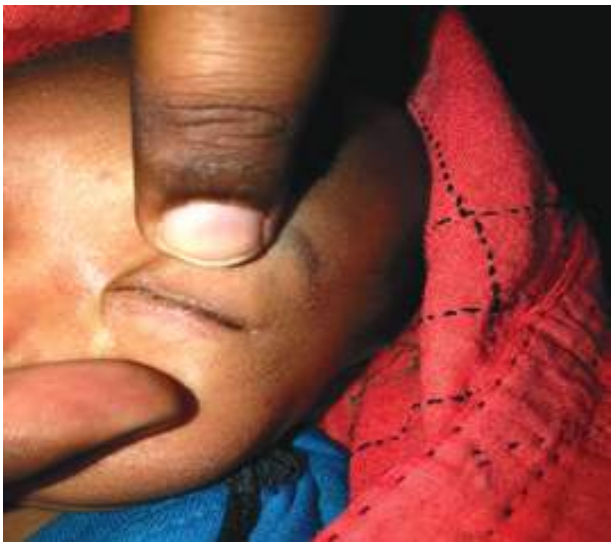
Discussion:

Ankyloblepharon filiforme adnatum (AFA) is a rare congenital anomaly[1,2] consisting of a partial or complete fusion of the eyelid margins. Such condition is normal during the fetal life (eyelids normally remain fused until the 5th month of gestation)[3], but not after birth. It is described as a single or multiple tissue band between the upper and lower eyelid margins. Rosenman's 1980 classification divides AFA into four subgroups (1, isolated; 2, associated with cardiac or central nervous system anomalies; 3, associated with ectodermal syndromes; 4, associated with cleft lip and/or palate)[4].

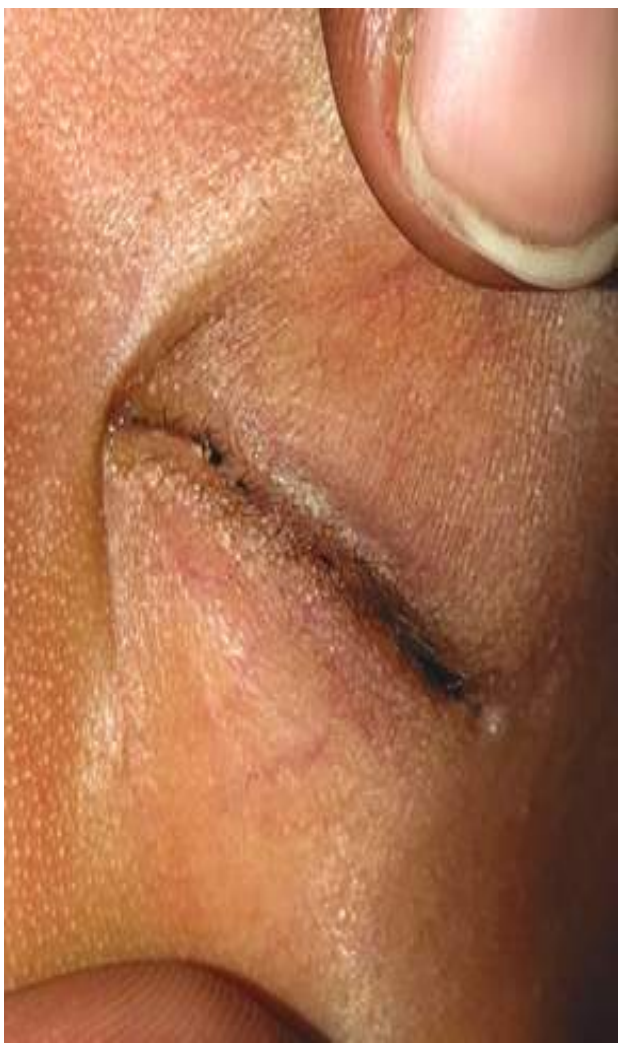
More recent findings show that AFA can indeed occur isolated as well as along with other ocular anomalies[5], trisomy 18[6,7], or with other multisystemic syndromes, particularly Hay-Wells syndrome[8,9], also known as ankyloblepharon-ectodermal defects–cleft lip/palate (AEC) syndrome[9]: this condition includes a great amount of congenital malformations and anomalies (ectodermal defects, cleft-lip/palate, limb anomalies) associated with AFA[9] . AFA, in isolation, is of unknown etiology with an incidence of 4.4 per 100 000 births. AFA describes single or multiple bands of tissue joining the upper and lower eyelids either unilaterally or bilaterally. It may present as an isolated congenital defect such as in our patient. However, it is always important to actively look for coexisting pathology.Furthermore, it might be associated with hydrocephalus and cardiac defects[2,3]. Therefore detailed systemic assessment by a paediatrician is imperative in the management of AFA. The ophthalmic association of AFA is iridogoniodysgenesis with juvenile glaucoma.

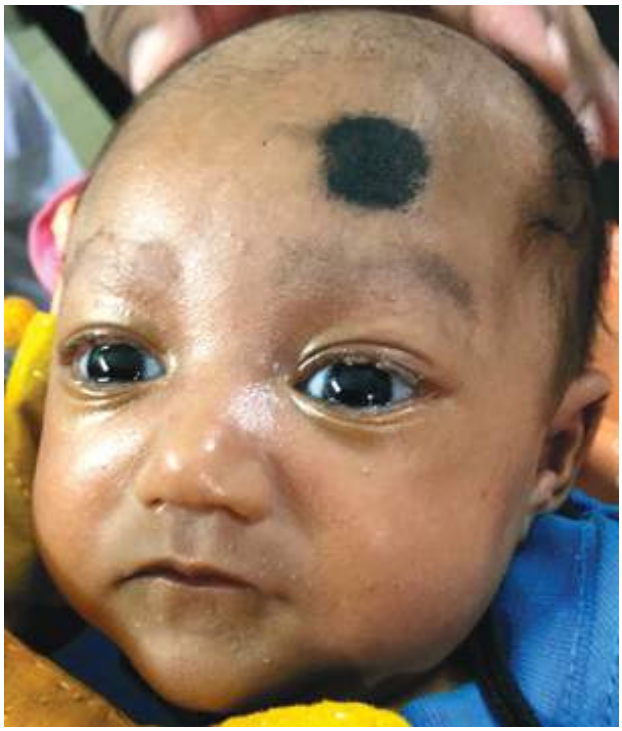
In Our report illustrates a simple surgical approach that is modified from previously published cases. It is safe and well tolerated with the aid of topical anaesthesia. Surgical correction should be performed promptly to minimise any risk of occlusion amblyopia, and enable full examination of the eye.





Picture 1,2,3: Multiple band of tissues in both eyes and picture 4 is the appearance after excision of the bands.





Conclusion:

Ankyloblepharon filiforme adnatum is a rare congenital condition, which may be easily missed at first clinical examination at birth. Examination of eyelids represents a fundamental part of neonatal physical evaluation to avoid future functional problems like impaired vision or amblyopia, and as eyelids malformations could be a sign of multisystemic disease a thorough evaluation of other body systems to exclude associated malformations is strongly recommended. Surgical correction should be performed promptly to minimise any risk of occlusion amblyopia, and enable full examination of the eye.

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