MYELOPATHY CAUSED BY ATLANTO-AXIAL DISLOCATION

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Injury to the upper cervical spine resulting in a separate odontoid process and atlanto-axial dislocation presents a serious problem. The patient may survive the initial episode and the injury may remain undetected and untreated. A sudden recurrence of the deformity may produce signs of damage to the spinal cord, or, over a period of many months or years, evidence of cord damage may slowly develop as the result of recurrent gliding of the atlas on the axis, with consequent narrowing of the vertebral canal.

Corner,* in 1907, describing a series of patients with dislocation of the atlas, recorded the case of a young woman in whom signs of damage to the upper cervical cord developed 1 year after fracturing the odontoid process, with death occurring 14 months following the injury. This author stressed the possibility of delayed myelopathy resulting from abnormality produced by trauma at the atlanto-axial level.

Recognizing the significance of dislocation of the atlas on the axis associated with odontoid fracture, Mixter and Osgood12 in 1910 successfully treated 1 patient by operation in which, by means of a stout braided silk suture, the posterior arch of the atlas was firmly fixed to the spine of the axis.

In 1912, Elliott and Sachs* reported the case of a man, aged 50, who had had a fracture of the odontoid process some 32 years before his final hospital admission, in the authors' words "a long earthly existence with a broken neck." Autopsy revealed a pseudo-arthritis between the body of the axis and the odontoid, the latter being ankylosed with the arch of the atlas. Sections of the spinal cord showed that the crossed pyramidal tracts were completely degenerated and atrophic. Similar spinal cord degeneration was found by List* in 1941, the result of cord compression by abnormalities, such as a loose odontoid, in the region of the foramen magnum and upper cervical spine.

Bachs et al.,† in 1955, reported finding in the literature only 9 cases of delayed myelopathy following atlanto-axial dislocation associated with a displaced odontoid process. These authors added 1 more case to the series but, unfortunately, autopsy was not permitted. However, in recording 2 cases from the literature in which autopsy was performed, mention was made of demyelination, disintegration and glial reaction within the spinal cord and thickening of the pia mater and arachnoid.

The present case likewise demonstrates the extent to which the cord may be damaged by atlanto-axial dislocation, the result of an injury some 25 years before the onset of symptoms.†

CASE REPORT

R.G. (50-6533), a man aged 41, was admitted to the Montreal Neurological Institute on April 26, 1950, referred by Dr. Reuben Rabinovitch. He complained of loss of strength in

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† This case was presented at the meeting of the Halsted Society in Montreal on February 24, 1955.
the right arm and leg, and soreness of the neck. Family and personal history were negative apart from gonorrhoea and syphilis 20 years previously. The man was married, had 7 children, and had been a chauffeur for the past 12 years. At the age of 14, over 25 years before the present admission, while balancing himself upside down on a fence, he fell and landed on his head, knocking himself unconscious for 15 minutes. Following this incident, and until 14 months before this admission, he was quite well except for some limitation in extreme rotation of his neck. Symptoms began with weakness and stiffness in the right lower limb which were followed in a few months by weakness of the right arm. About 4 months before admission the left arm became weak and he observed that the right hand was less sensitive in touching various objects. He was aware of an increasing unsteadiness of gait, which was more marked in the dark. For a period of 1 year he had experienced a sensation of pain and tiredness in the neck and a soreness in the lower back.

Examination. He was a well-developed male who walked in a very stiff-legged fashion on a wide base. There was marked tenderness on palpation of the upper cervical spine and a prominence of the spine of C3. The right pupil was smaller than the left and there was a slight unsustained nystagmus on extreme gaze to the right. There was slight weakness of the right sternomastoid and trapezius. Motor power was decreased throughout, but more markedly so on the right side, particularly in the right hand. There was slight wasting of both hands and forearms, more so on the right. Fine and rapidly alternating movements were done more slowly on the right than on the left. Sensation of touch was intact but there was altered temperature sensation with indistinct limits over the trunk anteriorly and posteriorly. Pain sensation was difficult to evaluate. There was thought to be an area of hyperaesthesia in the C3 distribution. Vibration sense was markedly decreased or almost absent below C3, particularly on the right side. Position sense was surprisingly well maintained, with only slight loss in the toes and more moderate loss in the fingers. The deep tendon reflexes were bilaterally hyperactive, the right slightly more than the left. There was plantar extension bilaterally, with ankle clonus and right wrist clonus. Tone was markedly increased throughout.

Roentgenograms of the cervical spine disclosed a marked atlanto-axial dislocation (Fig. 1A) which could be completely reduced by full extension of the head (Fig. 1B). With the head flexed there was anterior displacement of the atlas on the axis and an obvious narrowing of the cervical canal between the posterior arch of the atlas and the body of the axis. Tomographic films demonstrated a normally shaped dens, separated from the body of the axis by a distance of 3 to 5 mm.

Operation. Atlanto-axial fusion was done under general anaesthesia on April 29, 1950. (In this particular case traction was not applied but subsequent procedures of this type have been done under traction.) The posterior arch of the atlas was wired to the base of the spinous process of the axis and the cartilaginous plates of the articular facets between C1 and C2 were removed. There resulted a better alignment of the posterior arches of the atlas and axis. The intervening dura mater, which before had been stretched tightly, was relaxed and pulsated well. However, at the close of the operation, the anaesthetist reported that the patient's respirations were impaired. When the patient regained consciousness, it was apparent that he was unable to move any of the extremities. Respiration was assisted with the respirator for 48 hours but his condition deteriorated and he died on May 1, 1950.

Autopsy. The specimen of the upper cervical spine obtained at autopsy was cut in the midline, as shown in Fig. 2A. The mechanism by which the upper cervical cord was compressed is readily seen. The loose odontoid, with the pseudo-arthrosis between the dens and the body of the axis, is apparent in Fig. 2B. It is to be noted that, with any degree of movement, not only bone but the associated soft tissues encroach upon the cervical canal and thereby compress the cord. The marked degree of flattening of the cord with loss of substance is seen in Fig. 2A and B. At the level of the lesion the pia-arachnoid is greatly thickened. Above the level of cord compression, at the pyramidal decussation, degeneration can be seen in the lateral spinothalamic tracts and in the posterior columns, bilaterally (Fig. 2C). A cross-section through the lower cervical cord (Fig. 2D) shows marked demyelination in the crossed
Fig. 1. Lateral view of cervical spine, (A) in flexion, showing gliding of the atlas on the axis, and (B) in extension.
FIG. 2. Autopsy specimen of upper cervical spine showing (A) compression of cord, and (B) pseudo-arthritis between the dens and the body of the axis.
Fig. 3. (A, B) Cervical cord at level of cord compression. Weil, X5. (C) Above level of cord compression, at pyramidal decussation; degeneration in lateral spinothalamic tracts and posterior columns bilaterally. Luxol & cresyl violet, X5. (D) Below level of cord compression; degeneration in crossed pyramidal tracts. Luxol & cresyl violet, X5. (E) Thoracic cord; degeneration in crossed pyramidal tracts. Luxol & cresyl violet, X5. (F) Lumbar cord; degeneration in crossed pyramidal tracts. Luxol & cresyl violet, X5.

pyramidal tracts. A similar picture is to be noted in the thoracic cord section (Fig. 3E) and in the lumbar cord section (Fig. 3F).

An incidental finding, at the level of cord compression in Fig. 3A and B, are the several skeins of nerve fibers within what remains of the grey matter. The interlacing network of fibers is shown in greater magnification in the Bodian stained sections in Fig. 4A, B, and E. Adjacent to the skein in Fig. 4C can be seen a group of nerve fibers. A similar bundle of nerve fibers rims the periphery of a blood vessel lying within the anterior median fissure in
Fig. 4. (A) Skein of nerve fibers, at level of cord compression. Bodian, X86. (B) Smaller skeins of nerve fibers. Bodian, X86. (C) Group of nerve fibers adjacent to skein. Bodian, X172. (D) Bundle of nerve fibers rimming blood vessel. Bodian, X172. (E) Interlacing network of nerve fibers within a skein. Bodian, X303. (F) Schwann-like cells associated with nerve fiber skein. Cresyl violet, X172.
DISCUSSION

This patient had a symptom-free interval of approximately 25 years between the accident and the onset of his neurological complaints. It is difficult to know whether the cervical cord lesion with degeneration of ascending and descending tracts resulted from mechanical factors, per se, or whether it followed upon interference with blood supply to this level, thereby producing the degeneration.

Studies of cervical cord compression in cervical spondylosis have shown a similar degeneration of ascending and descending tracts (Bedford, et al.; Brain, et al.). These authors believed that the cord damage resulted from direct pressure, the cord being fixed by the dentate ligaments. Mair and Druckman, on the other hand, concluded that the important factor is compression of the anterior spinal artery, resulting in an impaired blood supply in its distal distribution, i.e., those areas of the cord showing degeneration.

While reviewing sections of the cord in cases of protruded cervical intervertebral discs, Mair and Druckman found evidence of aberrant nerve fibers at the levels of cord compression. These fibers appeared in bundles or large skeins and frequently were covered by Schwann cells. They are identical with those found in the present case, in which cord compression resulted from atlanto-axial dislocation associated with a loose odontoid process. It is thought that these collections of aberrant fibers represent an attempt at regeneration within the injured spinal cord.

The extensive destruction within the cord in this case emphasizes the importance of recognizing odontoid injuries and treating them to prevent the later development of this severe and progressive myelopathy. Congenital abnormality of the upper cervical spine with a separate odontoid process may produce a similar degree of myelopathy. In fact, McRae has stated that it is not possible to decide in an adult whether the radiological picture is that of an old ununited fracture of the odontoid or an anomaly.

Treatment consists of skeletal traction and fusion, as advocated by Cone and Turner. During operation it is probably important to maintain skeletal traction to facilitate the placement of bone wedges between the facets of C1 and C2, thereby keeping taut the redundant soft tissues which otherwise tend to encroach upon the cervical canal. When wound healing is complete, the tong-traction may be removed and the patient may be immobilized in a Minerva plaster.

SUMMARY

Reported is the case of a man in whom motor weakness in the extremities and impaired sensation over the body developed approximately 25 years after a fall on his head which, presumably, fractured the odontoid process. It is believed that the loose odontoid permitted excessive movement at the atlanto-axial joint, resulting in compression of the upper cervical cord and degeneration of both ascending and descending tracts. An incidental finding was the presence of so-called “aberrant regenerating nerve fibers” at the level of cord compression.

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REFERENCES


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