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Accessory fibrotic lateral rectus muscles in exotropic Duane syndrome with severe retraction and upshoot

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Article Summary

A 3.5-year-old boy presented at the University of California Los Angeles–Olive View Hospital with abnormal eye movements present since birth. His parents noticed that his left eye always appeared smaller than his right eye and that he had a right head turn. On examination, the patient demonstrated a 5° right head turn. In forced primary position, he had an exotropia of 6°. He had –2 limitation to adduction and abduction and +3 overelevation in adduction (upshoot) in the left eye.

The decision was made to perform surgery consisting of a lateral rectus recession with Y-splitting for presumed exotropic Duane syndrome with a significant upshoot. Preoperative forced duction testing revealed moderate restriction to adduction and no restriction to abduction. The surgical video demonstrates the presence of 2 accessory bands posterior to the lateral rectus muscle. All 3 structures (the lateral rectus and the 2 accessory bands) were recessed to 17 mm from the limbus in a V-shaped configuration (Figures 1 and 2). At postoperative month 1, the patient was orthotropic in primary position, and his upshoot had resolved. His adduction improved to –1 but his abduction worsened to –3.

First reported in 1893 by Nussbaum,¹ accessory muscles have been well described in disorders such as Duane syndrome and congenital fibrosis of the extraocular muscles.² They should be suspected in patients with atypical patterns of restrictive strabismus, globe retraction in directions other than adduction, or in patients with severe up- or downshoots. Lueder² described 3 types of accessory bands in his 2002 review: (1) structures arising from extraocular muscles and inserting in abnormal locations, (2) fibrous bands located beneath the rectus muscles, and (3) discrete anomalous structures in the posterior orbit. In a study of 453 strabismic subjects, Khitri and Demer³ demonstrated anomalous bands in 2.4% of subjects evaluated by magnetic resonance imaging.

Histopathological evidence implicates that these bands are made up of extraocular muscle tissue;⁴ however, pathological specimens are rare, because the bands are often difficult to

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access and not necessarily removed as part of routine treatment. In our case, no pathological specimen was obtained; however, the bands grossly resembled fibrous tissue with scattered muscle fibers. We theorize, therefore, that they are a combination of Lueder's types (1) and (2), that is, a fibrous band with some features of extraocular muscle.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

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Figure 1.

Figure 2.