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CLINICAL INVESTIGATIONS

Treatment of A-pattern Esotropia with Marked Mongoloid Slanting Palpebral Fissures

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Background: The association of oblique palpebral fissures and A- or V-pattern has not been clarified. We report two cases of A-pattern esotropia with marked mongoloid slanting palpebral fissures associated with vertical displacement of the horizontal rectus muscle.

Cases: Case 1 was a boy with Prader-Willi syndrome. He showed A-pattern esotropia with upward slanting palpebral fissures. Severe superior oblique muscle overaction was observed. Case 2 was a girl with meningocele. She also showed A-pattern esotropia with upward slanting palpebral fissures.

Observations: In case 1, weakening surgery of the superior oblique muscles did not improve the A-pattern. Coronal images of computed tomography showed one-half-muscle-width upward displacement of both lateral rectus muscles. After downward transposition surgery of the lateral rectus muscles, the preoperative A-pattern of 25 prism diopters (PD) was successfully corrected to 10 PD. In case 2 also, upward displacement of both lateral rectus muscles was shown by computed tomography. The preoperative A-pattern of 26 PD was corrected to 4 PD postoperatively after upward transposition surgery of the medial rectus muscles.

Conclusions: The vertical displacement of horizontal rectus muscles was considered the principal cause of A-pattern in these cases associated with marked mongoloid slanting palpebral fissures. **Jpn J Ophthalmol 2001;45:482–486**

Key Words: A-pattern esotropia, coronal CT, mongoloid slanting palpebral fissure, vertical displacement, vertical transposition.

Introduction

A- and V-patterns introduced by Urist[1] are very common signs of great importance in strabismology. The etiology of A- and V-patterns includes horizontal rectus muscle overaction,[2] vertical rectus muscle underaction,[3] oblique muscle dysfunction,[4] imbalance of insertions of oblique muscles,[5] or structural orbital anomaly.[6,7] Patients with craniosynostosis, such as Crouzon, Apert, Pfeiffer syndromes and plagiocephaly, often show A- and V-patterns owing to the orbital anomalies.[8]

Obliquity of palpebral fissures was once considered to be related to A-, V-patterns. Mongoloid (upward) slanting of the palpebral fissures was considered to be associated with superior oblique overaction and A-pattern, and antimongoloid (downward) slanting of the palpebral fissures, to be associated with inferior oblique overaction and V-pattern.[9] However, another study[10] could not find any reliable association between A-, V-patterns, and oblique palpebral fissures.[11]

We report two cases of A-pattern esotropia with marked mongoloid slanting palpebral fissures with superior oblique overaction. In our cases, we could elucidate vertical displacement of horizontal rectus

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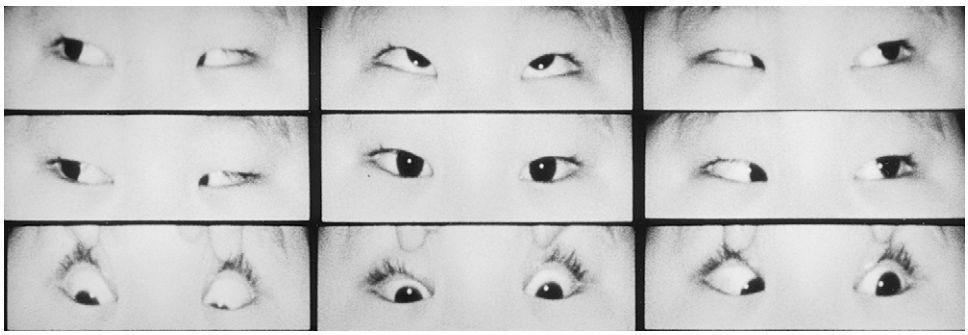

Figure 1. Preoperative status of A-pattern esotropia with bilateral +3 superior oblique overaction (case 1). Marked mongoloid slanting palpebral fissures can be seen.

muscles by coronal computed tomography (CT). Superior oblique weakening surgery did not improve A-pattern effectively. These cases were successfully treated with vertical transposition of horizontal rectus muscles. We describe herein the relationship between A-pattern esotropia with mongoloid slanting palpebral fissures and vertical displacement of horizontal rectus muscles.

Case Reports

Case 1

A 3-year-old boy was referred for evaluation of esotropia in December 1991. He had been followed-up for Prader-Willi syndrome in the Pediatric Department of our hospital. Corneal reflex test revealed A-pattern esotropia and superior oblique overaction in both eyes as well as marked mongoloid slanting palpebral fissures (20° upward slant) (Figure 1).

In July 1996, at the age of 8 years, the corrected visual acuity was 12/20 in the right eye and 18/20 in the left eye. The cycloplegic refraction was: OD:sph +1.0 D: cyl −3.0 D Ax180°; OS: cyl −1.5 D Ax180°. The alternate prism and cover test results were 60 PD esotropia (ET) with 5 PD left hypertropia on upward gaze, 45 PD ET with 4 PD left hypertropia in primary position and 35 PD ET with 3 PD left hypertropia on downward gaze (A-pattern of 25 PD). Version showed +3 overaction of each superior oblique muscle. Fundus examination revealed mild incyclotropia.

We performed surgery of 5.5-mm recession of the left medial rectus muscle and 7-mm resection of the left lateral rectus muscle with disinsertion of both superior oblique muscles. The postoperative alternate prism and cover test results showed 25 PD ET with 4 PD left hypertropia on upward gaze, 14 PD ET with 3 PD left hypertropia in primary position and 4 PD exotropia (XT) with 2 PD left hypertropia on downward gaze (A-pattern of 29 PD). The overaction of the superior oblique muscles still remained, and the A-pattern was not improved significantly (Figure 2). Coronal CT at the plane near the globe-optic nerve junction showed one-half-muscle-width upward displacement of both lateral rectus muscles. The orbits

Figure 2. Postoperative status of case 1 after bilateral superior oblique disinsertion and recession-resection of left eye. Overaction of superior oblique muscles still remained and A-pattern was not improved.

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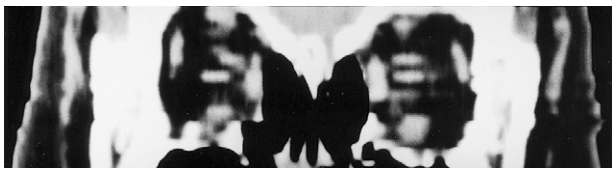

Figure 3. Case 1. Coronal computed tomography scan at plane near globe-optic nerve junction showing upward displacement of both lateral rectus muscles.

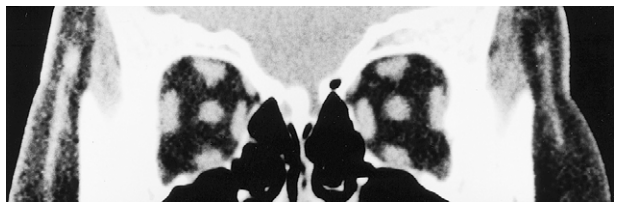

Figure 5. Case 1. Postoperative coronal computed tomography scan did not show much change in displacement of rectus muscles.

were not tilted and the trochleas were not displaced abnormally (Figure 3).

In January 1999, at the age of 11 years, the boy underwent a second surgery for residual esotropia and A-pattern. The second surgery performed was 3.5-mm resection of the right lateral rectus muscle with downward transposition of one-half-muscle-width of both lateral rectus muscles. Postoperative alternate prism and cover test showed 14 PD ET with 3 PD left hypertropia on upward gaze, 10 PD ET with 4 PD left hypertropia in primary position and 4 PD ET with 3 PD left hypertropia on downward gaze (A-pattern of 10 PD) (Figure 4). On version, there was +1 overaction of the left superior oblique muscle. Fundus photography of both eyes showed no significant torsional change before and after muscle transposition surgery (10° incyclotropia). Postoperative coronal CT scan showed little change of the rectus muscle displacement (Figure 5).

Case 2

In June 1996, a 7-month-old girl was referred for evaluation of esotropia. She had been followed up for meningocele in the Neurology Department of our hospital. The Krimsky test revealed 30 PD esotropia with marked mongoloid slanting palpebral fissures (20° upward slant). In June 1998, at the age of 31 months, preoperative evaluation was performed. The visual acuity with Teller acuity cards was 20/190 in the right eye and 20/190 in the left eye. The Krimsky test results were 45 PD ET on upward gaze, 30 PD ET in primary position and 14 PD ET on downward gaze (A-pattern of 26 PD) (Figure 6). Version showed +1 overaction of the right superior oblique muscle. Cyclodeviation and stereopsis were not detected by subjective examinations. Coronal CT at the plane near the globe-optic nerve junction showed downward displacement of both medial rectus muscles (Figure 7) The orbits were not tilted and the trochleas were not displaced abnormally

In June 1998, at the age of 32 months, she underwent surgery of 5.5-mm recession of both medial rectus muscles as well as upward transposition of one-half-muscle-width.

Postoperative results of alternate prism and cover test were 8 PD XT on upward gaze, 8 PD XT in primary position and 12 PD XT on downward gaze (A-pattern of 4 PD) (Figure 8). Postoperative coronal CT scan showed little change compared with preoperative scan (Figure 9).

Discussion

In these cases of A-pattern esotropia with mongoloid palpebral fissures, vertical displacement of horizontal rectus muscles was clarified by coronal

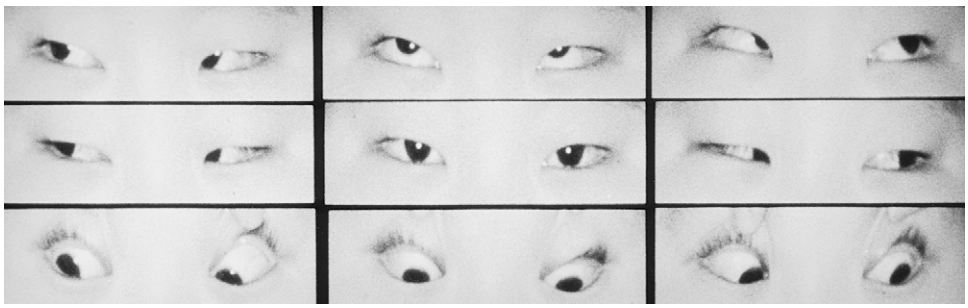

Figure 4. Postoperative status of case 1 after downward transposition of both lateral rectus muscles. A-pattern has disappeared.

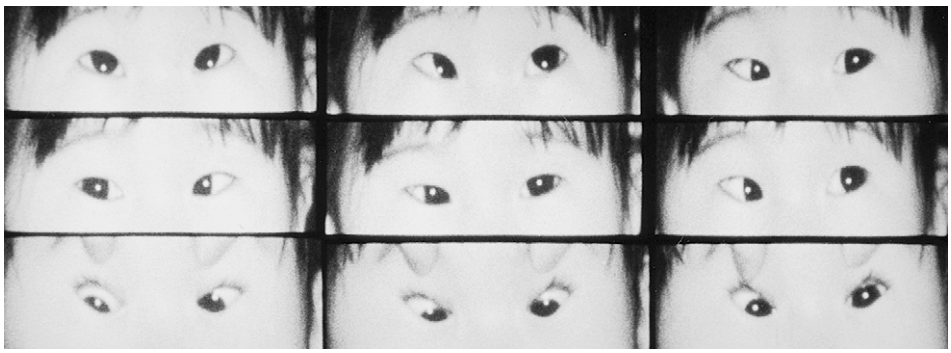

Figure 6. Preoperative status of A-pattern esotropia of case 2. Marked mongoloid slanting palpebral fissures can be seen.

CT, and A-pattern was effectively treated by vertical transposition of horizontal rectus muscles. Superior oblique weakening surgery in case 1 hardly improved A-pattern.

The association of palpebral fissures and A- or V-pattern has not been established.[10,11] Urrets-Zavalia et al[9] found that mongoloid slanting palpebral fissures tended to be associated with A-pattern and antimongoloid fissures with V-pattern. They found that mongoloid facial development consisted of well-developed malar bones, upward slanting palpebral fissures associated with A-pattern esotropia with overacting superior oblique muscles. They also found the association of antimongoloid facial features and V esotropia with overacting inferior oblique muscles.

Ruttum and von Noorden[10] could not find any relationship between facial characteristics and A-, V-exotropia. These studies suggested oblique muscle dysfunction might be related to A- and V-patterns in patients with characteristically slanting palpebral fissures.

The displacement of horizontal rectus muscles has not been discussed with regard to A- and V-patterns in association with mongoloid or antimongoloid slanting palpebral fissures. Clark et al[12] investigated the paths of extraocular muscles from 9 mm posterior to 6 mm anterior plane of the globe-optic nerve junction. They determined that there was less than 2-mm displacement of the horizontal and vertical rectus muscles around the orbital center in normal and strabismic patients with A- and V-patterns. In our 2 cases, we could obtain evidence of upward displaced lateral rectus muscle and downward displaced medial rectus muscle by coronal CT. As the superior oblique weakening surgery in case 1 failed to improve A-pattern esotropia, the overaction of superior oblique muscles was not considered to be the cause of the A-pattern. The displacement of the horizontal rectus muscles was considered the main cause of A-pattern in this esotropic case.

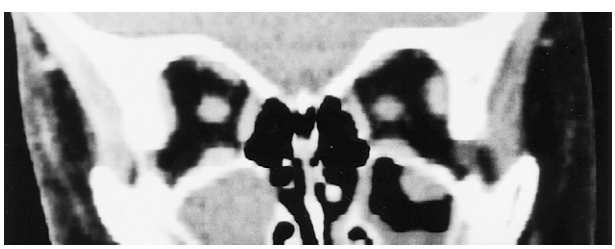

Figure 7. Case 2. Coronal computed tomography scan showing upward displacement of both lateral rectus muscles.

In a patient with meningocele, as our case 2, it was reported that A-pattern was usually associated with exotropia, superior oblique overaction and dissociated vertical deviation.[10] Although +1 superior oblique overaction was found in the right eye of our patient, surgery was not performed on the overacting superior oblique muscle, but on the displaced horizontal rectus muscles, and the A-pattern was successfully corrected by surgery. Overaction of superior oblique muscles in cases 1 and 2 are similar to the four cases of apparent A-pattern reported by Clark et al[12] with heterotopic muscle pulleys confirmed by magnetic resonance image. In those cases without mongoloid slanting palpebral fissures, the lateral rectus muscles deviated upward and the medial rectus muscles deviated downward.

The findings of coronal CT were extremely useful in determining the surgical procedures in these cases. The orbits were neither extorted nor intorted in both cases as Diamond et al[13] pointed out in a patient with plagiocephaly. The surgical amount of upward transposition of the medial rectus muscle or the downward transposition of the lateral rectus muscle in surgery was determined according to the amount of displacement of the images. There was little or no

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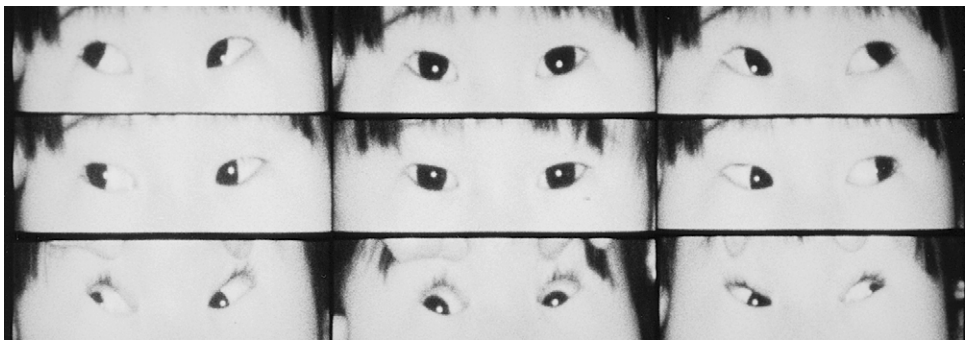

Figure 8. Postoperative status of case 2 after upward transposition of both medial rectus muscles. A-pattern has disappeared.

change of muscle displacement in coronal CT after transposition surgery because the rectus muscle bellies pass through pulleys fixed in the orbit.[14]

The surgical amounts of A-pattern correction in our cases was slightly larger than those of Ribeiro et al.[15] Urist proposed overaction of the medial rectus as the etiology of A-pattern and recommended recession of the medial rectus muscles with upward transposition.[16] We had the impression that the surgical correction of A-pattern would be equally effective whether the surgery was performed on the medial rectus or on the lateral rectus muscles.

In conclusion, the vertical displacement of the horizontal rectus muscles was considered the principal cause of A-pattern in these cases with marked mongoloid slanting palpebral fissures.

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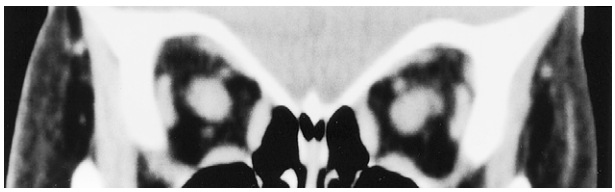

Figure 9. Case 2. Postoperative coronal computed tomography scan did not show much change in displacement of rectus muscles.