Case Reports

Spontaneous Dislocation of the Atlas

Report of a Case Simulating Syringomyelia with a Discussion of Etiology and Methods of Treatment

PAUL SKOK, M.D.,* JOHN KAPP, M.D., AND CHARLES E. TROLAND, M.D.
Department of Neurological Surgery, Medical College of Virginia, Richmond, Virginia

Spontaneous dislocation of the atlas is an entity discussed infrequently in American medical literature. Its true incidence is difficult to estimate since the dislocation often is attributed to trivial trauma. In addition to the presentation of one such case it is our purpose to discuss possible etiologic factors, to emphasize how such spontaneous dislocation may mimic neurologic syndromes and to discuss the possible avenues of therapy in these cases.

Case Report

No. B-35-86-87. M.B.S., a 32-year-old colored female, was admitted on Nov. 4, 1960 complaining of weakness of all extremities and pain in the neck for 3 months. She had been well until December 1959, when she was operated upon for a retropharyngeal abscess.

Without any antecedent history of trauma she next noted the gradual onset of a stiff neck with inability to turn the head to the right, numbness of both upper extremities, and severe weakness of the arms and hands. She was unable to stand well and complained of unsteadiness. There were no difficulties with bowel or urinary incontinence. She complained of headache radiating from the occipital to the frontal regions; however, there had been no vomiting or visual disturbance. The patient claimed a loss of 19 lbs. in weight because of inability to chew. There was no dysphagia or dysarthria.

Examination. The vital signs were normal. The patient appeared to be an alert, emaciated colored female with severe bitemporal hollowing. There was torticollis to the left. The pupils were round, regular and equal, reacting to light directly and consensually. The extraocular movements were normal except for slight nystagmus on horizontal gaze. The fundi were normal. The neck was markedly twisted to the left and she was unable to turn her head back to the midline. Any attempt to turn the head to the right caused severe pain.

Neurologically there was no demonstrable abnormality of cranial nerves. However, there was hypalgesia from C2 to L3 bilaterally. Below this level pain was well perceived. Sensations of position and touch were intact. There was diffuse weakness, especially severe in the upper extremities, associated with moderate spasticity bilaterally in both lower extremities. However, no clonus was demonstrable and the plantar reflexes were both flexor.

Received for publication June 24, 1963.

* Present address: 277 Alexander St., Rochester, N.Y.

Routine roentgenograms of the cervical spine (Fig. 1) disclosed a severe forward dislocation of the atlas without fracture of the odontoid process. Also noted was a retropharyngeal mass which was thought to be local swelling secondary to the dislocation. The spinal canal measured barely 0.5 cm. from the posterior arch of the atlas to the odontoid.

The impression at the time of admission was spontaneous dislocation of the atlas with symptoms of syringomyelia, secondary to compression of the cord.

Course. The patient was placed in cervical halter traction overnight and by the next morning had a progressively decreasing extent of hypalgesia, which now was present from C4 to L1. There was no appreciable improvement in the weakness.

Operation. Since no further improvement occurred, the patient was operated upon 8 days later. Under general endotracheal anesthesia a decompressive procedure was carried out with removal of the posterior arch of the atlas and a suboccipital craniectomy. The dura mater
was not opened. No traction was applied either at or after the operation.

Course. The next day the sensory level was barely perceptible although there was still no improvement in the motor signs. By the 3rd postoperative day there was no sensory level obtainable at all. The remainder of her course was uneventful with strength noticeably returning by the 2nd postoperative week.

During this time the patient was treated by rest in bed without traction. Thereafter a Zimmer brace was used to maintain the atlanto-axial articulation. It was decided that fusion would be undertaken if in the course of this conservative therapy any further dislocation occurred.

The patient was discharged on Dec. 8, 1960, only to return 3 weeks later with renewed complaints of pain in the neck and weakness. She had not been wearing her brace. Neurological findings were normal but the roentgenograms (Fig. 3) disclosed a more marked dislocation of the atlas with a persistence of the retropharyngeal swelling. Crutchfield tongs were therefore inserted.

The patient was seen by the Department of Oral Surgery who found fistulae and chronic inflammatory changes at the apex of her few remaining roots. These were therefore extracted.

Evaluation of ear, nose and throat failed to disclose any pathology. No significant pharyngeal pathology was noted.

The patient was kept in traction at this time for 8 weeks, following which physiotherapy was instituted.

On March 23, 1961 it was noted that the patient had an abscess on the right side of her neck. This drained spontaneously and cultures yielded acid-fast bacillus. The patient was therefore transferred to the Sanatorium, where she remained until May 14, 1962, undergoing treatment for her tuberculosis with INH, PAS and streptomycin. Although the initial cultures were positive for acid-fast bacillus, all subsequent cultures failed to grow out any acid-fast bacillus. There were no changes on plain roentgenograms compatible with tuberculosis.

During this time, progressive ambulation was accomplished, and the atlanto-axial joint appeared stable. The patient was discharged on May 14, 1962 without a collar or brace.

She was last seen on May 8, 1963, at which time she had regained complete mobility of her neck. There was no pain on motion of the neck and her neurological findings were normal. Roentgenograms at this time showed improved alignment of the atlanto-axial joint which was maintained in the upright position without support (Fig. 3). There were no signs of reactivation of her tuberculosis.

Discussion

The first report of dislocation of the atlas without fracture is attributed to Bell in 1830. Since that time this entity has become a recognized complication of many inflammatory processes in the cervico-occipital area, including upper respiratory infections, otitis media, scrofula, rheumatic fever and rheumatoid arthritis. There are now over 200 cases reported in the literature. However, the fact that these cases are reported by a small number of authors suggests that this entity frequently is undiagnosed. Sharp and Purser found atlanto-axial dislocations in 17.8 per cent of the 79 patients admitted to their Rheumatism Service in 1958 with rheumatoid or Marie-Strumpell arthritis. Bell found atlanto-axial displacement in 40 per cent of 20 autopsy cases of rheumatoid arthritis.

Although seen in all age groups this condition occurs most frequently in children. In Sullivan's...
Fig. 4. View from above of atlanto-odontoid articulation. (A) Odontoid process of axis. (B) Cartilaginous buffer anteriorly and posteriorly. (C) Transverse ligament.

series from the literature 77 per cent were below the age of 13. This is not surprising since inflammatory diseases in this area, that is, tonsillitis, otitis media, etc., are seen more frequently in this age group.

A brief review of the anatomy of the atlantoaxial joint is helpful in considering the mechanism of spontaneous dislocation. The odontoid is surrounded by a ring composed of the anterior arch of the atlas anteriorly and the transverse ligament buffered by cartilage posteriorly (Fig. 4). Normally the transverse ligament exerts no checking effect on the cranivertebral movements. However, it does represent part of this annulus osteofibrosis, and with loss of function of the transverse ligament the atlanto-occipital unit can move in a horizontal direction. 19

Several theories have been proposed to explain this lesion of the transverse ligament. Wittek 20 at the turn of the century proposed a theory of "metastatic effusion" of the ligaments secondary to infection which stretched the ligaments. Jones 8 in 1932 suggested that the vertebra became involved in hyperemia which occurred with an inflammatory process in the neck. This resulted in decalcification of the bone and loosening of the transverse ligament. In 1953 Lippmann 10 demonstrated experimentally that aseptic inflammation about a knee joint of the rabbit caused decalcification of the neighboring bone and severely weakened the adjacent ligaments. He showed that when a ligament was subjected to graded strain it would tear prematurely within the ligament rather than at its insertion into the bone. He noted that weakening of the ligaments occurred before radiographic evidence of bony changes. He proposed no mechanism for this phenomenon. Lourie and Stewart 21 suggested that the prolonged use of steroids in rheumatoid arthritis may contribute to the weakening of the ligaments and their attachments in this area.

Pain in the neck, stiffness and torticollis are the common presenting symptoms. However, in 18 of the 22 patients in Sharp and Purser's 18 series with rheumatoid arthritis the symptoms did not suggest spontaneous dislocation, and the diagnosis was made only on the basis of the routine roentgenographic studies.

Boever and Hennebert estimated that 10 to 15 per cent of atlanto-axial dislocations are associated with signs of compression of the cord below the level of the dislocation. These may take the form of quadriplegia, Brown-Séquard's syndrome, or syringomyelia as in our case, but any combination of cord symptoms is possible. 2

Physical examination may reveal in addition to the neurological abnormalities and cervical tenderness, an abnormal position of the head. Abnormal atlanto-axial mobility may be elicited clinically. Rotation of the atlas with counterrotation of the axis, placing the spinous process of the axis on the normal side (Sudeck's sign), may be found. If the dislocation is pronounced, the anterior arch may be felt as a forward bulge in the posterior pharynx.

The final diagnosis of atlanto-axial dislocation can be only made radiologically. After routine anterior-posterior, lateral and oblique views of the cervical spine have ruled out fracture, lateral films with the neck in flexion and extension should be obtained to show the interval between the anterior arch of the atlas and the odontoid process. The normal interval is still the subject of debate. Jackson 21 stated that the upper limits of normal in adults is 2.5 mm. in flexion and Jacobson and Bleecker 22 felt that up to 5 mm. in flexion is normal in children under the age of 10.

Treatment can be divided into the broad categories of immobilization, fusion, or decompression. Immobilization may be obtained by Crutchfield tongs, braces or plaster casts. Skeletal traction has the added advantage over immobilization of decompressing the cord. Decompression usually consists of laminectomy, with or without suboccipital craniectomy.

The ideal treatment would seem to be to reduce the dislocation with Crutchfield tongs and traction until realignment of the dislocation and clearing of neurological symptoms are achieved. Thereafter, particularly if the realignment proves unstable after an appropriate trial of tongs and traction (usually 6–8 weeks), fusion without laminectomy would seem to be indicated. In our case, laminectomy was resorted to after halter traction seemed to have reached a plateau regarding neurological recovery. In retrospect, it would have been wiser to attempt skeletal traction with Crutchfield tongs before resorting to laminectomy since, following laminectomy, fusion in this region is difficult if not impossible, and without fusion the potentially dangerous situation of an unstable C1-C2 joint is left untreated.
Summary

One example of spontaneous atlanto-axial dislocation is presented with a review of the possible mechanisms involved, the symptomatology of these patients, and the methods of treatment used to date, with suggestions as to the ideal surgical therapy. It is our contention that more awareness of this entity on the part of clinicians will result in the more frequent detection of this removable lesion. It remains an axiom that all patients with spinal-cord disease, regardless of how typical the signs and symptoms are, merit complete roentgenological evaluation.

References