Orbital bands are anomalous muscle tissue connections between recti muscles or between a rectus muscle and the globe. Also known as supernumerary extraocular muscles, they have been identified by magnetic resonance imaging in 0.8% of orthotropic and 2.4% of strabismic patients.1 Gomez-Lopez-Hernandez syndrome (OMIM 601853) is a rare sporadic syndrome reported in only 27 patients worldwide to date.2 This syndrome, also known as cerebellotrigeminal dermal dysplasia, has not received any attention in the ophthalmic literature. The characteristic triad is rhombencephalosynapsis, trigeminal anesthesia, and bilateral alopecia in the parietal or parieto-occipital regions. Additional features are listed in the Table.

Report of a Case. A 9-month-old girl was referred for assessment of strabismus with abnormal head posture. She was the first child of nonconsanguineous parents and was delivered at full term. There was no family history of strabismus. Suspicion of gross motor delay had been raised. Her fixation was central, steady, and maintained in each eye. Teller card visual acuity was estimated at 20/960 OU, improving to 20/94 OU at the next visit with full correction of refractive error (+2.50 + 1.00 × 75° OD, +2.50 + 1.50 × 125° OS) determined by cycloplegic refraction. She adopted an abnormal head position with 20° chin elevation and 30° right head tilt. Esotropia (ET) with an A pattern was detected: 30 prism diopters (Δ) ET in the primary position, 40Δ ET in upgaze, and 14Δ ET in downgaze. There was no evidence of superior oblique overaction in either eye. Bilateral incyclotorsion was noted on funduscopy. With these findings and possible facial dysmorphism (Figure 1), a computed tomographic scan was ordered to assess the orientation of the orbits and to exclude craniosynostosis. Thin bands connecting the lateral aspects of the superior and inferior recti bilaterally were noted. Magnetic resonance imaging clearly delineated these bands, which had signal characteristic of extraocular muscle (Figure 2A-C). The orbits were incyclorotated as determined by a plane joining the medial and lateral recti (Figure 2A). The cranial sutures appeared normal. The cerebellum showed fusion of the cerebellar hemispheres (rhombencephalosynapsis) characteristic of Gomez-Lopez-Hernandez syndrome (Figure 2D).

Further examination, including evaluation by a medical geneticist, confirmed the presence of all features listed in the Table with the exception of corneal opacities.

The patient subsequently underwent 4.5-mm medial rectus recessions with full tendon width upshift for the A pattern. The initial postoperative result was microtropia but with persistence of 15Δ of A pattern. Three years later, she underwent 5-mm lateral rectus recessions for 20Δ exotropia plus two-thirds tendon width downshift. One year later, she has 20Δ ET with 10Δ of A pattern. Fundus examination still shows moderate bilateral incyclotorsion.
Comment. High-resolution magnetic resonance imaging of the orbits, aided particularly by the use of surface coil techniques, allows detection of anomalous attachments between muscles or to the globe, previously detectable only during postmortem examination or occasionally intraoperatively. These structures can be an incidental finding or a cause of restrictive strabismus. The imaging findings alone are not a reliable predictor of the strabismus pattern. The case described here of an A-pattern ET with thin bands connecting the lateral borders of the superior and inferior recti differs from the cases reported by Khitri and Demer showing the same band anatomy in the setting of a V-pattern ET, right hypertropia, or orthophoria with elevation deficit. In our case, the contribution of the anomalous bands to the strabismus pattern cannot be apportioned from the rotated axis of the horizontal recti. A-pattern strabismus has been reported in association with rhombencephalosynapsis in 3 patients who did not have focused imaging of their orbits.

Most orbital bands cannot be accessed during conventional strabismus surgery, but there have been reports of treatment. This case highlights the difficulties in obtaining stable eye alignment postoperatively when orbital bands possibly interfere with normal globe rotations.

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