Ankyloblepharon filiform adnatum: a case report
Ankyloblepharon filiform adnatum : à propos d’un cas

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Abstract:
Ankyloblepharon filiform adnatum (AFA) is defined by partial or complete fusion of the lid margins. It can cover a part of the eyelid or its totality. Diagnosis should be done at an early stage of life. It needs a surgical correction. We describe the case of a newborn at his twentieth day of life, referred to us for assessment of his both right and left eyelids. He was born on term. Pregnancy and delivery went normally. The baby was healthy. However, we found multiple bands of tissue between upper and lower eyelids of both of his eyes. These bands were axial, covering the pupil. Full eyelid opening was incomplete. The baby underwent surgery which consisted on excising the bands of tissue with scissors at the level of each eyelid margin. Eye examination did not reveal any abnormality. Two months later, the follow up showed no anomaly. Ankyloblepharon filiform adnatum may be rare but it is potentially amblyogenic. The ophtalmological examination must always be completed in order to look for coexisting congenital anomalies. The eyelid malformation should be treated surgically as soon as the diagnosis is done.

Keywords: Ankyloblepharon filiform adnatum ; eyelid ; surgery

Résumé:
L’ankyloblepharon filiform adnatum (AFA) est défini par une fusion partielle ou complète des paupières par des toiles de peau. Il peut intéresser une partie de la paupière ou sa totalité. Le diagnostic doit être fait à un stade précoce de la vie. Le traitement est essentiellement chirurgical. Nous décrivons le cas d’un nouveau-né à son vingtième jour de vie qui a été référé pour prise en charge de sa malformation des paupières. Il est né à terme. La grossesse et l’accouchement se sont déroulés normalement. Le bébé était en bonne santé sauf à l’exception de multiples bandes de tissu entre les paupières supérieures et inférieures de ses deux yeux. Ces bandes étaient axiales, recouvraient la pupille et empêchaient l’ouverture complète des deux paupières. Le bébé a subi une intervention chirurgicale consistant à exciser les bandes de tissu avec des ciseaux au niveau de chaque bord de paupière. L’examen oculaire n’a révélé aucune anomalie. Deux mois plus tard, le suivi n’a montré aucune anomalie. L’Ankyloblepharon filiform adnatum peut être rare mais il est potentiellement amblyogène. Cependant, l’importance majeure de cette anomalie est d’alerter le médecin sur la présence d’autres anomalies congénitales associées. Le traitement doit être effectué dès le diagnostic.

Mots-clés : Ankyloblepharon filiform adnatum ; paupières ; chirurgie

Introduction
Ankyloblepharon filiform adnatum is defined by partial or complete adhesion of the ciliary edges of the upper and lower eyelids at the gray line, by single or multiple fine bands of extensible tissue, which reduces the opening of the eyelid. It is amblyogenic and its association with other congenital malformations may account for high mortality and morbidity [1]. We report here a case of Ankyloblepharon filiform adnatum in a male newborn and describe its management.

Clinical case
A newborn at his twentieth day of life was referred for assessment of his both right and left eyelids. He was born on term. Pregnancy and delivery went normally. The antenatal, intranatal, and postnatal periods were uneventful. The mother denied taking any drugs except iron and vitamin supplements. There was no history of any X-ray exposure. There was no family history of congenital anomalies. A detailed clinical assessment by a paediatrician was normal. Ocular examination

Figure 1 : (before treatment) Multiple fibrous bands between the upper and lower eye lids
showed bands of tissue between upper and lower eyelids of both of his eyes. These bands were axial, covering the pupil, and the full opening of both eyelids was incomplete (figure I).

The baby underwent surgery which consisted on excising the bands of tissue with scissors at the level of each eyelid margin. Eye examination did not reveal any abnormality. Two months later, the follow up showed no anomaly (figure II).

**Discussion**

First described by Von Hasner in 1881, Ankyloblepharon filiforme adnatum is a rare benign congenital anomaly, usually associated with multiple and complex malformations, suggestive of an autosomal dominant inheritance of varying degrees of penetrance. Sporadic cases have, however, also been described [2]. No gender predominance has been reported [3]. The fusion of eyelids is a normal stage in human development. The developing eyelid margins remain fused until the fifth gestational month but may take up to the seventh month of gestation to be completely separated [4]. Fusion seen at birth as in Ankyloblepharon filiforme adnatum is abnormal.

The etiology of Ankyloblepharon Filiform Adnatum is unknown, but failure of apoptosis at a critical stage in eyelid development has been suggested. Usually, Ankyloblepharon Filiform Adnatum constitutes a solitary malformation, as in our case, with sporadic occurrence. However, it can be associated with several disorders [5].

Diagnosis of ankyloblepharon is based on clinical features. It is defined by partial or complete fusion of the eyelid margins [6]. A significant central ankyloblepharon may interfere with vision and lead to amblyopia.

The length of the bands varies from 1 to 10 mm and the width varies from 0.3 to 0.5 mm and are invariably extensible. The bands always lie between the cilia and orifices of the tarsal gland. The band is always composed of a central vascular connective tissue strand surrounded by pavement epithelium. The connective tissue is usually highly cellular and embryogenic in nature, but Cordero found muscle fibers and numerous sub-epithelial glands in the histological study of the band. When the bands are cut, bleeding may occur [1]. Pathogenesis of the condition is disputed. Various theories have been proposed including a simple defect in separation, pathologic growth of the skin with an inflammatory origin, an epithelial defect in fetal life through which connective tissue grows, and traumatic cause as fingernail trauma in utero. But the most accepted theory is that of pure aberrance of development, due either to a temporary arrest of the growth of epithelium or more probably, an abnormally rapid proliferation of mesoderm allowing union at certain points of the mesenchyme of the lid folds without epithelial interposition [1].

It is usually a solitary malformation of sporadic occurrence, but can occur in an autosomal dominant pattern associated with cleft lip and palate in most familial cases. In some patients, it appears as a part of Edward’s syndrome (Trisomy 18), Hay-Wells syndrome (a variant of ectodactyly-ectodermal dysplasia-cleft lip-palate syndrome), popliteal pterygium syndrome (characterized by intercrural webbing of the lower limbs), and curly air-ankyloblepharon-nail dysplasia (CHANDS). It may also be reported in association with hydrocephalus, meningocele, imperforate anus, bilateral syndactyly, infantile glaucoma, and cardiac problems such as persistent ductus arteriosus and ventricular septal defect [7-10].

An unusual report of a multiple malformation syndrome with Ankyloblepharon filiforme adnatum, with cleft lip and palate, bilateral popliteal pterygia, bilateral complete syndactyly of 2nd and 3rd toes and hypoplastic nails, accessory nipple, and partially descended testis has also been reported [2].

Management of ankyloblepharon is mainly surgical. Detailed systemic assessment by an experienced pediatrician is essential to rule out coexisting pathology and proper management of congenital ankyloblepharon. Fine bands of adhesions can be broken by forcibly separating the lids or with use of muscle hook, sharp scissors or a scalpel [11]. The fine remnants at the lid margin usually shrink and resolve. Bipolar cautery forces can be used at the bases of fine filaments to release the adhesions. In congenital ankyloblepharon, visual prognosis is usually good if timely intervention is performed. Cosmetic appearance after separation of the bands is usually very good [12]. A spontaneous resorption of the bridles can be observed after a few months. The prognosis depends on the associated anomalies [13].
Conclusion

Ankyloblepharon filiform adnatum may be rare but it is potentially amblyogenic and it requires further investigation. Treatment should be performed as soon as the diagnosis is done.

References


CONFLICTS OF INTEREST:
None