Sir,

Management of ankyloblepharon filiforme adnatum

Ankyloblepharon filiforme adnatum (AFA) is a rare congenital malformation affecting the eyelids. We report a case describing its management.

Case report

A male neonate was referred for assessment of his left eyelids. He was born at term, to a primigravid mother, weighing 3150 g. Pregnancy and delivery were unremarkable. There was no family history of ophthalmic or systemic disease. A detailed systemic assessment by a paediatrician was clear.

Ocular examination showed partial fusion of his left upper and lower eyelids by a central, narrow band of tissue, arising from the grey lines (Figure 1). Full eyelid opening was impaired and interpalpebral aperture was limited to 3.5 mm. Right eye examination was normal. The band of tissue was retracted anteriorly with a squint hook, clamped for 10 s, and excised with Vannas scissors at the level of each eyelid margin. The procedure was performed under corneal anaesthesia using benoxinate 0.4%. There was no sign of distress and no bleeding. Subsequent left eye examination did not reveal any other pathology. At 2 years' follow up, logMAR visual acuity was +0.1 in both eyes and examination was normal.

Discussion

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.

The ophthalmic association of AFA is iridogoniodysgenesis with juvenile glaucoma.[1] Systemically, AFA is associated with cleft lip and palate,[2] occasionally in the context of ectodermal dysplasia syndromes[3] and popliteal pterygium syndrome.[4] AFA has also been reported in association with Edward's syndrome,[5] and CHANDS[6] (curly hair, ankyloblepharon, nail dysplasia). Other associations include hydrocephalus, meningomyelocoele, and imperforate anus,[7] cardiac defects and syndactyly.[4] Detailed systemic assessment by an experienced paediatrician is therefore imperative in the management of AFA.

Our report illustrates a simple surgical approach that is modified from previously published cases.[1,2,4] 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.

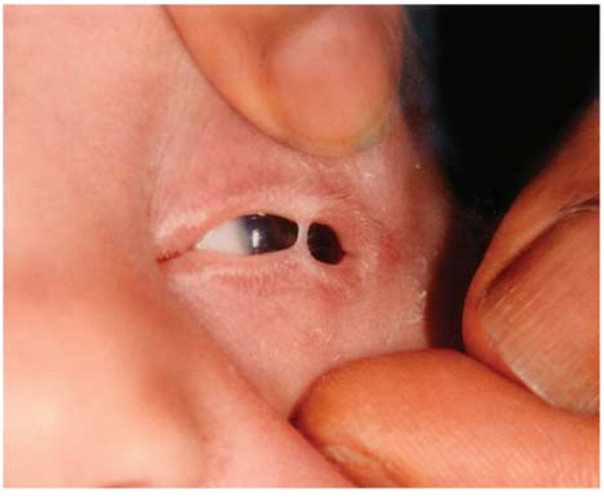

Figure 1 Photograph of the neonate with ankyloblepharon filiforme adnatum showing partial fusion of the left upper and lower eyelids by a band of tissue.

Conflict of interest

The authors declare no conflict of interest.

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Sir,

Enhanced rod–cone interaction with progressive macular dysfunction

Case report

A 22-year-old man was referred with decreased visual acuity of 4 years duration and bilateral 'positive'

scotomata. Past medical and ocular history was unremarkable. Visual acuity (VA) was 0.30 logMAR in the right eye (OD) and 0.60 in the left (OS). Slit lamp and fundus examination were normal. Automated perimetry (Humphrey 24–2) showed small bilateral central scotomata. Electrophysiologic tests, including full field and pattern electroretinography (PERG), were normal, but modified dark adaptometry was consistent with enhanced rod–cone interaction (E-RCI) (Figure 1). After 6 years, he was complaining of poor central vision and constant flickering, especially when going from a dark to an illuminated environment and VA had dropped to 0.79 OD and 0.83 OS. Fundus exam remained normal. PERG elicited significantly reduced activity from both eyes, and a multifocal ERG (mfERG) showed central macular dysfunction. Full-field ERGs remained normal showing the dysfunction to be confined to the maculae. Three dimensional Optical Coherence Tomography (3D-OCT) showed bilateral disruption of the photoreceptor inner segment/outer segment (IS/OS) layer (Figure 2).

Comment

E-RCI, first described by Arden[1,2] in 1985, is a rarely reported disorder in which the normal elevation of dark adapted cone thresholds by dark adapting rods is greatly enhanced. A modified dark adaptometer is needed to make the diagnosis, as conventional Goldman-Weekers dark adaptometer cannot adequately differentiate rod and cone thresholds and cannot evaluate dark adapted cone sensitivity/thresholds. In this case, modified dark adaptometry[1] demonstrated enhanced elevation of cone

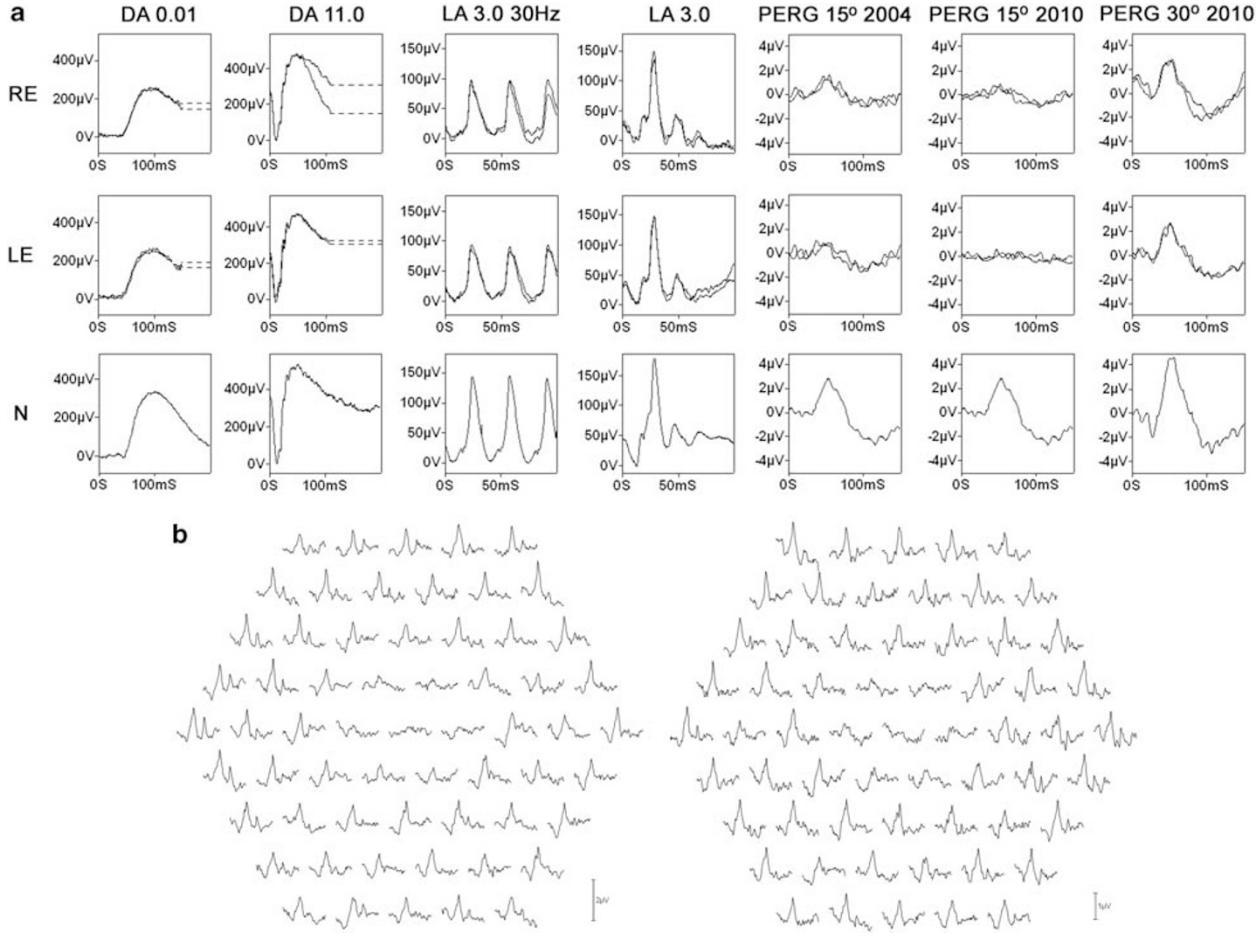

Figure 1 Electrophysiology and psychophysics. (a) ERGs and PERGs. Full field ERGs were normal in both 2004 and 2010, and only the later data are shown. PERG shows significant deterioration with a standard field size between 2004 and 2010. Large field PERG data, only available for the recent visit, suggest relative sparing of paramacular function. Data from a representative normal subject are shown in the lower row. The paramacular sparing is confirmed by the mfERG (b) where there is marked loss of the responses to central hexagons, but preservation of the responses to peripheral hexagons (field size = 55 degrees). (c) Dark adaptometry: dark adaptation (DA) shows enhanced rod–cone interaction. The dashed line shows the conventional DA response from a normal subject; the solid line shows the threshold measurements to a red 15 Hz flicker (cone mediated). Note the normal elevation of the cone threshold by 0.7–1.2 log units as rod sensitivity increases. The hollow squares show the rod data from the patient; the filled squares are the cone-mediated responses showing abnormal threshold elevation (note that the responses shown as 0.0 log units include (undetected) responses where the threshold response exceeded the range of the instrument). The two blocks of data at 40 min are those to a 1 Hz 200 ms flash using blue (rod) and red (cone) stimuli for final threshold measurement.

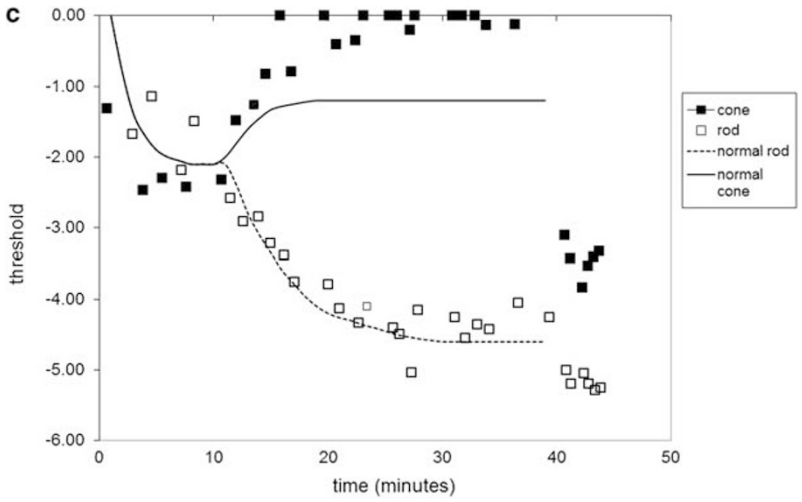

Figure 1 Continued.

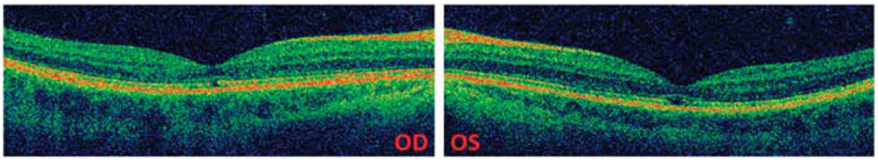

Figure 2 The 3D-OCT showing bilateral disruption of the photoreceptor inner segment/outer segment layer and intact outer limiting membrane.

thresholds by dark adapting rods and established the diagnosis of E-RCI. The condition is usually benign and previous reports have not shown progression.[3–5] The continuing reduction in VA in the present case and the markedly reduced PERG and mfERG suggested progressive macular dysfunction, which was confirmed by 3D-OCT demonstration of bilateral disruption of the photoreceptor IS/OS layer. To our knowledge, this is the first case of progressive macular dysfunction in association with E-RCI. It demonstrates the value of electrophysiology, psychophysics, and OCT in revealing functional and structural abnormalities despite the presence of a normal fundus exam.

Conflict of interest

The authors declare no conflict of interest.

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Sir,

A case of African crystalline maculopathy

We present the first reported case of West African crystalline maculopathy in an East African patient from Egypt.