Fibrous Band between Extraocular Muscles in Unilateral Coronal Synostosis

Dear Editor,

Unilateral coronal synostosis is an abnormal premature fusion of one of the coronal sutures. As a consequence of the shortened orbital roof secondary to the fused coronal suture, the ipsilateral trochlea is displaced posteriorly [1-3]. Therefore, unilateral coronal synostosis is frequently associated with development of strabismus, most typically superior oblique (SO) palsy on the ipsilateral side of the synostosis [1-3]. However, contrary to syndromic craniosynostosis, anomalies of the extraocular muscle structure themselves have not been reported in unilateral coronal synostosis [4]. We report a case of unilateral coronal synostosis and SO palsy with a fibrous band, which was found intraoperatively to course between extraocular muscles.

An 18-month-old boy with non-syndromic unilateral coronal synostosis in the right coronal suture was referred for a persistent left head tilt despite cranioplasty. He had undergone an expansion cranioplasty with trans-suture distraction osteogenesis at 14 months of age. He had a 15-degree head tilt to the left. Motility examination showed flick right hypertropia in primary position with a positive head tilt test on right head tilt. The fundus was normal in each eye, but that of the right eye was mildly excyclorotated. After an additional cranioplasty at 34 months of age, asymmetry of the orbits was improved. A computerized tomography scan of the orbit demonstrated that all rectus muscles and SO muscles were present in both eyes, but the right SO muscle was posterior compared with the left SO muscle (Fig. 1A). At 5 years of age, the patient had right hypertropia of 8 prism diopters (PD) in primary position that increased to 16 prism diopters on right head tilt, associated with +2 right inferior oblique (IO) overaction. He was orthotropic on left head tilt (Fig. 1B). Fundus examination revealed moderate excyclotorsion in the right eye (Fig. 1C).

A right IO myectomy was performed. At the time of surgery, Guyton’s exaggerated forced duction test for the right SO or IO muscle was not significant, but a thick and fibrous band that connected the inferior edge of the right lateral rectus muscle 3 mm posterior to the insertion to the right IO muscle was found and removed (Fig. 1D).

One year after muscle surgery, the patient’s corrected visual acuity was 20 / 20 in each eye with myopic correction of -1.00 diopter. He had no head tilt and orthotropia in primary position with normal oblique function (Fig. 1E). Fundus examination revealed no excyclotorsion in either eye (Fig. 1F).

It is well known that the retro-displaced trochlea in patients with unilateral coronal synostosis may result in relative underaction of the SO muscle on the synostotic side, mimicking SO palsy [1-3]. Thus, surgical correction for asymmetry of the orbits might improve the imbalance between SO and IO muscles. However, SO palsy in the present case persisted despite successful correction of orbital asymmetry. It has been reported that, after fronto-orbital advancement, commonly performed in unilateral coronal synostosis, resolution of strabismus is rare, while a new strabismus developed [1,2]. During fronto-orbital advancement, the bony bandeau only displaces anteriorly after dissection of the periorbit, so that the trochlea may be further posteriorly displaced, aggravating the retro-positioning of the SO muscle [1,2]. Although expansion cranioplasty with distractor in the present case involved only the coronal suture, different in the surgical plane from fronto-orbital advancement, retro-displacement of the trochlea remained based on the postoperative images. It is possible that correction for retrusion of the superior-temporal orbit with expansion cranioplasty was not sufficient to resolve or reduce the retrodisplacement of the trochlea at the superior-nasal orbit. In addition, resultant changes in sarcomere length of the extraocular muscles in pattern strabismus have been reported in early infancy [2].

It is well known that there is a high prevalence of anomalies of the extraocular muscles in syndromic craniosynostosis, such as those related to Crouzon, Apert, or Pfeiffer...
syndrome [4]. Mutations in fibroblast growth factor receptor (FGFR) 2 are mostly responsible for syndromic craniosynostosis, and FGFR-2 has been reported to be predominantly expressed within extraocular muscles [4]. However, a mutation in the FGFR-3 gene is responsible for some cases of unilateral coronal synostosis. On the other hand, a non-ocular rotary anomalous extraocular muscle band could coincidentally exist [5]. Nevertheless, this is the first report of an anomalous extraocular muscle in a patient with unilateral coronal synostosis.

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Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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