Acute acquired comitant esotropia

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SUMMARY Acute acquired comitant esotropia has been used to describe a dramatic onset of a relatively large angle of esotropia with diplopia and minimal refractive error. We describe six children aged 5 to 11 years who developed an acute non-accommodative esotropia with diplopia. Neurological examination, including CT scan, in each of these children gave negative results. We suggest that this is an unusual presentation of esotropia of undetermined aetiology. The diagnosis, clinical characteristics, and management are discussed.

Acute acquired comitant esotropia (AACE) is an unusual presentation of esotropia which occurs in older children and adults. It is characterised by acute onset of esotropia with diplopia. The refractive error is insignificant and neurological examination gives normal results. The purpose of this paper is to discuss the diagnosis and management of this unusual condition in children.

Patients and methods

We reviewed the records of patients aged 3 years or older who presented with the acute onset of diplopia and comitant esotropia from January 1984 to December 1987, at the paediatric ophthalmology services of Wills Eye Hospital, Albany Medical Center, Children’s Hospital of New Jersey, and Schneider Childrens’ Hospital. We excluded all patients who had a reduction in their esotropia with hyperopic spectacle correction.

Results

Of the 11 patients aged 3 or older who presented with acute onset of comitant esotropia and diplopia six met the above criteria for inclusion in this study. The patients ranged in age from 5 to 11 years, median 7 years. All complained of diplopia. In three patients the esotropia and diplopia were intermittent initially but became constant within a few weeks. The onset of esotropia was an average of six months prior to our examination, range one to 10 months. The esodeviation was comitant and ranged from 20 to 55 prism dioptres (Figs. 1, 2, 3). The near and distance measurements were equal. Visual acuity was 20/20 in all eyes. Ocular rotations, pupils and results of fundus examinations were normal. A cycloplegic retinoscopy under pentolate 1% or 2% was performed on each patient. The full cycloplegic refraction was prescribed for all patients, initially, with no change in the deviation noted (Table 1). None of the patients had a preceding infection or any neurological symptoms. In addition to the ophthalmic examination each child was examined by a paediatric neurologist and underwent CT scanning; all results were within normal limits. Other specific neurological tests in individual cases, as recommended by the paediatric neurologist, were completed. The negative results

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Fig. 1 Patient No. 6 in primary gaze.
Acute acquired comitant esotropia

Fig. 2 Patient No. 6 in right gaze, illustrating comitance of esotropia.

Fig. 3 Patient No. 6 in the left gaze, illustrating comitance of esotropia.

are listed in Table 1. All six patients underwent a bilateral medial rectus recession appropriate for the amount of esotropia on average eight months after initial presentation, range 3–24 months.

At follow-up ranging from three months to three years the angle has been found to have remained stable at orthophoria, except in one patient who developed a recurrent esotropia of 25 dioptres. This patient was previously described by Goldman and Nelson. The results of sensory tests are listed in Table 2.

Discussion

Previous case reports divide AACE into three types. Burian and Miller in 1958 reviewed the features of each type of AACE. Type I AACE, first described in 1947 by Swan, follows occlusion of one eye or loss of vision in one eye secondary to injury or disease. Von Noorden postulates that the esotropia is the result of interruption of a previously well functioning fusion mechanism, causing a latent deviation to become manifest. The course of this deviation may be variable, resulting in spontaneous resolution or becoming constant and requiring treatment.

Table 1 Clinical characteristics and neurological testing

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age</th>
<th>Sex</th>
<th>Refractive error</th>
<th>Neurological testing</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>10</td>
<td>M</td>
<td>Plano</td>
<td>MRI – ve</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Plano</td>
<td>Tensilon – ve</td>
</tr>
<tr>
<td>2</td>
<td>7</td>
<td>M</td>
<td>OD+1.75</td>
<td>CT – ve</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>OS+1.50</td>
<td>MRI – ve</td>
</tr>
<tr>
<td>3</td>
<td>6</td>
<td>M</td>
<td>OD+1.50</td>
<td>CT – ve</td>
</tr>
<tr>
<td>4</td>
<td>7</td>
<td>F</td>
<td>OD+1.75</td>
<td>Tensilon – ve</td>
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<tr>
<td></td>
<td></td>
<td></td>
<td>OS+1.00</td>
<td>CT – ve</td>
</tr>
<tr>
<td>5</td>
<td>11</td>
<td>F</td>
<td>OD+1.00</td>
<td>Tensilon – ve</td>
</tr>
<tr>
<td>6</td>
<td>10</td>
<td>M</td>
<td>OD+6.75</td>
<td>CT – ve</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>OS+6.00</td>
<td></td>
</tr>
</tbody>
</table>

Type II AACE, which is also termed the Franceschetti type, is characterised by an acute onset of a relatively large angle of comitant esotropia and diplopia. The refractive error is usually mildly hyperopic but does not influence the angle of deviation. There is no history of infection, and the results of neurological examination are negative. Burian reported on four patients aged six to 72 years. Their angle of esotropia ranged from 30 to 70 prism dioptres, and treatment with glasses did not improve the angle of esotropia. In one of Burian’s patients esotropia began while he was ill with pneumonia.

Type III AACE, reported by Bielschowsky in 1922, is characterised by the acute onset of esotropia in patients with uncorrected myopia of −5.00 dioptres or more, presumably following physical or mental stress. These patients were esotropic at distance but orthophoric at near vision. The mechanism was thought to be uncorrected myopia with excessive close work, leading to strengthening of the medial recti and weakening of the lateral recti. These patients have subsequently been excluded from the classification of AACE because the esotropia is present only at distance fixation. These patients are more appropriately classified as having divergence insufficiency.

Our patients had type II AACE with an acute onset of moderate to large angle comitant esotropia.

Table 2 Deviation and stereoacuity results

<table>
<thead>
<tr>
<th>Patient</th>
<th>Surgery after onset (months)</th>
<th>Angle (pre-op.)</th>
<th>Angle (post-op.)</th>
<th>Stereo (post-op.)</th>
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<tbody>
<tr>
<td>1</td>
<td>3</td>
<td>30 ET</td>
<td>Ortho</td>
<td>80 arc s</td>
</tr>
<tr>
<td>2</td>
<td>9</td>
<td>25 ET</td>
<td>Ortho</td>
<td>40 arc s</td>
</tr>
<tr>
<td>3</td>
<td>12</td>
<td>45 ET</td>
<td>25 ET</td>
<td>Unknown</td>
</tr>
<tr>
<td>4</td>
<td>12</td>
<td>45 ET</td>
<td>Ortho</td>
<td>40 arc s</td>
</tr>
<tr>
<td>5</td>
<td>6</td>
<td>55 ET</td>
<td>Ortho</td>
<td>40 arc s</td>
</tr>
<tr>
<td>6</td>
<td>9</td>
<td>35 ET</td>
<td>Ortho</td>
<td>40 arc s</td>
</tr>
</tbody>
</table>

ET=esotropia.
with diplopia. The refractive correction did not influence the angle of deviation. Unlike Burian's series, our patients did not have either preceding or coincidental illness. In all of them the results of neurological examination were normal.

Because of the concern about the possible development of suppression, these patients can be treated with base-out prisms to maintain binocular function until the angle of deviation stabilises. However, four of our six patients had 40 arc seconds of stereoacuity, which would eliminate the possibility of a previous monofixation syndrome that decompensated. The excellent stereopsis also showed that these patients did not develop suppression.

Type II AACE may be a dramatic and alarming condition. We recommend that patients who present with acute onset of diplopia and comitant esotropia receive a complete ophthalmologic and neurological examination and a careful history taken to rule out cyclic esotropia, divergence insufficiency, paretic strabismus, and myasthenia gravis, any of which may also present acutely. Whether further examination including CT scanning, should be performed is still not clear. In our series the results of all neurological tests were negative, but we believe that, owing to the relatively small size of our series, a larger number of patients must be studied before a recommendation to eliminate neurological testing is made. Since invasive studies (such as Tensilon testing and neuroradiologic testing) in children carry some risk, this is an important question to answer. We also suggest that the term AACE type II be reserved for patients with acute onset of diplopia and comitant esotropia unrelated to refractive error and of unknown aetiology.

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