

CASE REPORT

A NEWBORN WITH ANKYLOBLEPHARON FILIFORME ADNATUM WITH CLEFTING: A CASE REPORT

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HOW TO CITE THIS ARTICLE:

Yogesh R. B, Anusha Arvind, K. Prahallada Reddy, G. Pavan, Deepashri Mutalik. "A Newborn with Ankyloblepharon Filiforme Adnatum with Clefting: A Case Report". Journal of Evolution of Medical and Dental Sciences 2014; Vol. 3, Issue 62, November 17; Page: 13787-13790, DOI: 10.14260/jemds/2014/3844

ABSTRACT: Ankyloblepharon filiforme adnatum is congenital anomaly involving abnormal palpebral fissure opening. We report a case of bilateral ankyloblepharon filiforme adnatum in a newborn Indian male who had associated features of clefting. Ankyloblepharon filiforme adnatum can present as an isolated finding or in association with other anomalies, or as part of a well-defined syndrome.

KEYWORDS: Ankyloblepharon, clefting, fibrous band, release of band occlusion amblyopia.

INTRODUCTION: Ankyloblepharon filiforme adnatum (AFA) is a rare benign congenital anomaly where the developing ciliary upper and lower eyelid margins remain fused until the fifth month of gestation, but may not be completely separated until the seventh month. Ankyloblepharon filiforme adnatum is usually a sporadic isolated malformation.⁽¹⁾

CASE PRESENTATION: A male neonate was born to 26-year-old primi of a non-consanguineous marriage at 40 weeks of gestation by a normal vaginal delivery following an unremarkable pregnancy. The baby boy adapted without difficulties with Apgar scores of 7, 9, and 10 at 1, 5 and 10 minutes, respectively. His birth weight was 3100 g. Anomaly scan done at 20 weeks was normal.

The neonatal examination revealed a single band of tissue vertically attached to the upper and lower eyelids in both the eyes, covering the pupil and preventing full opening of the eyelids (Figure 1 & 2). A detailed systemic examination revealed associated cleft lip & palate, low set ears (Figure 3), scattered cafe-au lait spots, nuchal pad of fat and prehensile toe. Further investigations such as 2D echocardiography and ultrasonography of the abdomen revealed no abnormalities. There were no similar congenital malformation in the family, no family history of eye or systemic diseases, and the mother denied taking any drugs besides iron, folic acid and calcium supplementation.

The band of tissue was divided by one cut using a scissors, without necessitating either sedation or administration of a local anesthetic on post natal day 12. No bleeding or distress was seen immediately after the procedure. The baby had bleeding from the excised area of the right upper eye lid on the following day, which was sutured using absorbable vicryl suture to control the same. Following division of the vertical bands thorough ocular and fundus examination was done and found to be normal (Figure 4).

CASE REPORT

Fig. 1 & 2: A fibrous band connects the upper and lower eye lids (1) and prevents opening of the eye (2).



Fig. 1

Fig. 2

Fig. 3 & 4: Associated features of low set ears, cleft lip and palate. Normal appearance of the right eye following division of the band of tissue.



Fig. 3

Fig. 4

DISCUSSION: Ankyloblepharon filiforme adnatum is characterized by the presence of isolated strands of extensile tissue passing between the upper and lower lid margins. This aberrant tissue attaches to the lid margin between the cilia and meibomian gland orifices and can involve one or both lids to varying degrees.⁽²⁾ AFA was first described by Josef von Hasner in 1881.

Classification of AFA into four subgroups by Rosenman⁽³⁾ et al is as follows: Group I includes sporadic cases without any associated defects. Group II is also sporadic, but AFA is found in association with cardiac, gastrointestinal and CNS defects. Group III represents cases of AFA in association with ectodermal syndromes (ankyloblepharon-ectodermal dysplasia-clefting AEC syndromes) syndromes as seen in this case. Group IV represents cases of AFA with cleft lip and palate defects in the patient or in the extended family.

CASE REPORT

The pathogenesis of this condition remains unclear. It has been proposed that a relative arrest of epithelial growth allows a more rapid proliferation of mesodermal tissue to bridge the future lid fissure. The subsequent separation of the lid fold stretches these bands and contributes to their sometimes-elongated appearance.⁽⁴⁾ Usually, AFA constitutes a solitary malformation, with autosomal dominant or sporadic occurrence as in our case, and an incidence of 4.4 per 100,000 births.^(5,6)

However, it can be associated with several disorders such as trisomy 18 (Edward's syndrome),⁽⁷⁾ Hay-Wells syndrome (a variant of the ectodactyly-ectodermal dysplasia-cleft lip palate syndrome),⁽⁸⁾ popliteal pterygium syndrome (characterised by intercrural webbing of the lower limbs),⁽⁹⁾ CHANDS (curly hair-ankyloblepharon-nail dysplasia)⁽¹⁰⁾ and cleft lip and palate.⁽¹¹⁾

Other associations may include hydrocephalus, meningomyelocele, imperforate anus,⁽¹²⁾ bilateral syndactyly,⁽⁹⁾ infantile glaucoma,⁽¹³⁾ and cardiac problems such as patent ductus arteriosus and ventricular septal defects.⁽⁹⁾

Timely separation of the eyelids is crucial to avoid the development of occlusion amblyopia, enable full examination of the eye, alleviate parental stress and for neonatal comfort.

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CASE REPORT

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Date of Submission: 29/09/2014.
Date of Peer Review: 30/09/2014.
Date of Acceptance: 13/11/2014.
Date of Publishing: 17/11/2014.