Infection-related spontaneous atlantoaxial dislocation in an adult

Case report

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This paper reports the third described case of infection-related atlantoaxial subluxation in an adult. Like most of the similar cases seen in the pediatric literature, this case was associated with a parapharyngeal β-hemolytic streptococcal abscess. Based upon this experience, the authors advocate intravenous antibiotic therapy and 1) immediate reduction followed by application of a halo brace; 2) immobilization in a halo brace for at least 3 months; and 3) a C1–2 wiring and fusion procedure for patients who fail this trial of conservative therapy.

KEY WORDS • atlantoaxial dislocation • infection • spine stabilization

Atlantoaxial dislocations are usually categorized as congenital, traumatic, or acquired ("spontaneous") in association with pharyngeal infection or inflammatory joint disease. The first case of spontaneous atlantoaxial dislocation was reported by Sir Charles Bell in 1830, and was associated with destruction of the transverse ligament by an eroding syphilitic ulcer of the posterior pharyngeal wall. Since that time more than 200 cases of spontaneous atlantoaxial dislocation have been reported. The infection-related type has occurred almost exclusively in children; adult cases are most commonly associated with rheumatoid arthritis or other noninfectious inflammatory conditions. There are two exceptions to this: one in a 22-year-old soldier reported by Swanberg and one in a 62-year-old woman reported by Wilson.

The purpose of this paper is to add to the literature a third case of an infection-related atlantoaxial dislocation in an adult, to describe the relevant anatomy and pathophysiology of this rare entity, and to discuss an approach to the management of these cases in light of the experience gained in treating this patient.

Case Report

This 23-year-old muscular black man presented with a complaint of a sore throat and progressively increasing neck pain of 2 to 3 weeks' duration. Over the 3 days just prior to admission he developed dysphagia and the additional complaint of a heavy feeling in his throat. He was otherwise healthy, and specifically denied any history of injury or trauma.

Examination. The patient was evaluated in the emergency department by the ear, nose, and throat service. Examination revealed no torticollis or rotation of the head; the head and shoulders were turned as a unit, presumably secondary to neck pain. No sternocleidomastoid muscle spasm was palpable. There was marked edema in the posterior pharyngeal wall. The epiglottis was normal and the tonsils were not enlarged. The white blood cell count was 18,600/cu mm with 92% segmented neutrophils, and the hematocrit was 41.8%. The erythrocyte sedimentation rate was 36 mm/hr.

A contrast-enhanced computerized tomography (CT) scan of the neck showed a 5 × 3-cm retropharyngeal abscess (Fig. 1). Cervical spine x-ray films were taken but were not commented upon at that time. A course of intravenous penicillin and clindamycin was begun empirically and the patient was taken to surgery where the abscess was drained of about 10 ml of purulent material.

Course. A review of the cervical spine x-ray films the next morning disclosed a subluxation of C-1 on...
FIG. 1. Left: Computerized tomography (CT) scan of the neck showing a large retropharyngeal abscess. Right: Scout view from the CT scan demonstrating the increased atlanto-dens interval, but this went unnoticed at first.

FIG. 2. Left: Cervical radiograph on flexion showing about 11 mm of subluxation of C-1 on C-2. Right: Cervical radiograph in extension showing reduction of the atlanto-dens interval to about 3 mm.

C-2 of approximately 10 mm. A neurosurgery consultation revealed the patient to be neurologically intact. A Philadelphia collar was placed temporarily, and cervical spine x-ray films were obtained in active flexion and extension in order to evaluate spinal stability. On flexion there was 11 mm of subluxation, which reduced to 3 mm on extension (Fig. 2). The orthopedic surgery department was then consulted with regard to placement of an orthosis. The patient was placed in a halo vest which reduced the subluxation to 3 mm. Final culture results of the abscess contents showed a Group A β-hemolytic streptococcal infection, and intravenous penicillin was continued for a total of 4 weeks, at which time a repeat erythrocyte sedimentation rate was within normal limits. This was followed by a 2-week course of oral penicillin. A gallium citrate scan showed no focal increase in activity in the C1–2 area.

The patient was maintained in the halo apparatus for 2 months, during which he presented several times for adjustment of loosened pins. A cervical spine x-ray film obtained 1 month after removal of the halo device showed 4 mm of anterior subluxation of C-1 on C-2 with flexion, and complete reduction on extension. At this time, a C1–2 fusion was recommended, but the patient refused surgical intervention. When he returned for routine evaluation 1 month later, repeat cervical spine x-ray films on flexion and extension showed a persistently increased atlanto-dens interval of 3 mm, but no subluxation. He has since been followed at 6-month intervals. He remains neurologically intact, but his flexion and extension cervical radiographs show a persistent abnormal atlanto-dens interval without abnormal movement.

Discussion

The transverse ligament prevents excessive anterior shift of the atlas of the axis, and many authors consider that this is the site of the lesion in cases of spontaneous dislocation. Several theories have been proposed to explain the lesion of the transverse ligament. Wittek was the first to draw attention to the fact that subluxation may follow local cervical spine infection. In 1931, Greig postulated that hyperemia due to an inflammatory process decalcified the vertebrae, particularly around the sites of ligamentous attachment to bone, allowing a loosening of the transverse ligament. This was consistent with the earlier work by Leriche and Policard that demonstrated decalcification of bone in the presence of hyperemia and sclerosis resulting from ischemia. Radiological evidence and support for this concept were provided in the case reports of Watson Jones. He noted the following characteristics of the syndrome: 1) the condition is peculiar to children; 2) it is always associated with some type of inflammation in the upper cervical area; 3) there is a latent period of 7
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to 10 days between the onset of infection and the dislocation; and 4) the anterior arch of the atlas shows decalcification but not destruction.

In 1953, Lippmann demonstrated experimentally that aseptic inflammation around a knee joint of the rabbit caused decalcification of bone and weakening of the ligaments. While he proposed no mechanism for these and other changes, more importantly, he noted that when subjected to a graded strain a ligament will tear from within before it separates at its insertion into bone. He further stated that these ligamentous changes proceeded any radiographically demonstrable bone changes.

Although these data would seem to clarify why decalcification of bone is only inconsistently observed in cases of spontaneous dislocation, other serious questions remain. First, Werne showed that the transverse ligament will not become totally insufficient unless it is entirely ruptured or weakened in its entire transverse section. While this “all or none” phenomenon would help account for the lack of a correlation between the occurrence of subluxation and the severity of the infection noted by Hunter, it leaves serious questions unanswered regarding the role of the alar and capsular ligaments in the occurrence of this dislocation. Second, the time interval required between the presence of infection and the resulting dislocation is too variable. For instance, dislocation has occurred from 1 day after the onset of infection to several years later. Also, although many young children have cases of cervical sepsis, atlantoaxial dislocation is rare. Finally, there are questions regarding the contribution that spasm in the deep cervical muscles may make to the amount of subluxation seen. This was originally recognized by Grisel, and probably is related to the rotary torticollis that often accompanies spontaneous atlantoaxial dislocation in children. The total clinical syndrome is most likely the result of a combination of factors including decalcification, ligamentous laxity, and deep cervical muscle spasm.

Anatomic predisposition to atlantoaxial subluxation occurs in certain subgroups during adulthood. Up to 20% of patients with Down’s syndrome have been found to have atlantoaxial instability, and this has been correlated with the degree of laxity of the transverse ligament. Both a congenital ligamentous weakness and an intrinsic connective tissue defect have been postulated as possible etiologies. It must be quite rare, but it is presumed that this anatomic predisposition to ligamentous laxity might also occur in certain otherwise normal adults and, in the setting of hyperemia and spasm of the deep cervical muscles, may lead to frank subluxation.

While high-resolution CT with reformatted sagittal images and any one of the many software programs showing bone density may shed new light on the pathophysiology of this condition, the current standard for making the diagnosis remains true lateral cervical radiographs in flexion and extension. These allow an assessment of the extent of the subluxation as well as the stability of the atlantoaxial articulation.

Because this condition is relatively rare, no one center has a wide range of experience in managing these cases. Watson Jones favored reduction under anesthesia followed by fixation in plaster. Sullivan considered reduction under anesthesia to be too dangerous, and favored reduction with traction. Werne agreed with the use of traction, and recommended dorsally directed traction in order to achieve reduction.

The present patient responded to a course of halo brace stabilization, but because of the relatively few similar cases and the lack of much experience at any one center, management must be individualized. Based upon our limited experience with adult spontaneous atlantoaxial subluxation we recommend: 1) immediate reduction followed by application of a halo brace; 2) halo brace immobilization for at least 3 months; and 3) performing a C1-2 wiring and fusion procedure for patients who fail this trial of conservative therapy.

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References


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