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CASE REPORT

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Pathogenesis of non-traumatic atlanto-axial subluxation (Grisel's syndrome)

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Abstract Non-traumatic atlanto-axial subluxation (AAS) is an uncommon complication of upper neck inflammatory processes and head and neck surgery. It is also known under the eponym of Grisel's syndrome (GS). We present a case report of a 6-year-old boy with GS that resulted from a retropharyngeal abscess. A diagnosis was not made until 2 months after the occurrence of infection. Re-evaluation of repeated CT scans of the neck showed the sequential development of AAS. These findings implied that the pathogenesis of GS is a distention of the ligaments between the atlas and axis rather than loosening of the ligaments caused by the spread of an inflammatory edema from the soft tissues of the neck as has been proposed by others.

Key words Atlanto-axial joint subluxation · Grisel's syndrome · Retropharyngeal abscess · Torticollis · Computed tomography

Introduction

Non-traumatic atlanto-axial subluxation (AAS) associated with head and neck disease or otolaryngologic procedures is a rare condition. It typically occurs in children with an infection in the head and neck [16]. The presenting symptom other than that of the infection is persisting torticollis. There are different proposed explanations for the pathogenesis. We describe a case in which sequential CT scans of the neck were obtained. Re-evaluation of these scans showed the step-by-step development of AAS, indicating a new consideration for its pathogenesis.

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Case report

A 6-year-old white Swedish boy was referred to the ENT Department of Länssjukhuset Ryhov because of a sore throat, swelling on the left side of his neck, fever and left torticollis. Physical examination revealed grossly enlarged lymph nodes over the left sternocleidomastoid muscle without fluctuation. Pharyngitis was seen, as well as enlargement of the left tonsil but without peritonsillar swelling. White blood cell count was elevated to 19,500 with 86% polymorphonuclear forms. The red blood cell count was normal.

Oral penicillin V was given, but no improvement was seen at follow-up 2 days later. Manual attempts to correct the torticollis caused pain over the vertex. There was no neurologic impairment. Intravenous (IV) penicillin therapy was then started, and tonsillectomy was performed the next day as definitive treatment for tonsil infection. However no peritonsillar abscess was found and histological examination of the tonsils showed only inflammatory changes.

Eight days after admission recovery from the infection was seen, but the torticollis persisted. A retropharyngeal abscess was then suspected and indicated by CT (Fig. 1). Antibiotic treatment was changed to IV cefuroxime, and surgical exploration of the neck performed under general anesthesia. At surgery no abscess was found.

Repeat CT 2 days later showed only some remaining edema. Since the torticollis did not subside skeletal scintigraphy using technetium-99 as radionuclide was performed. A small area of increased uptake was seen on the right side of the neck at the C1-C2 level. This was not typical for osteomyelitis and was interpreted as an artifact or possible inflammation of muscle attachments. The patient was then sent home on oral clindamycin.

At check-up 22 days after admission the only complaint was persistent torticollis. CT was again repeated but was interpreted as normal. Physiotherapy was started, but two months after the initial admission the patient was still unable to rotate his head to the right past a neutral position. A diagnosis of AAS was finally confirmed by means of dynamic CT performed under general anesthesia. Complete repositioning of the atlas on the axis was then seen when the head was carefully rotated to the right. Skull traction was used for 2 weeks, after which a halo-vest was applied for 4 weeks. However, 4 days after removal of the halo-vest reluxation occurred.

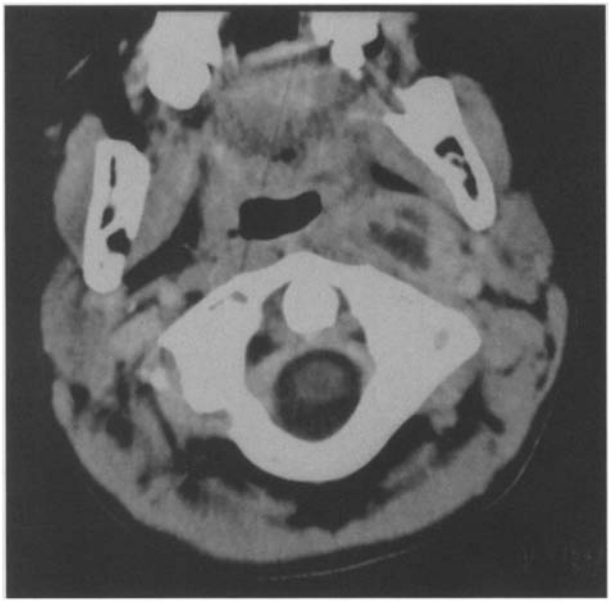

Fig. 1 CT scan of the upper neck showing a retropharyngeal abscess on the left side

Skull traction and halo-vest treatment was again applied, and maintained for 12 weeks. After this no torticollis was seen and normal range of motion of the neck was restored. A final CT scan was obtained 1 year later and normal relationships between the atlas and axis were seen at maximum rotation of the head to both sides. There have been no further sequelae to date.

Discussion

The first report on non-traumatic AAS secondary to an inflammatory process in the upper neck was published by Sir Charles Bell [1] in 1830. He described a male patient who died with a pharyngeal ulcer that was possibly syphilitic. Autopsy showed erosion of the transverse ligament of the axis. Cases have since been reported sporadically [16, 19]. At present a total of approximately 100 cases can be found in the medical literature [1–3, 5, 7–12, 14–16, 18–23]. Several names have been given to this condition, including "distention luxation," "nasopharyngeal torticollis," Grisel's syndrome (GS), spontaneous hyperemic dislocation of the atlas and atlanto-axial rotary fixation. The terms most frequently used in contemporary literature are GS or non-traumatic AAS.

The associated upper neck inflammatory process in GS includes an upper respiratory infection, otitis media, mastoiditis, tonsillitis, parotitis, cervical adenitis and upper neck masses. Retropharyngeal cellulitis or abscesses as the cause of transient torticollis are probably more common than generally thought [4, 17]. AAS has also resulted from a complication to ENT surgery, including tonsillectomy, adenoidectomy, mastoidectomy, choanal atresia repair, or repair for cleft palate [14, 16]. AAS has occurred in patients with systemic sepsis, been a feature in rheumatic disease or following trauma, and has been idiopathic.

The joint between atlas and axis is unique and complex. It allows flexion-extension, vertical approximation

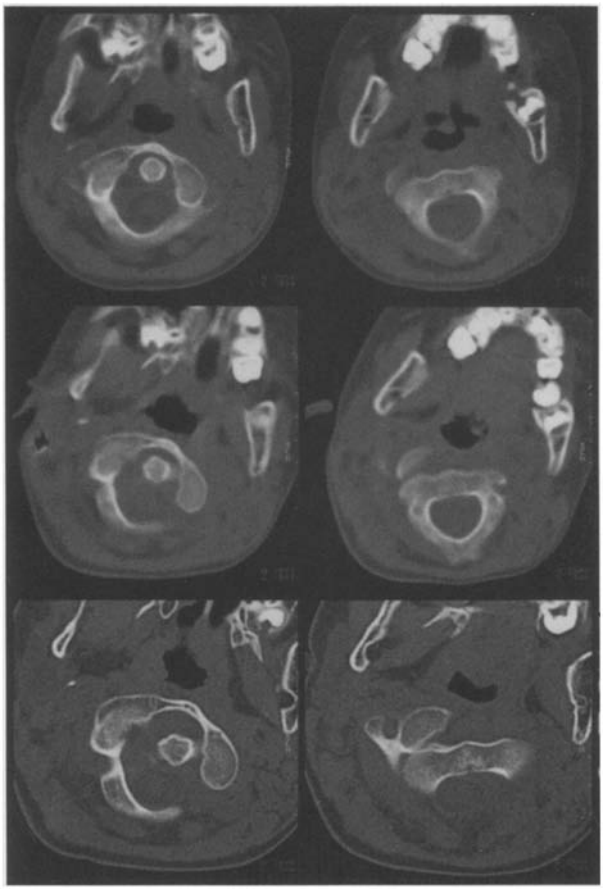

Fig. 2 Sequential CT scans of the neck showing the development of atlanto-axial subluxation (AAS) over time. *Top* Eight days after admission. Bone windows show rotation within a normal range. The distance between the posterior border of the anterior ring of the atlas and the anterior aspect of the dens axis is 1.5 mm, representing a type I AAS. *Middle* Four weeks after admission. Further progression of rotational dislocation is present, but the dens is still in a normal position. The distance between the atlas and dens is 3 mm. Soft tissue window pictures at this point show no remaining edema or other inflammatory signs. *Bottom* Two months after admission. A type II AAS is seen. The atlas has rotated with the left lateral mass acting as the pivot, and the dislocated dens indicating overdistention of the transverse ligament. The distance between the atlas and dens is 7–8 mm. Subluxation has developed on the side contralateral to the resolved retropharyngeal abscess

as well as lateral translation movements, but its principal movement is that of rotation [6, 16]. The transverse ligament prevents anterior dislocation of the atlas on the axis. The alar ligaments assist in this function but primarily prevent excessive rotation. That the loosening or distention of these ligaments is necessary for non-traumatic AAS to occur was understood by early clinicians [8, 21]. How the loosening or distention of these ligaments can result from an inflammatory process having no direct connection with the joint structures has been the subject of many theories.

Identification of the pharyngovertebral veins draining the posterior pharyngeal region gave a possible anatomic rationale for AAS [13]. It was then proposed that edema was spread via this route to the ligaments responsible for

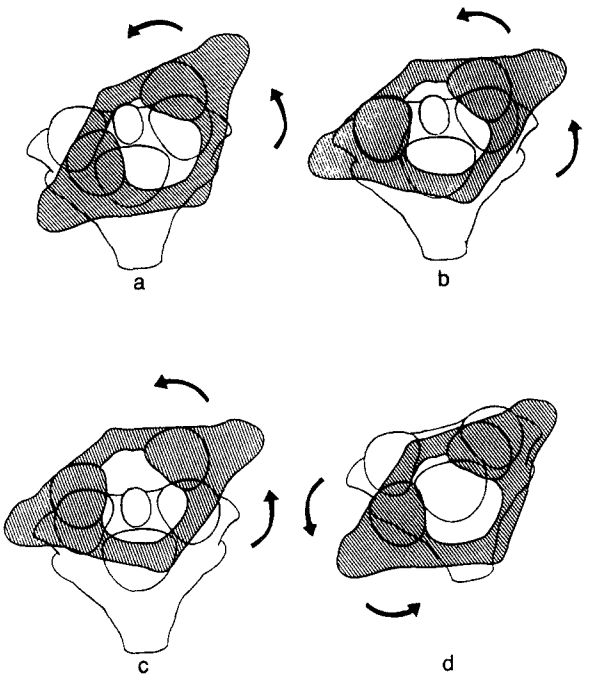

Fig. 3a–d Fielding's classification of AAS. **a** Type I is not an actual subluxation but a rotation within the normal range of movement between the atlas and the axis. ***b*** Type II shows unilateral displacement of one lateral mass of the atlas while the opposite, intact lateral joint acts as the pivot. **c** Type III shows how both lateral aspects of the atlas can be displaced anteriorly. **d** Type IV shows posterior subluxation of the atlas with the odontoid process fractured or absent. (Reprinted with permission from [5])

the stability between atlas and axis, and that these ligaments were loosened and pathologic rotation of the atlas on the axis resulted.

Exactly when actual subluxation occurs in relation to torticollis has not been discussed in earlier reports. Our case provides new information in this respect. Our initial CT scans did not demonstrate any abnormal rotation between the atlas and axis, despite the impressive torticollis present. Rotation increased with time without dislocation of the dens. The final subluxation includes dorsal displacement of the dens in relation to the atlas and occurred long after the inflammatory process and edema in the soft tissues of the neck had resolved (Fig. 2a–c). Furthermore the subluxation was seen on the side contralateral to the retropharyngeal abscess, as also seen in Wittek's [21] case.

We believe that our case proves that the pathogenesis of AAS is not a matter of the spread of edema causing abnormal loosening of ligaments between the atlas and the axis. The condition begins rather as a typical torticollis caused by spasm of irritated neck muscles. If long-standing, this results in distention of the ligaments involved, and eventually in subluxation. The prerequisite is probably elastic or lax ligaments, explaining why AAS is almost exclusively seen in children. Lax ligaments are also a known feature in Down's syndrome with atlanto-axial joint instability, increasing the risk for non-traumatic AAS [9].

As shown in Fig. 3, non-traumatic AAS has been classified by Fielding and Hawkins [5] into four types. Types I and II are the most frequently found clinically, with no neurologic deficits. However, there are also cases in which pure anterior dislocation occurs without rotation [11]. Our case initially represented a type I disorder (Fig. 2a), followed by a type II subluxation (Fig. 2c).

The clinical findings in AAS include torticollis with the chin turned away from the subluxated side. The head is tilted towards the affected side and flexed anteriorly ("cock-robin" look) making it difficult to open the mouth. Attempts to turn the head cause pain in the occiput and vertex as well as the neck. The spinous process of the axis can be palpated in the neck away from the midline toward the affected side (Sudeck's sign). The bulging anterior arch of the atlas can sometimes be seen or palpated in the nasopharynx. The mass effect can also produce a nasal quality of the voice [15].

Actual neurologic impairment involves exaggerated reflexes of the limbs, tetraplegia and bladder dysfunction but is extremely rare in otherwise healthy patients [2, 5, 20–22]. However, this occurrence is more common in patients with Down's syndrome [9].

Radiographic confirmation of AAS can be difficult using a plain X-ray technique. Our review of the reported cases showed that several initial roentgenograms were reported as "normal." The use of CT in selected cases was proposed by Fielding et al. [7], although an adequate diagnosis is best achieved with a dynamic CT scan. As active and passive movements in affected patients can be very painful, the investigation usually has to be performed under general anesthesia. Scintigraphy can possibly be of some help [18], but the missing of a correct diagnosis (as in our case) is a hazard because of non-specific findings [3, 10].

The recommended treatment of AAS involves the initial application of a skull traction tong under general anesthesia followed by a dynamic CT scan in which the head is rotated maximally to either side. If the movements of the atlas on the axis are substantially restricted, a diagnosis of fixed atlanto-axial rotation can be made and traction applied. As soon as the patient can rotate his head symmetrically to left and right a halo-vest can be applied. The vest is generally maintained for a total of 6–12 weeks depending on the length of the history. If reluxation occurs, an additional attempt with traction and halo-vest has proven successful [3]. Surgical fusion can become necessary if the joint remains unstable or the subluxation unreduced [6].

When effectively treated, the prognosis of non-traumatic AAS is excellent. Without treatment AAS becomes permanent, and will result in persistent torticollis with possible facial flattening [5, 16]. A few cases have regained normal, or almost normal, mobility of the neck despite persistent or recurring AAS seen roentgenographically [5, 11, 21]. However, such cases can still have future problems with neck pain and premature degenerative disease.

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