Restrictive myopic myopathy: computed tomography, magnetic resonance imaging, echography, and histological findings

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Restrictive ocular motility disturbances associated with high myopia (myopic myopathy) is a rarely seen phenomenon. Though the exact aetiology of myopic myopathy (MM) is not known, progressive neurogenic palsy,^{1,2} structural changes of muscles,³ paralysis,⁴ and myositis⁵ are suggested as the cause of MM.

A case of MM with computed tomography (CT), magnetic resonance imaging (MRI), echographic findings, and histological examination of medial recti is reported.

Case report

A 67-year-old man presented with strabismus for 6 years. He had a history of lens extraction and intraocular lens implantation 1 year previously in the left eye and cataract in the right eye. Best corrected visual acuity was hand movements in the right eye and 1/10 in the left eye.

Ophthalmoscopy showed annular conus, chorioretinal atrophy, and posterior staphyloma in the left eye. Axial lengths of the eyes were 32.7 mm and 33.9 mm in the right and left eyes respectively. The patient exhibited 30 ΔD esotropia with bilateral limitation of abduction more marked in the left eye (Fig 1).

Axial CT and MRI scan of the orbits showed significant elongation and enlargement of both eyes. Both lateral recti were extremely thin and there was almost no space between the eyeballs and anterior parts of lateral orbital walls (Fig 2). Examination of thyroid gland and thyroid tests were normal.

With echography, flattening of the postero-

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Figure 1 Positions of the eyes in three patients with myopic myopathy in all fields of gaze showing bilateral limitation of abduction, more marked in the left eye.

Fig 2A

Fig 2B

Fig 2C

Figure 2 Computed tomography scans of the orbits. (A) Laevoversion. (B) Primary position. (C) Dextroversion.

lateral part of the globe and angulation was seen during adduction. Histological examination showed fibrosis and minimal atrophy of the medial rectus.

Comment

Hugonnier and Magnard⁵ claimed that the aetiology of MM is myositis. Our patient showed limited abduction and positive forced duction test as in Hugonnier and Magnard's cases, but in addition there was atrophy and fibrosis of medial rectus muscle fibres by histological examination. Knapp⁶ also reported the same ocular motility abnormalities in patients with high myopia without histological abnormalities. Zolog⁷ found disappearance of muscular fibres of lateral recti in two similar patients.

Bagolini and coworkers⁴ claimed that pressure of the large globe on the lateral rectus results in paralysis of the muscle with subsequent esotropia in six patients with MM. They stated that in addition to paralysis of the lateral rectus, direct pressure on the muscle or capillary bed may play an important role in the aetiology. Instead of a marked forced duction test at adduction, they found only a slightly increased cytoplasmic component of muscle fibres and moderate hypertrophy on histological examination of the medial rectus; but we found substantially increased connective tissues and diminished dimension of myofibres.

Demer and Von Noorden⁸ reported a high myope patient with restrictive motility disturbance owing to the contact between the elongated globe and the medial wall of the orbit. But orbital CTs and MRIs of our patient demonstrated enough space at maximum abduction

between the medial rectus and the globe. Ruttum *et al*⁹ demonstrated flattening of the postero-medial wall of the globe against the medial orbital wall at abduction in a patient with MM. We have seen flattening of the posterolateral wall of the globe at adduction but not at abduction, so we do not think that the limitation of abduction is caused by contact between the globe and medial wall of the orbit.

In patients with myopia, the globe continues to enlarge during adulthood while the orbit ceases to grow. When the eye continues to grow, it begins to compress on the lateral rectus. As a result of this compression, lateral rectus becomes atrophic and esotropia occurs. Long standing esotropia may cause medial rectus fibrosis, in turn further increasing esotropia. In the later decades, atrophy of the soft orbital tissues may contribute to lateral rectus dysfunction by decreasing the protective effect of the muscle.

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Trisomy 4p and ocular defects

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Trisomy of the short arm of chromosome 4 is a relatively well studied pathology. At least 85 cases have been reported,^{1–12} 74 of them were reviewed by Kleczkowska *et al*.⁹ Despite the fact that most patients with trisomy 4p have no serious eye defects, some recent data suggest that some forms of ocular pathology may be relatively common for this condition.

Case report

A girl was born at term after the first pregnancy of healthy 22-year-old unrelated parents. Her

birth weight was 3200 g and length was 52 cm. Severe micropthalmos on the right and uveal tract coloboma on the left were mentioned in the delivery room. Further ophthalmic examination revealed coloboma of the iris, choroid, and retina. The right eye was enucleated and replaced by a prosthesis.

Examination at the age of 4 years and 4 months showed a relatively short girl (97 cm, just below the 5th percentile) with normal weight (17.2 kg, above 50th percentile) and head circumference. Her fine motor development and speech were delayed. She had brachycephaly, a large nose