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Atlando-axial rotary fixation

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Objectives. To analyse the causes of atlanto-axial rotary fixation (AARF) and discuss the diagnosis and treatment.

Design. Retrospective case studies.

Setting. Medical University of Southern Africa — Ga-Rankuwa referral hospital.

Patients. A total of 10 patients admitted to and treated in the Department of Orthopaedics, Ga-Rankuwa Hospital, between July 1989 and June 1993.

Outcome measure. Dynamic computed tomography (CT) scan.

Results. Upper respiratory tract infection and trauma were the commonest causes of AARF. There was a delay in diagnosis ranging between 4 weeks and 2 years 6 months. Clinical and radiological reduction was obtained by gradual skeletal traction in 6 patients. Two patients had improvement of the torticollis but still had subluxation on the CT scan. In 1 patient no reduction was obtained on occipitocervical fusion and transoral decompression was necessary. In 1 case the parents refused any form of treatment. There was no recurrence in the 7 patients followed up (minimum 6 months).

Conclusion. This study shows that AARF is often diagnosed late. The patients diagnosed early responded well to skeletal traction followed by external support. In patients diagnosed late the AARF could not be reduced completely and needed surgical fusion. If untreated, condition can be complicated by tetraparesis.


Torticollis is not a uncommon condition in children. There are many causes, including soft tissue pathology, bone pathology and intradural pathology. Atlanto-axial rotary subluxation is one of the commonest causes of torticollis. Many of these subluxations are easily correctable. However, a few may be difficult to reduce. Fielding and Hawkins coined the term 'atlanto-axial rotary fixation' (AARF) for those subluxations that are difficult to reduce.

The purpose of this paper is to review this condition in our patients and to emphasise early diagnosis and proper management to prevent late complications.

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Method

A retrospective study of patients seen at our hospital between July 1989 and June 1993 was undertaken. There were 10 children with the diagnosis of AARF during that period (Table I). Their ages ranged from 4 to 13 years with an average of 7 years 6 months. There were 6 girls and 4 boys. Their clinical presentation is shown in Table II. Six patients presented with neck pain. All patients presented with torticollis and diminished range of movement. Patient 5 presented with progressive tetraparesis over a period of 1 year. On examination their heads were tilted, flexed and rotated to one side like that of a cock robin. None of the patients had facial asymmetry or associated fracture of the clavicle. Five had spasm of the sternomastoid muscle on the opposite side of the torticollis.

The causes of AARF in our patients are shown in Fig. 1. In 4 patients the AARF followed upper respiratory tract infection (Grisel's syndrome). Three were due to trauma. One patient had juvenile rheumatoid arthritis. In 1 child, the AARF followed treatment of idiopathic scoliosis with an ill-fitting Milwaukee brace. In 1 child the cause was unknown.

Table I. Patients with AARF seen at Ga-Rankuwa Hospital between July 1989 and June 1993

<table>
<thead>
<tr>
<th>Patient no.</th>
<th>Age (yrs)</th>
<th>Sex</th>
<th>Aetiology</th>
<th>Delayed diagnosis</th>
<th>Hawkins type</th>
<th>Final treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>13</td>
<td>F</td>
<td>Ill-fitting Milwaukee brace</td>
<td>16 wks</td>
<td>I</td>
<td>Halo traction and C1C2 fusion</td>
</tr>
<tr>
<td>2</td>
<td>7</td>
<td>F</td>
<td>Grisel (URTI)</td>
<td>12 wks</td>
<td>II</td>
<td>Halo traction and C1C2 fusion</td>
</tr>
<tr>
<td>3</td>
<td>5</td>
<td>F</td>
<td>Grisel (URTI)</td>
<td>14 wks</td>
<td>II</td>
<td>Halo traction and C1C2 fusion</td>
</tr>
<tr>
<td>4</td>
<td>6</td>
<td>F</td>
<td>Grisel (URTI)</td>
<td>12 wks</td>
<td>II</td>
<td>Halo traction and C1C2 fusion</td>
</tr>
<tr>
<td>5</td>
<td>13</td>
<td>F</td>
<td>Trauma</td>
<td>2 yrs</td>
<td>III</td>
<td>Transoral decompression and occipito-C2 fusion</td>
</tr>
<tr>
<td>6</td>
<td>4</td>
<td>M</td>
<td>Grisel (URTI)</td>
<td>4 wks</td>
<td>I</td>
<td>Halo traction and Minerva jacket</td>
</tr>
<tr>
<td>7</td>
<td>6</td>
<td>M</td>
<td>Trauma</td>
<td>4 wks</td>
<td>I</td>
<td>Halo traction and Minerva jacket</td>
</tr>
<tr>
<td>8</td>
<td>8</td>
<td>F</td>
<td>Trauma</td>
<td>5 wks</td>
<td>I</td>
<td>Halo traction and hard collar</td>
</tr>
<tr>
<td>9</td>
<td>9</td>
<td>M</td>
<td>Rheumatoid arthritis</td>
<td>21/2 yrs</td>
<td>II</td>
<td>Parents refused any treatment</td>
</tr>
<tr>
<td>10</td>
<td>7</td>
<td>M</td>
<td>Unknown</td>
<td>6 wks</td>
<td>I</td>
<td>Halo traction and Minerva jacket</td>
</tr>
</tbody>
</table>

The duration of symptoms before diagnosis ranged from 4 weeks to 2 years 6 months (Table I). Seven of the patients had some form of treatment before the definitive diagnosis of AARF was made. The treatments given were physiotherapy, soft-collar and halter traction.

Radiographs were often difficult to interpret as it was difficult to obtain good open-mouth views. For that reason, a computed tomography (CT) scan was requested in all patients to confirm the diagnosis and classify the condition according to Fielding and Hawkins. In type I disease, there is rotation of the atlas over the axis with an intact transverse ligament and no subluxation. In type II there is unilateral anterior displacement of the lateral mass with the opposite joint acting as a pivot and an atlanto-dens interval (ADI) of 3 - 5 mm. In type III there is bilateral anterior displacement of the lateral masses with an ADI of more than 5 mm. In type IV there is posterior subluxation of the atlas. Of our patients, 5 were type I, 4 type II and 1 type III (Table I).

Magnetic resonance imaging was only undertaken in patient 5, who presented with tetraparesis (Fig. 2). All patients were initially treated with halter neck traction for 5 days. If correction of the torticollis was not achieved the diagnosis of AARF was considered. A dynamic CT scan was then done to confirm the diagnosis and a halo ring was applied for correction. The traction weights were gradually increased by 0.5 kg every 5 days from 2 kg to a maximum of 6 kg, depending on the size of the child and the rate of correction.

In 4 patients who presented with symptoms of less than 3 months' duration, the reduction was maintained with an external support for a total of 12 weeks. In another 4 patients, whose symptoms were of more than 3 months' duration, the reduction was maintained with an external support for a total of 12 weeks. In another 4 patients, whose symptoms were of more than 3 months' duration, the reduction was maintained with an external support for a total of 12 weeks.
duration, an atlanto-axial fusion was performed. The child with tetraparesis and Hawkins type III AARF did not improve after the 6 weeks of halo-ring traction. A one-stage transoral decompression with a microscope, followed by an occipitocervical fusion, was performed.

The parents of 1 child, who had severe rheumatoid arthritis, refused any form of treatment.

Results

Clinical and radiological reduction of the AARF was obtained in 5 patients with Hawkins type I AARF and in 1 patient with type II. In 2 patients, clinical reduction of the torticollis was obtained but CT scans at 7 weeks of traction showed incomplete correction. Seven patients were followed up for between 6 months and 4 years. Two were lost to follow-up (patients 9 and 10). Patient 5, who underwent transoral decompression and occipitocervical fusion, experienced neurological improvement from Frankel S to a Frankel C postoperatively. Unfortunately she died 4 weeks postoperatively following obstruction of the tracheostomy tube that had been inserted as part of the transoral approach to the ClC2 complex.

None of the 7 patients followed up experienced recurrence of symptoms. The children who underwent atlanto-axial fusion only had a slight limitation of lateral rotation.

Discussion

The description of atlanto-axial subluxation associated with pharyngeal pathology was credited to Bell in his 1830 report. Since then case reports have been reported by several authors. Grisel described his cases in 1930 and the condition was subsequently named after him. Atlanto-axial rotatory fixation was first described in detail by Corner in 1907. There have been many case reports since. Several anatomical studies have helped us to understand the pathology and pathogenesis of AARF. Werne, among others, gave a detailed study of the major role of occipito-atlanto-axial ligaments in the stabilisation of the atlanto-axial joint. Coutts noted that with an intact transverse ligament, atlanto-axial rotation of 65° resulted in narrowing of the spinal canal at this level to 7 mm. He also noted that in a deficient transverse ligament with 5 mm ADI, rotation of 45° resulted in narrowing of the spinal canal to 12 mm.

In 1984, Parke et al. published a study describing the pharyngovertebral veins which, for the first time, explained how inflammation spread from the retropharynx to the atlanto-axial ligaments in Grisel's syndrome.

The pathogenesis of AARF in Grisel's syndrome is controversial. Wittek postulated that effusion of the atlanto-axial joint led to laxity of the capsule and transverse ligament, which allowed the mass of the atlas to displace anteriorly. Greig and Watson-Jones proposed that regional hyperaemia led to ligamentous laxity and decalcification of the attachment of the transverse ligament, with subluxation of the atlas as the final result. Grisel hypothesised that paravertebral and neck muscles went into spasm in an attempt to decompress the inflamed joints. This resulted in the subluxation of the atlanto-axial joints.

Where trauma was the cause of the AARF, the mechanism was forced flexion with rotation, as in a fall on the shoulder and the side of the head. Goddard found a high incidence of associated fracture of the clavicle in their patients who had AARF due to trauma. We did not have this experience.

There are several reasons why AARF is difficult to reduce. Blockage of the reduction by synovial fringes in the inflamed joint is one reason that has been put forward. Fielding and Hawkins postulated that in the late stage, ligamentous capsular contracture was the main reason for the difficulty in reduction. In cases of trauma, associated fracture of the articular surfaces has been advanced as a reason for the difficulty in reduction of the atlanto-axial displacement.

One Keir et al. postulated that in the quiescent stage, failure to reduce the AARF was the result of dislocation of the occiput over the atlas while the atlas was still dislocated over the axis. The occipito-atlantal dislocation was the result of a compensatory rotation of the head to reverse the initial atlanto-axial subluxation. The displaced atlas thus became fixed between the occiput and the axis. They called this pseudo-reduction.

Seventy-five to eighty per cent of patients with AARF reported in the literature were children aged under 13 years. In this study all patients were under 13 years.

As in the literature there was a delay in the diagnosis of this condition. Torticollis that does not resolve within 5 - 7 days of soft-collar or halter traction treatment should raise suspicion of AARF. Other clinical features are: (i) restriction of neck movements; (ii) spasm of the sternomastoid muscle on the contralateral side of the head; and (iii) the spinous process of the axis is prominent and palpable at the opposite side of the dislocation (Sudek's sign). A thorough ear, nose and throat examination should also be done.

Wortzman and Dewar described features of AARF on radiographs but these are difficult to interpret as it is difficult to get good open-mouth views in this condition. Dynamic CT scan is the examination of choice to confirm the diagnosis. In this study the CT scan was taken with the head first in neutral position, then rotated maximally one way and the other. Magnetic resonance imaging must be done where there is a neurological fall-out. Fielding and Hawkins gave a helpful classification of this condition after reviewing their 17 cases. We used this classification in our study as we believed it defined the pathology better.

Unfortunately the natural history of AARF is not fully known. Patients have been reported to develop facial asymmetry if left unattended. Neurological complications seem to be rare, possibly because of the large diameter of the spinal canal at this region. However, one of Fielding and Hawkins' patients died of spinal cord transection by the atlas. Watson-Jones also described a patient who died as a consequence of spinal cord transection by the atlas. It would seem that while AARF may be difficult to reduce the chances of it progressing are there. It is therefore important to reduce the AARF and obtain stability to prevent complications. The difficulty in obtaining the reduction was directly related to the delay before the final diagnosis was...
made. Manipulation of patients with AARF can be very dangerous and should not be attempted. At our clinic we started with halter traction for 5 days to exclude the easily reducible atlanto-axial subluxation. When no reduction was obtained, the diagnosis of AARF was confirmed with a CT scan and halo-ring traction was applied for the gradual reduction of the AARF.

In patients whose symptoms are of less than 3 months' duration we maintained the reduction by external support for a minimum of 6 weeks. In patients in whom complete radiological reduction failed, patients who presented with neurological fall-out, patients with symptoms of more than 3 months' duration and those who showed some instability after 6 weeks of immobilisation, a posterior atlanto-axial fusion is required.

In a Fielding type III patient with persistent neurological symptoms and failure to reduce after 6 weeks of attempted gradual reduction we would advocate an occipitocervical fusion followed by an anterior decompression by excision of the odontoid process. If the transoral approach is chosen, prophylactic antibiotics and aseptic technique should be used to prevent meningitis and encephalitis. Other approaches, like mandible- and tongue-splitting approaches, have been used but we do not have personal experience of these.

Conclusion

AARF is often diagnosed late, yet can result in serious complications and may be difficult to treat. Early and effective treatment of children who present with torticollis caused by atlanto-axial subluxation is necessary as it is not known which of them will progress to AARF.

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MEDUNSA's origin — the end obliterates the means

To the Editor: The Medical University of Southern Africa comes of age as a unique medical institution that has acquired undeniable stature. It has blazed a trail unequalled in the medical and health training of black Africans, with high standards in a wide range of health and health-related fields. MEDUNSA was conceived as a typical 'bush' university during the apartheid era. Its origins were steeped in rumours and controversies. The previous regime was noted for weird distortions, such as taking correct actions for wrong reasons or achieving good results while implementing perverted courses of action. Ironically, for the very reasons currently being debated, this was the problem with the desirability of producing more black health manpower. Rural areas were under-served and neglected and white missionaries had provided most services in rural black areas, at a sacrifice. However, the plans to meet this legitimate need went awry from the outset. In a widely read article1 a doctor claiming first-hand knowledge and authority, and also claiming to represent the MASA, made startling revelations about the MEDUNSA plans. His information sparked off a furor in the press,2 accompanied by intense public debate, and evoked outright rejection. The situation was not improved by those who claimed to speak on behalf of the government.3 There were also various reports on how the new institutions would provide careers for 'Bantu specialists'. This is a matter of history, given that when MEDUNSA was founded, the Bantu specialists who existed and applied could not be accommodated because the apartheid system had lagged behind the MEDUNSA experiment. At the time MEDUNSA opened, the Public Service Commission had not yet created posts for 'Bantu specialists' (PSC — personal communication, 1976). MEDUNSA's origins were therefore a unique experiment in terms of lecturing staff and students! These pioneers, both staff and students, functioned in an environment characterized by many gaps between them — political, social, cultural and linguistic. This was not surprising. The name 'MEDUNSA' was also cause for debate. Why 'Southern' Africa? It is open to question what this term meant or with what objective it was formulated. One view (African) on 'southern Africa' was that the medical architects and supporters of the grand apartheid plan of balkanisation of South Africa, were preparing to justify a southern African community born out of 'white' South Africa and autonomous Bantustans. All this is history now, thanks to the eventual beginning of a democratic South Africa in 1994. MEDUNSA can justifiably bask in its present well-deserved glory (whatever the mechanisms whereby it was achieved) of being a true pioneer in community-based tertiary education (African) on 'southern Africa' was that the (Whatever the mechanisms whereby it was achieved) of being a true pioneer in community-based tertiary education and teaching of disadvantaged black students.

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There is life after the diagnosis

To the Editor: For the past 20 years I have listened to case presentations by registrars, interns and medical students at four medical schools; two of them in South Africa. I must have heard at least 7 000 such renditions over this period. These case presentations have a lot in common, but I would like to dwell on the peculiarity of the South African presenter, who tends to give 'the diagnosis' in the first statement he or she makes. The presentation typically begins as follows: 'I am presenting a case of pneumonia', or whatever the case may be. Even when the diagnosis is unjustifiable or suspect, the format is the same. To me this is like putting the cart before the horse.

The obsession about mentioning the diagnosis immediately one opens one's mouth might have its origins in the mistaken belief that once the diagnosis has been given the battle has been won. Medical students in particular seem to think that a diagnosis is the end point or the bottom line. They seem to believe registrars and senior colleagues can now take over the show. My contention is that there is life after the diagnosis. The diagnosis has to be confirmed through investigations and case management needs to be discussed.

Could this near obsession with the diagnosis have its origins in the message we send to the medical students? Doesn't 'passing' or 'failing' a clinical examination often depend on whether a candidate at undergraduate level has made the diagnosis or not?

Here is a real-life scenario I witnessed not long ago. A final-year M.B. Ch.B. student was given a history of a 4-year-old child who could no longer walk but had no neuron signs in the lower limbs. Instead of giving a list of possible diagnoses at the end of her presentation, the candidate started off with (you guessed it), 'I am presenting a case of lower motor neuron disease ... '. The actual diagnosis in this case was tuberculosis of the spine with upper motor neuron signs in the lower limbs.

Unless we all agree that the method of arriving at the diagnosis through history, clinical examination and investigations and a discussion of what to do with the diagnosis are as important as the diagnosis itself, we shall always have the cart before the horse. Diagnosis will be achieved by any means at all costs.

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