Treatment of nontraumatic atlantoaxial dislocation and fibrous fusion

Case report

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The authors describe a case of nontraumatic atlantoaxial dislocation secondary to acute rheumatic fever, in which there appeared to be fibrous fusion between the axis and the atlas in the subluxed position. The dislocation was reduced by means of combined traction and steroid therapy. Fibrous fusion occurred in the realigned position after immobilization.

KEY WORDS:
- spinal cord compression
- spinal fusion
- atlantoaxial dislocation
- rheumatic fever
- cervical traction

Numerous causes of nontraumatic dislocation of the atlas on the axis have been described. The etiology can be classified into three groups: congenital defects of the cervico-occipital junction; primary or metastatic tumors of the upper cervical spine and occiput; and inflammatory diseases. In this report, the authors describe the nonoperative treatment of a patient with marked atlantoaxial dislocation secondary to rheumatic fever, with apparent fibrous fusion of the dislocation in the subluxed position.

Case Report

On October 23, 1975, this 9-year-old girl was admitted to the Neurosurgical Service at the Medical University of South Carolina Hospital for further management of an apparently spontaneous fusion of a severe atlantoaxial dislocation. She had been treated at another hospital in October, 1974, for acute rheumatic fever. In March, 1975, she was reevaluated at the same hospital because of pain and stiffness in her neck for the previous 4 months. She was noted to have limitation of flexion and extension of the neck, and neck rotation was not possible. There was a firm, bone-like prominence over the upper cervical spine posteriorly, moderate weakness of abduction of the right arm at the shoulder, and winging of the right scapula. The right knee jerk was increased. X-ray films of the cervical spine at that time revealed a marked dislocation of the atlas on the axis, and no movement was seen on the flexion and extension views. In an attempt to reduce the dislocation, cervical traction with skull tongs was started. However, despite 5 weeks of continuous traction of up to 20 lb, there was no movement of the atlas on the axis.

On admission to the Medical University of South Carolina Hospital, the following findings were noted: a coarse pansystolic murmur; marked limitation of flexion and extension of the neck; limited neck rotation to 15° either side of the midline; a protuberance of bone that could be felt over the upper cervical spine posteriorly; winging of the right scapula; weakness of abduction of the right arm at the shoulder; and a hyperactive right knee jerk. Diagnostic blood studies, including ASO titer and sedimentation rate, were normal. Cervical spine x-ray films revealed marked anterior dislocation of the atlas on the axis (Fig. 1). Cineradiography of the cervical spine during flexion and extension revealed no movement between the occiput, atlas, and axis. A myelogram demonstrated that the spinal cord had been displaced to the left of the odontoid, but was not widened at that level. Somatosensory evoked potentials from the ulnar and

median nerves in both arms were mildly abnormal (right more than left), but those from the posterior tibial nerves at the ankles were normal on both sides.

On November 18, 1975, treatment was begun with 40 mg of prednisone daily for 3 days, then the dose was gradually decreased and stopped on January 9, 1976. On November 21, 1975, 15 lb of cervical traction with Cone-Barton tongs was begun, and counterbalancing pelvic traction was applied as the cervical traction was increased. On December 1, 1975, with the patient at 30 lb cervical and 20 lb of pelvic traction, x-ray films revealed relative widening of the C1–2 interspace. At this time, 10 lb of anteriorly directed cervical traction was added, by means of a soft cervical collar applied to the back of the neck (Fig. 2). Two days later, at 40 lb of cervical traction, the alignment of C-1 on C-2 was greatly improved. Cervical tong traction was then gradually reduced while anteriorly directed soft cervical traction was maintained for another 2 weeks. At that time, the distance between the anterior surface of the odontoid process and the posterior surface of the anterior arch of the atlas had decreased to 6 mm. It was thought that as much reduction of the dislocation as possible had been accomplished, so traction was discontinued. A Minerva jacket was applied for 3 months and was then replaced by a firm cervical collar. X-ray films at that

Fig. 1. Lateral cervical spine radiograph before traction, showing marked anterior dislocation of the atlas on the axis.

Fig. 2. Three types of traction are being applied to the patient. From left to right: conventional cervical traction with Cone-Barton tongs; soft low posterior cervical traction with vector directed anteriorly to reduce anterior subluxation of the atlas on the axis; and pelvic traction directed caudally.
Nontraumatic atlantoaxial dislocation
time revealed a persistent C1-2 subluxation of 6 or 7 mm (Fig. 3). Somatosensory evoked potentials from the arms and legs were essentially normal.

Discussion
The first case of atlantoaxial subluxation due to an inflammatory process was reported in 1830 by Sir Charles Bell. In that case, autopsy revealed erosion of the transverse ligament of the atlas by an inflammatory process which had spread from a pharyngeal ulcer. Atlantoaxial subluxation associated with acute rheumatic fever was first reported in 1905, and since then numerous other cases have been described in relation to inflammatory states. In 1961, Rogers classified the inflammatory diseases into two groups: local inflammatory processes, primarily occurring in children (such as tonsillitis and rheumatic fever); and systemic inflammatory diseases with local effects, primarily occurring in adults (such as rheumatoid arthritis and ankylosing spondylitis). The incidence of atlantoaxial dislocation associated with rheumatic fever is not known; however, it varies from 17% to 40% for rheumatoid arthritis.

Although numerous explanations for the subluxation have been proposed, the mechanism is still obscure. A widely held view is that the inflammatory process establishes a hyperemic state in the bone-ligament structure of the atlantoaxial complex, which may cause weakening or dissolution of the joint.

Lourie and Stewart suggested that prolonged steroid treatment in patients with rheumatoid arthritis could be a contributing factor to the atlantoaxial subluxation. Our patient was given steroids before and during the reduction of the dislocation, primarily as a protective measure for the neural structures involved, but also in the hope that there might be some dissolution of the fibrous adhesion that united the atlas and axis.

The goal of therapy in these patients is reduction of the dislocation, decompression of the upper cervical spinal cord, and stabilization of the atlas and axis. In this case, surgical treatment would have required some form of decompression (laminectomy and/or odontoidectomy) followed by occipitocervical fusion. In an effort to avoid the risk of surgical correction of the deformity and to avoid the functional limitation of occipitocervical fusion, conservative therapy was carried out.

Our patient illustrates several points of importance in cases of nontraumatic atlantoaxial dislocation. The occasional association of this condition with rheumatic fever is noteworthy, and the need for early detection of the dislocation is evident if subsequent fibrous fusion in an abnormal position is to be prevented. In atlantoaxial dislocation, it may be possible to achieve a successful reduction by the conservative measures described, even when fibrous fusion has occurred. However, it is not clear whether the increased traction, the steroid therapy, or a combination of both was the key element which succeeded in reducing the dislocation after an earlier trial of traction had failed. It is suggested that steroids may be of benefit in such cases, particularly when fibrous fusion is present.

References

Fig. 3. Lateral cervical radiograph after removal of the Minerva jacket. Flexion and extension radiographs, not demonstrated, revealed no movement of the atlas in respect to the axis.


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