Unusual Association Between Highly Myopic Strabismus And Orbital Tumor

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Citation

Abstract
We present a case of highly myopic strabismus in a 70-year-old woman. The right eye was fixed in a position of extreme adduction, and ocular motility was severely restricted in all directions. Magnetic resonance imaging demonstrated that the posterior globe appeared to be dislocated from the muscle cone in the right eye. An orbital tumor was detected incidentally. To our knowledge, there are no previous reports of highly myopic strabismus associated with orbital tumor. We speculate that the orbital tumor may have exacerbated the outward dislocation of the right globe.

INTRODUCTION
Axial high myopia sometimes gives rise to a particular type of strabismus, characterized by mechanical restriction in both abduction and sursumduction, resulting in esotropia and hypotropia. At the most advanced stage, the affected eye is so tightly fixed in an esotropic and hypotropic position that movement in any other direction is impossible. This condition has been called convergent strabismus fixus or myopic strabismus fixus. The posterior half of the globe is dislocated from the muscle cone. Recently, Yokoyama proposed the term “highly myopic strabismus” for this condition.1, 2 In this study, we present an unusual case of highly myopic strabismus associated with orbital tumor in a 70-year-old woman.

CASE REPORT
A 70-year-old woman with high myopia was referred to Jichi Medical University Hospital because of progressive esotropia of the right eye for the past year. The patient had no specific medical history. Best-corrected visual acuity was light perception and 1.2 in her right and left eyes, respectively. Slit-lamp examination showed cortical opacities in both lenses. Fundus examination showed myopic chorioretinal atrophy with staphyloma in the right eye; however, there was no specific abnormal finding in the left eye. The right eye was fixed in a position of extreme adduction, and ocular motility was severely restricted in all directions (Figure 1).

Figure 1
Ocular motility photographs of the patient in nine positions of gaze. The right eye was fixed in a position of extreme adduction, and ocular motility was severely restricted in all directions.

Ultrasonic B-scan echography (UD-8000, TOMEY, Nagoya, Japan) revealed staphyloma in the right eye (Figure 2a), but no abnormal findings were observed in the left eye (Figure 2b). The axial length was 29.5 mm in the right eye and 24.2 mm in the left eye.
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Figure 2
Ultrasonic B-scan echography shows staphyloma in the right eye (a). No abnormalities are evident in the left eye (b).

Magnetic resonance imaging (MRI) demonstrated that the posterior globe appeared to be dislocated from the muscle cone in the right eye (Figure 3). In addition, displacement of the lateral rectus muscle inferiorly was clearly visible. Furthermore, an irregularly shaped mass was detected in the right superonasal intra-orbital space.

Figure 3
Coronal T1-weighted MRI demonstrated that the globe appeared to be dislocated from the muscle cone. The lateral rectus muscle was displaced inferiorly. An irregularly shaped mass was detected in the superonasal intra-orbital space.

Axial T1-weighted images showed an isointense-signal tumor relative to muscle (Figure 4a). Axial T2-weighted images revealed that the tumor was mildly hyperintense relative to muscle (Figure 4b).

Figure 4a
MRI: (a) Axial T1-weighted image shows an oval isointense-signal tumor relative to muscle.

Post-contrast axial image using gadolinium-diethylenetriamine pentaacetic acid showed that the tumor enhanced heterogeneously (Figure 4c).

Figure 4b
MRI: (b) Axial T2-weighted image reveals that the tumor is mildly hyperintense relative to muscle.

Figure 4c
MRI: (c) Post-contrast axial image shows that the tumor enhances heterogeneously.

Based on these MRI findings, we speculated that the orbital tumor might be as schwannoma. We elected to follow-up the
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patient with observation alone because surgery was not possible. The reported findings did not change at any point during the 1-year follow-up period.

DISCUSSION

This case demonstrated an unusual association between highly myopic strabismus and orbital tumor. To our knowledge, there are no reports of highly myopic strabismus associated with orbital tumor.

Schwannomas are encapsulated, slowly progressive, benign proliferations of Schwann cells. 3

They are rarely found in the orbit and account for 0.7% to 2.3% of all histopathologically proven orbital tumors. 4, 5 Schwannomas most commonly arise from branches of the trigeminal nerve; however, they may arise from peripheral branches of the oculomotor, trochlear, and abducens nerves; parasympathetic and sympathetic fibers; and the ciliary ganglia. 3, 6-9

MR imaging features may be helpful in differentiating a schwannoma from other tumors.

Generally, schwannomas appear as isointense with respect to the extraocular muscle and cerebral gray matter on T1-weighted images and hyperintense on T2-weighted images. After the administration of intravenous contrast material, heterogeneous uptake is typically observed. 3, 4, 9 Based on these MRI findings, we speculated that this orbital tumor might be a schwannoma.

Interestingly, this patient demonstrated progressive esotropia for one year prior to presenting at our clinic. We speculate that the orbital tumor may have exacerbated outward dislocation of the right globe.

References

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