Congenital Bilateral Medial Rectus Muscle Aplasia

To the Editors:

Ocular motility problems related to congenital abnormalities of the extraocular muscles are rare. Such congenital abnormalities may present in a spectrum from accessory additional rectus muscles to the absence of extraocular muscles. Absence of one or more extraocular muscles is a rare condition usually seen in association with craniofacial syndromes. We present a case of bilateral medial rectus muscle aplasia with a constant exodeviation in the absence of associated cranial facial anomalies.

An 8-year-old girl presented to our clinic with abnormal eye movement since the age of 8 months. She was otherwise healthy. On examination, the patient’s best-corrected visual acuity was 20/20, with a refraction of -1.75 -2.0 × 180° in the right eye and 16/20 with a refraction of -1.0 -1.5 × 180° in the left eye. No abnormal head position or chin depression was observed. A 30 prism diopter (PD) exotropia was present in primary and near gazes and a 25 PD exotropia was present in distance gaze. She showed mild overaction of both inferior oblique muscles and moderate limitation of adduction in both eyes. On down gaze, a 10 PD exotropia was present that increased to 40 PD on up gaze. A significant V-pattern exodeviation of 40 PD exotropia on up gaze was present. The remaining ocular examination was entirely normal. A computed tomography (CT) scan of the orbits was performed before surgery, but all extraocular muscles appeared normal. Surgical correction with bilateral inferior oblique muscle myectomy, resection of the medial rectus muscle, and recession of the lateral rectus muscle of the left eye was planned.

Bilateral inferior oblique muscle myectomy was performed. The left medial rectus muscle showed thin fibrous tissue and was biopsied (Figure 1A). Medial rectus muscle exploration of the right eye was performed and showed marked fibrotic tissue (Figure 1B). A biopsy of the right medial rectus muscle was also performed. After the biopsies, bilateral lateral rectus muscle recession was performed. The lateral rectus muscles were visible and appeared to be normal. The biopsy results of both medial rectus muscles showed fibrous tissues with scattered vascularity and an absence of muscle fiber (Figures 1C and 1D). After surgery, the patient was orthophoric in primary gaze with glasses, and the V-pattern was greatly diminished. She continued to do well for 3 years after the surgery.

Orbital CT or magnetic resonance imaging can
aid in the diagnosis of congenitally absent extraocular muscles before surgery and can allow for effective surgical planning to correct the congenital abnormality. However, in our patient, although all of the extraocular muscles appeared normal throughout the coronal orbital CT scans, bilateral medial rectus muscle aplasia was found intraoperatively. The unusual findings in the presented case suggest that developmental abnormalities of the extraocular muscles should be considered in the differential diagnosis for constant exotropia.

REFERENCES

Dong-Wook Lee, MD
Sam Lee, MD
Min Ahn, MD, PhD
Jeonbuk, South Korea

The authors have no financial or proprietary interest in the materials presented herein.

doi: 10.3928/01913913-20130416-01