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Atlanto-axial subluxation (Grisel's syndrome) following otolaryngological diseases and procedures

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Abstract
Grisel's syndrome is a rare condition of uncertain aetiology characterized by atlanto-axial subluxation following an infection in the head and neck region. The condition most frequently affects children and characteristically presents in the post-operative period with cervical pain and torticollis. We describe three cases and discuss the aetiology and pathogenesis. A high index of suspicion may lead to early diagnosis in its natural history thereby preventing potentially serious complications.

Key words: Grisel's syndrome; Atlanto-axial joint; Torticollis

Introduction

Non-traumatic subluxation of the atlanto-axial joint secondary to metastatic infection in the head and neck is a rare condition. Although first described by Sir Charles Bell, in 1830, it bears the eponymous name of the French physician, Grisel who described two cases in 1930 (Grisel, 1930). Characteristically the condition most frequently affects children, although cases occurring in adults have been described (Wetzel and LaRocca, 1989). The infection starts from a focus in the head and neck and then spreads to the cervical vertebrae, but the exact route is uncertain. The inflammation causes ligamentous laxity with the risk of atlanto-axial subluxation and in severe cases spinal cord injury (Roach *et al.*, 1984). The condition has been infrequently noted in the ENT literature, but has been described following tonsillectomy (Wittek, 1908), mastoid surgery (Fitzsimmons, 1915), nasopharyngitis (Jones, 1932) and repair of choanal atresia (Hopla *et al.*, 1983). We describe three cases and briefly discuss the aetiology and pathogenesis.

Case reports

Case 1

An 11-year-old boy underwent a left modified radical mastoidectomy for cholesteatoma. The procedure was uneventful as was the initial post-operative recovery. Eight days following surgery he developed a torticollis with neck pain and stiffness. On examination he was mildly pyrexial at 37.5°C. He held his neck flexed to the side of the mastoidectomy and all neck movements were painful and restricted by approximately 50 per cent. The aural pack was removed revealing a mild otitis externa. He had no neurological signs and examination of the pharynx was unremarkable. The sternomastoid muscle on the side of the torticollis was lax but that on the opposite side was in spasm.

A full blood count was normal but the erythrocyte sedimentation rate (ESR) was elevated at 19 mm/h. Plain radiographs of the cervical spine including odontoid views were unremarkable. High resolution computer tomogram (CT) scans with 3D reconstruction showed the dens to be sitting eccentrically within the anterior arch of the atlas and transverse ligament (Figure 1). The facet joints were intact, but the facet joint space on the side of the torticollis had been reduced resulting in a lateral tilt of the atlas on the axis (Figure 2).

An orthopaedic opinion was sought and it was felt that the atlanto-axial subluxation was stable. He was therefore treated symptomatically with analgesics, a soft collar and gentle physiotherapy to reduce the stiffness. The torticollis resolved completely after five months of treatment leaving him with a full range of painless cervical movement.

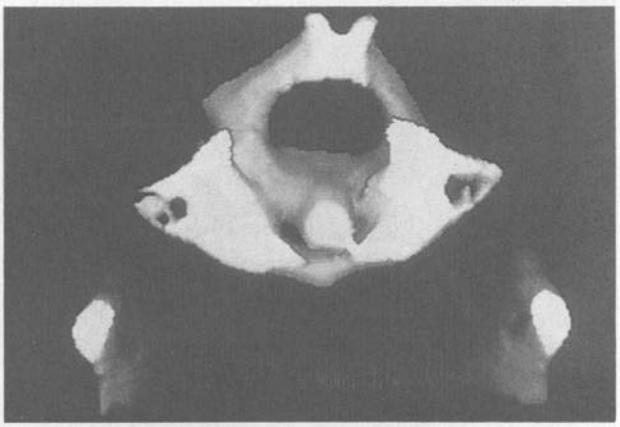

FIG. 1
A 3D reconstruction of trans-axial CT scan of *Case 1* showing the dens to be sitting eccentrically within the anterior arch of the atlas and transverse ligament.

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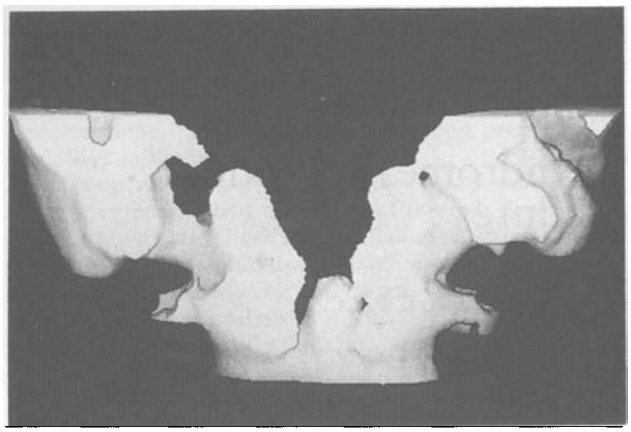

FIG. 2

Further scan of *Case 1* showing lateral tilt of the dens in relation to skull base and atlas.

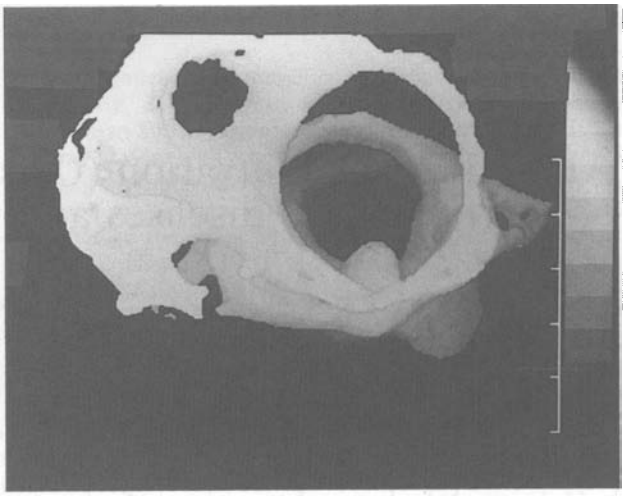

FIG. 3

A 3D reconstruction of trans-axial CT scan of *Case 2* showing rotation of atlas relative to axis with bilateral facet joint subluxation.

Case 2

A 10-year-old girl was admitted with severe follicular tonsillitis. She was commended on intravenous fluids and antibiotics. Over the next two days she developed a painful torticollis. On examination she was pyrexial at 38°C. All neck movements were severely restricted and painful, with a torticollis to the right. As with *Case 1* the sternomastoid muscle on the side of the torticollis was lax but the contralateral muscle was in spasm. She had no neurological signs or symptoms.

A full blood count showed a leucocytosis with predominant neutrophilia and the ESR was elevated at 25 mm/h. A high resolution CT scan with 3D reconstruction demonstrated significant atlanto-axial subluxation (Figure 3). She was transferred to a spinal injuries unit where she was managed using cervical traction to reduce the subluxation. External stabilization was required and halo traction was used. Repeat CT scans showed that the subluxation had been reduced and a further scan at three months showed that the atlanto-axial joint had become ankylosed but stable. Clinically she was pain-free and had no residual torticollis. There was full range of movement of her neck and she was discharged six months post-operatively.

Case 3

A five-year-old girl underwent an uneventful adenoton-

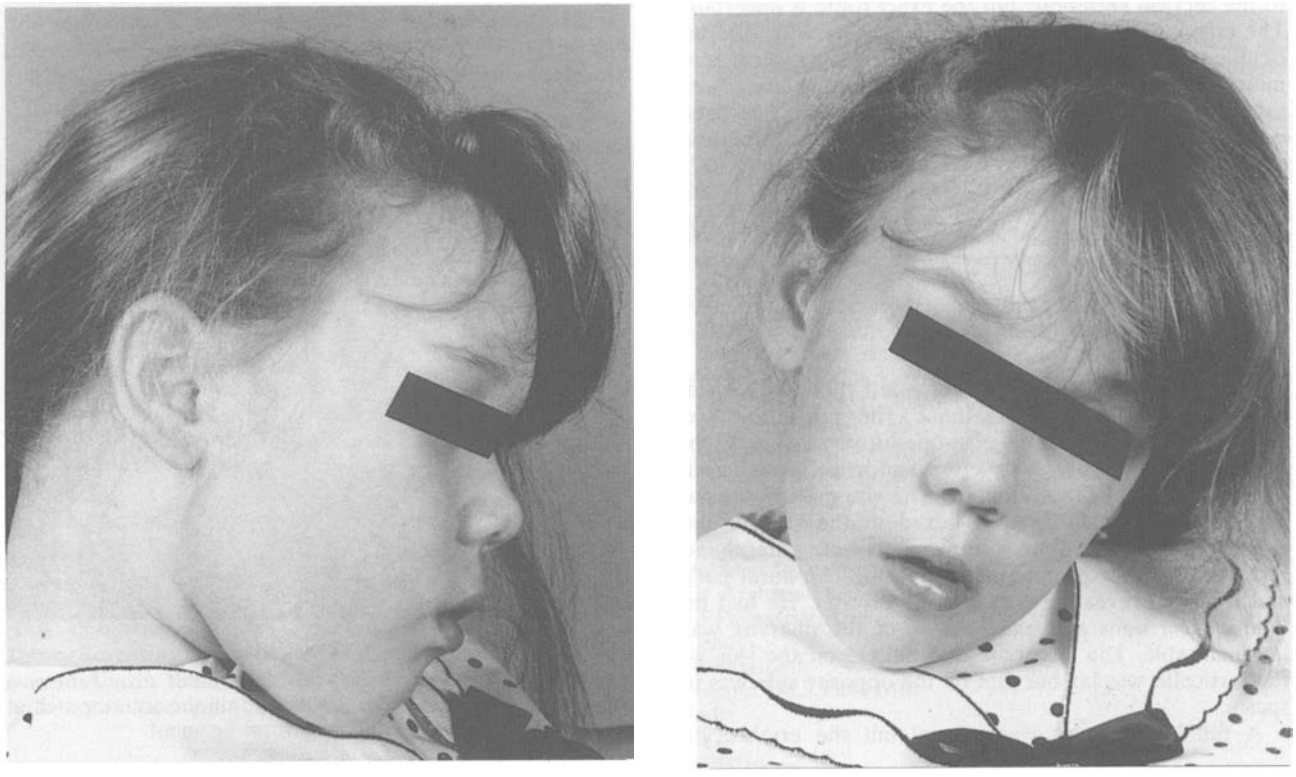

FIG. 4

Case 3 demonstrating a left-sided torticollis with the chin rotated to the right. The sternocleidomastoid muscle on the right side was in spasm.

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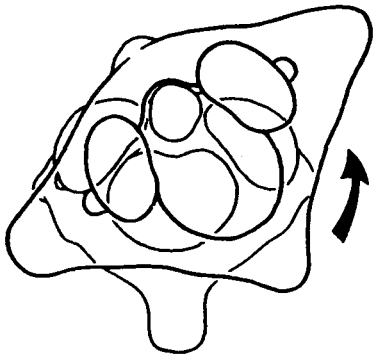
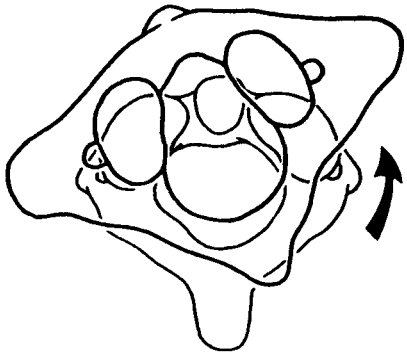
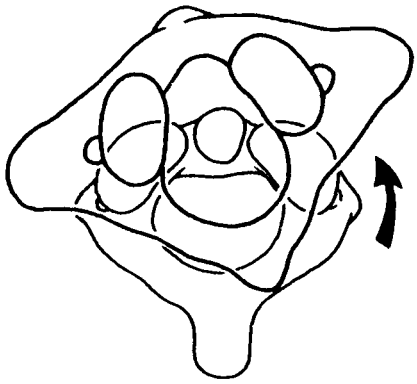


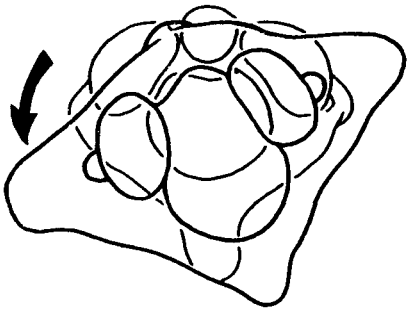
Type I - Rotary fixation without anterior displacement of the atlas (displacement of 3mm or less)



Type II - Rotary fixation with anterior displacement of the atlas of 3 to 5mm



Type III - Rotary fixation with anterior displacement of greater than 5mm



Type IV - Rotary fixation with posterior displacement

FIG. 5
Fielding classification of atlanto-axial subluxation.

sillectomy for recurrent tonsillitis. Four days post-operatively she developed a painful torticollis to the left (Figure 4). On examination she was apyrexial and both tonsillar fossae were clear of clot and debris. The head was tilted to the left, with the chin rotated to the right. The right sternocleidomastoid muscle was in spasm. The clinical diagnosis was of Grisel's syndrome. Plain radiographs of the neck were unhelpful and a request for a CT scan under general anaesthesia was made but refused by her parents. She was treated expectantly with analgesics and a soft collar. The torticollis completely resolved after three weeks with no residual symptoms.

Discussion

Non-traumatic atlanto-axial subluxation is an uncommon, poorly understood condition which occurs mainly in children and is thought to result from ligamentous laxity following an inflammatory process (Wetzel and LaRocca, 1989). The first published case was described by Sir Charles Bell, in 1830, in a patient with a syphilitic pharyngeal ulcer. Postmortem examination showed a C1/C2 dislocation secondary to rupture of the transverse ligament of the atlas. Similar case reports followed (Thompson, 1834; Ballance, 1884) but the condition bears the eponymous name of the French physician Grisel who described two cases in 1930 (Grisel, 1930). The association of infection in the pharynx or skull base and acquired torticollis has appeared under a variety of names including rotatory dislocation (Corner, 1907), hyperaemic dislocation (Jones, 1932), inflammatory dislocation (Fitzwilliams, 1934) and spontaneous non-traumatic subluxation (Frank, 1936). The varied nomenclature reflects the uncertain aetiology, all of which imply that subluxation of the atlanto-axial joint is secondary to ligamentous laxity due to an infectious process. Grisel's syndrome is primarily a disease of children and young adults (Wetzel and LaRocca, 1989), Sullivan *et al.* (1958) showed that 77 per cent of cases were younger than 13 years of age.

Integrity of the transverse ligament of the atlas is considered to be the most important factor in the stability of the atlanto-axial joint. In the event of total rupture of the transverse ligament the extent of subluxation depends on the functional length of the adjacent alar ligaments. This length decreases with age. Several authors (Greig, 1931; Fielding and Hawkins, 1977; Parke *et al.*, 1984)

believe that the greater laxity and vascularity of the atlanto-axial area in the first decade explains why the disease is more frequent in children. Fielding *et al.* (1978a) studied 17 patients with fixed rotatory subluxation and formulated a working classification into four distinct types (Figure 5). Type I demonstrates a rotatory fixation without anterior displacement of the atlas (displacement of 3 mm or less). This is the most common deformity and the transverse ligament remains intact. Type II has a rotatory fixation with anterior displacement of the atlas of 3 to 5 mm. This is the second most common deformity and is associated with some deficiency of the tranverse ligament and unilateral anterior displacement of one lateral mass while the contralateral joint acts as a pivot. Type III gives a rotatory fixation with anterior displacement of more than 5 mm. This type of deformity is associated with a deficiency of both the transverse and the other ligaments. Both lateral masses of the atlas are displaced anteriorly and asymmetrically, producing a rotatory deformity. Type IV is associated with rotatory fixation and posterior displacement. This is a rare lesion and occurs when a deficient dens allows asymmetrical posterior shifting of the lateral masses to produce a rotational deformity. The Fielding classification is also a prognostic indicator of the potential for complications. Type I is the most benign lesion, whereas Types III and IV although rare, are invariably associated with spinal cord compression with possible fatal consequences.

Three main theories have been suggested to explain the pathogenesis of Grisel's syndrome. Grisel (1930) believed that the torticollis was due to spasm of the cervical muscles pulling the occiput down onto the axis and partially extruding the atlas resulting in instability. However, most authors now believe that muscular spasm is a protective reflex rather than the cause of the displacement (Wetzel and LaRocca, 1989). Grieg (1931) proposed that local infection causes regional hyperaemia resulting in decalcification of the bony arch of the atlas near the insertion of the tranverse ligament. Watson-Jones and Roberts (1934) found radiographical evidence of decalcification in the anterior part of the atlas associated with subluxation, with recalcification occurring as the infection disappeared. However, in an extensive literature review, Pandya (1972) found no strong evidence of decalcification or postmortem proof of avulsion of the transverse ligament from its insertion to osteopenic bone. As early as 1908 Wittek (1908) suggested the theory that metastatic infection leads to peri-ligamentous inflammation resulting in laxity and consequent subluxation. The venous and lymph drainage of the pharynx was demonstrated by Parke *et al.* (1984) and explains a possible route for spread of infection lending support to the latter theory.

Patients with this condition often present with non-specific symptoms making diagnosis more difficult. Most patients complain of neck pain and stiffness, although dysphagia and odynophagia are frequent symptoms. Patients may develop a conductive hearing loss due to obstruction of the eustachian tube. Rhinolalia clausa due to obstruction of the nasopharynx by the anteriorly displaced anterior arch of the atlas has been reported (Watson-Jones and Roberts, 1934).

A mild pyrexia is usually present but physical findings will depend on the severity of the subluxation. The head will be rotated to the side opposite the facet dislocation. The neck is generally tender to palpation and attempted movement is painful. The spinous process of the axis may be palpable on the opposite side to the dislocation (Sudeck's sign) (Sudeck, 1923) and the sternomastoid muscle on the side of the torticollis is lax while the contralateral muscle is in spasm. This is in contrast to spasmodic torticollis where the converse is true. Neurological signs and symptoms are uncommon and can range from simple parasthesia to quadriparesis (Boever and Hennebert, 1953; Wetzel and LaRocca, 1989). Death has been reported (Boever and Hennebert, 1953; Fielding *et al.*, 1978a) and is invariably due to respiratory paralysis secondary to medullary compression.

Radiological investigations play the most important part in diagnosis. Plain antero–posterior and lateral radiographs may show asymmetry between the facet joints and increased distance between the anterior arch of the atlas and the dens. Decalcification can be seen on plain radiographs, but is only visible after four to six weeks. These investigations may be difficult to interpret and provide limited diagnostic yield. The investigation of choice is either high resolution CT scanning (Fielding *et al.*, 1978b), with 3D reconstruction to demonstrate bony subluxation, or magnetic resonance imaging (MRI) to detect signs of inflammation in the ligaments concerned, ideally both investigations should be performed (Roach *et al.*, 1984; McAfee *et al.*, 1986).

The key to adequate management is early diagnosis, misdiagnosis occurs frequently (Sullivan *et al.*, 1958) and the rate of complications is directly proportional to the delay in diagnosis. Broad spectrum antibiotics should be prescribed initially until the organism is identified either by blood culture, local aspiration or biopsy.

Debate exists in the literature as to the further management of the atlanto-axial subluxation, but generally depends on the Fielding classification and the chronicity of the disease. For Type I subluxation simple support in a soft collar and muscle relaxants is sufficient (Boiten *et al.*, 1986). For more severe subluxation most authors recommend cervical traction in the acute stage to reduce the subluxation followed by immobilization in a soft collar for at least three months (Phillips and Hensinger, 1989). Repeat scans are obtained to assess the adequacy of the reduction and immobilization. If the deformity has been present for more than three months, arthrodesis or joint immobility has usually occurred. In these situations traction will not effect reduction and permanent residual deformity may then develop. If the spine is stable conservative measures and regular review may be appropriate. However, several authors (Sullivan *et al.*, 1958; Masalawala, 1977) have suggested that in this situation the above measures may not be sufficient. If there are neurological signs or suspected instability operative decompression and arthrodesis is required (Fielding and Hawkins, 1977; Phillips and Hensinger, 1989).

Otorhinolaryngologists should be made aware of this condition and be vigilant of its early presentation so that effective treatment can be given before the development of potentially disastrous complications.

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