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Increased restriction from an accessory lateral rectus in exotropic Duane syndrome

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Supernumerary extraocular muscles can cause restrictive strabismus, unusual ocular movements, and a persistent positive forced duction test. Even among patients with clinically typical strabismus, intraoperative testing and surgical exploration may reveal the presence of supernumerary extraocular muscles. We report the case of a patient with exotropic Duane syndrome found intraoperatively to have an accessory lateral rectus muscle, with histopathologically confirmed striated fibers.

Case Report

An otherwise healthy 11-year-old girl presented at the Hospital Federal dos Servidores do Estado, Rio de Janeiro, Brazil, with right head turn and abnormal left eye movements since birth. The family denied any previous ocular surgery. On ophthalmological examination, her visual acuity was 20/20 in each eye, and there was a 15° right head turn, in which stereopsis was 40 arcsec. Motility examination revealed −3 limitation of adduction and −3 limitation of abduction of the left eye as well as a great retraction of the bulbi in adduction and intense up- and downshoot (Figure 1). In forced primary position, she had a left exotropia of 30$^{\Delta}$. She was diagnosed with exotropic Duane syndrome in the left eye. Periosteal fixation of the lateral rectus muscle in the orbital wall was planned to try to eliminate or alleviate all of her anomalies—retraction, exotropia, anomalous head position, up- and downshoot—in a single surgery.

Intraoperatively there was no sign of a previous surgery, and forced duction testing still revealed a mild restriction to adduction of the left eye after lateral rectus disinsertion. We explored further and found an accessory muscle 4 mm behind to the original insertion of the lateral rectus muscle that continued far posterior to the globe (Figure 2). The muscle was extirpated and sent for histopathological analysis, which showed striated muscle fibers (Figure 3). After removal of the accessory muscle, forced duction testing was negative.

At postoperative month 3 (Figure 4) the patient was orthotropic, without anomalous head position or significant retraction, there was full adduction, no up- or downshoot, but the abduction remained very weak. At final follow-up, 1 year postoperatively, the patient remained orthotropic with no head turn or up- or downshoot.

Discussion

Supernumerary muscles can cause anomalous attachments between structures of the eye and the orbit. According to Duke-Elder,[1] anatomical abnormalities of extraocular muscles are relatively common. Khriti and Demer[2] found that 2.4% of the population had structures consistent with supernumerary extraocular muscles on magnetic resonance imaging. There are few reported cases of accessory muscles in Duane syndrome.[2-5] To our knowledge, this is the first report of an accessory lateral rectus muscle confirmed by histopathological examination in a patient with exotropic Duane syndrome.

Orbital imaging examination are indicated in patients with unusual ocular movements to investigate anomalous structures.[2,3,5] Because our patient had a typical presentation of exotropic Duane syndrome, we did not perform MRI before the surgery.

Intraoperative forced duction testing is particularly important, given the restrictive nature of anomalous structures.[2,3] Lueder[3] suggests that the persistence of restriction after disinsertion of a rectus muscle is usually a clue to the presence of an anomalous muscle. Accordingly, in our patient forced duction testing showed continued limitation after disinsertion of the the lateral rectus muscle, and thus we continued surgical exploration and discovered an anomalous structure adherent to the sclera. After its extraction, we could finally move the eye freely. The patient became orthotropic, with neither anomalous head position nor significant ocular retraction or up- or downshoot. Other authors have also described similar favorable outcomes.[3-8]

Lueder[3] classified anomalous orbital structures into three types. The first includes structures arising from the extraocular muscle and inserting in abnormal locations. The second includes abnormal fibrous bands located under the extraocular muscle. The third type arises in the posterior orbit and inserts on the globe or on the extraocular muscle. Pineles and Velez[4] presented a similar case of exotropic Duane with two accessory bands posterior to the lateral rectus muscle that resembled fibrous tissue with scattered muscle fibers as a combination of Lueder's first two types. The accessory muscle described in our case resembles Lueder's third type, because its insertion was

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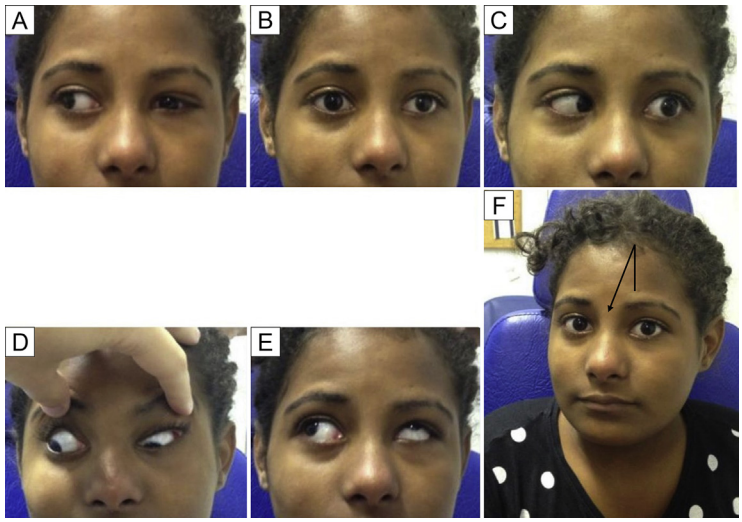

FIG 1. A, In the left eye there is adduction restriction and globe retraction in attempted adduction. B, Exotropia in primary ocular position. C, Left abduction restriction. D, Left downshoot. E, Left upshoot. F, Anomalous head position (15° right head turn).

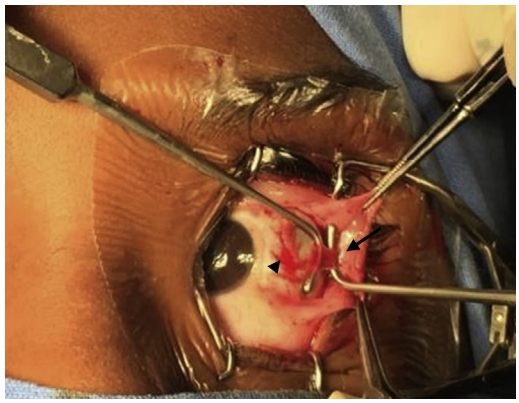

FIG 2. Surgical view of the accessory muscle (arrow), found 4 mm posterior to the original insertion of the lateral rectus muscle (arrowhead).

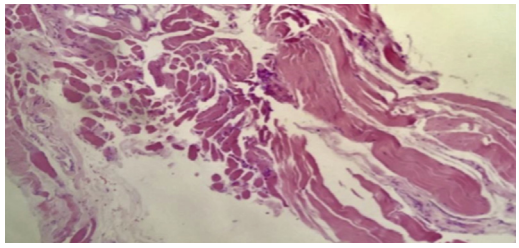

FIG 3. Cross section of the excised extraocular structure showing striated muscle fibers (hematoxylin-eosin, original magnification ×40).

behind the lateral rectus muscle, and it continued far posterior to the globe's equator. The histopathological analysis confirmed that the structure consisted of striated muscle fiber tissue.

The origin the anomalous extraocular muscles remains unknown. It is suggested that they could be atavistic remnants of the retractor bulbi muscle found in inferior mammals[9] or errors of cleavage in the development of mesoderm.[1] In addition, accessory extraocular muscles might be atrophied residual orbital structures that were not innervated or lost innervation during development.[2]

FIG 4. Clinical photograph of patient 3 months after surgery, when the patient was orthotropic and had no head turn.

Exotropic Duane syndrome is a characteristic form of restrictive strabismus. In our case, although the expected abduction remained very weak, removal of the accessory muscle extraction was fundamental to the excellent postoperative result.

Literature Search

PubMed was searched on April 7, 2017, without date or language restriction, using the following terms: *supernumerary*

OR *accessory* OR *anomalous* AND *extraocular muscle*; *Duane syndrome* AND *accessory* AND *extraocular muscle*; *accessory* AND *lateral rectus* AND *Duane syndrome*; *accessory* AND *lateral rectus* AND *type 2* OR *exotropic* AND *Duane syndrome*.

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Ophthalmic diagnosis and optical coherence tomography of abetalipoproteinemia, a treatable form of pediatric retinal dystrophy

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A 16-year-old boy with early-childhood-onset retinal dystrophy and developmental delay was diagnosed with abetalipoproteinemia based on ophthalmic examination, history, and results of a peripheral blood smear. The diagnosis was confirmed by lipid profile and genetic testing, and an older sister was confirmed to be affected as well. Although abetalipoproteinemia is treatable in early childhood, most cases are diagnosed late if at all. We highlight clinical features that should raise suspicion for this treatable but likely under-diagnosed form of early-onset retinal dystrophy and document retinal optical coherence tomography findings for a genetically proven case.

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Case Report

A 16-year-old boy was evaluated at King Khaled Eye Specialist Hospital for poor vision since early childhood. His 19-year-old sister was similarly affected, whereas his 3 eldest siblings (a brother, a sister, and a brother, in order of decreasing age) and a younger sister were asymptomatic. The boy's parents were from the same tribe. Poor vision had been noted since early childhood, particularly at night, and the parents were unsure whether it had been progressive. Review of systems revealed that both affected siblings had been "weaker" and delayed in their milestones compared to their siblings. Both affected siblings had not attended school because of perceived intellectual disability. Prior evaluation included brain magnetic resonance imaging, which was normal by report. Further questioning revealed that both siblings had problems with diarrhea after eating certain foods, particularly meat which they thus avoided. There was no history for polydactyly, labored breathing after birth, difficulty hearing, or cardiac disease.

The boy was under average for height and weight (153 cm and 45.60 kg). On ophthalmological examination, best-corrected visual acuity was 20/300 in each eye. There was pendular horizontal nystagmus and poor tracking. Anterior segment examination was normal. Retinal examination revealed a dystrophic retina, with frank central macular atrophy and overlying central gliosis. Optical coherence tomography revealed retinal thinning, particularly in the central macula. Autofluorescence showed an increased signal in the central macula. Electroretinography revealed cone-rod dysfunction.

Abetalipoproteinemia was suspected because of the atypical retinal dystrophy (with early central involvement) in the context of developmental delay and meat intolerance. A peripheral blood smear confirmed that the boy had marked acanthocytosis. Retinal findings and results of the blood smear are shown in Figure 1. Lipid profiles in millimoles/liter showed absent plasma triglyceride level (0.0 [normal, 5.2]) and decreased cholesterol profile (total cholesterol of 1.0 [normal 1.3]), with low-density lipoprotein (LDL) virtually absent (LDL of 0.1, HDL of 0.9). The affected sister had very similar ophthalmic findings and laboratory profile, and in addition to