Odontoid compression of the brain stem in a patient with rheumatoid arthritis

Case report

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Cervical spine involvement by rheumatoid arthritis is common; brain-stem compression secondary to vertical subluxation of the odontoid in patients with rheumatoid arthritis is rare. Vertical subluxation results from 1) destruction of the transverse atlantal, apical, and alar ligaments of the atlas and odontoid, and 2) bone resorption in the occipital condyles, lateral masses of the atlas, and basilar processes of the skull. Neurological symptoms result from direct compression of the brain stem or from ischemia secondary to compression of vertebral arteries, anterior spinal arteries, or small perforating arteries of the brain stem and spinal cord. A case is reported in which a slowly progressive neurological deficit developed in a woman with rheumatoid arthritis following a fall from a stretcher. Neurological symptoms represented direct compression of the medulla by the dens, a mechanism confirmed at operation and autopsy.

Recognition of progressive neurological deficit is often difficult in patients with rheumatoid arthritis because of their inactivity and their atrophic and immobile joints, but is essential if appropriate decompressive or stabilizing procedures are to be done. In patients with vertical subluxation of the dens, the transoral approach with removal of the odontoid is recommended. Decompression should be extensive, including the fibrous capsule around the odontoid and overlying synovial tissue as well as the odontoid itself.

KEY WORDS • brain-stem compression • brain-stem decompression • odontoid subluxation • rheumatoid arthritis • cervical spine

Cervical myelopathy secondary to involvement of the cervical spine by rheumatoid arthritis is well documented in the literature, but reports of brain-stem compression as a result of vertical subluxation of C-2 in rheumatoid arthritis are rare.\(^2\),\(^7\),\(^8\) We report a case of such brain-stem compression, demonstrate the importance of early recognition of the lesion, and discuss the mechanism that produces the neurological symptoms.

Case Report

This 62-year-old woman was admitted to North Carolina Baptist Hospital (NCBH) with a history of slowly progressive rheumatoid arthritis since the age of 31 years. She had been confined to a wheelchair for 5 years before admission following a fall from a stretcher. The details of injuries sustained in the fall could not be obtained, but the patient had been told that the base of her skull and neck had been fractured. She had refused therapy at the time. Over the ensuing years, she had slowly lost motor function of both arms; for 8 to 9 months before her admission to NCBH, she had suffered spasms and contractures of both legs. Over the 6 weeks before admission, she had developed progressive bowel and bladder incontinence, difficulty in breathing, loss of ability to sit up, difficulty in swallowing, and intermittently weak and hoarse speech.

Examination. Examination of her head and neck revealed normal facial movement and sensation, normal movement of the tongue with no atrophy, normal
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FIG. 1. Anteroposterior (left) and lateral (right) tomograms of the foramen magnum and posterior fossa demonstrating projection of the odontoid process (arrows) into the posterior fossa and the region of the medulla. Destruction of the posterior arch of C-1 can be seen on the lateral tomogram.

FIG. 2. The ventral surface of the medulla oblongata showing the deep impression made by the odontoid process (arrow) to the left of midline. Postmortem clots are present in the vertebral arteries.

movement of the soft palate, and a normal gag reflex. She was unable to turn her head from side to side, and could not flex or extend her neck.

There was dissolution of the metacarpophalangeal joints with deviation of her fingers to the ulnar side bilaterally. She had complete flaccid paralysis of the upper extremities, and rheumatoid nodules over the extensors of her left elbow. She could voluntarily flex and extend the legs at the hips and the knees, but had no motion of the toes; her ankles could not be moved passively or actively. She had flexion spasms and continuous involuntary adductor spasms of the lower extremities.

There was loss of sensation to pinprick over the trunk from about the T10-12 level to the level of C-5. Reflexes were normal except for the bilateral absence of the Achilles tendon reflex. A plantar response was not obtainable due to joint deformities in the feet and toes.

Tomograms of the cervical spine showed projection of the odontoid into the posterior fossa (Fig. 1). There was a 7-mm anterior subluxation of C-3 on C-4; the available sagittal diameter of the bones across the spinal canal at that level was 11 mm. The posterior arch of C-1 appeared either to be assimilated into the occiput or to be totally eroded and reabsorbed as a result of destruction by the rheumatoid disease.

Vertebral angiography demonstrated a tortuous left vertebral artery but no other abnormal findings. Superimposition of the tomograms on the angiograms demonstrated that the two vertebral arteries at their junction straddled the vertically subluxed dens exactly as a rider's legs straddle the back of a horse.
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On the 4th hospital day, the patient had spontaneous respiratory arrest. An endotracheal tube was placed, and her respirations were controlled by a ventilator. Respiratory arrest was believed to be secondary to compression of the brain stem by the odontoid, and was accompanied by loss of pain sensation in her extremities but by no other change in her neurological status. Cranial computerized tomography (CT) showed the dislocated odontoid pressing on the medulla and pons. The fourth ventricle was not well seen and was believed to be compressed, but there was no evidence of hydrocephalus or of infarction. She subsequently recovered spontaneous respiration and response to pain.

On the 12th hospital day, a tracheostomy was established. Because of her potential long-term survival, but the chance of repeated respiratory arrest due to medullary compression, she was taken to the operating room on the 17th hospital day.

Operation. Through a transoral approach, the anterior portions of the bodies of C-1 and C-2, the lower portion of the clivus, and the odontoid process were removed in that order. A remnant of fibrous capsule around the bone of the odontoid process was not removed because its manipulation created additional pressure against the brain stem.

Postoperatively, the patient could be weaned from the respirator, but her neurological status was otherwise unimproved. On the 27th hospital day, she developed fever, hypotension, and respiratory distress, and died.

Postmortem Examination. At autopsy, a deep impression made by the odontoid process was seen on the anterior (ventral) surface of the medulla oblongata slightly to the left of the midline (Fig. 2). Microscopic sections at that level showed demyelination of the posterior columns (Fig. 3 left) and gliosis of the gracilis and cuneate nuclei. Less marked demyelination was seen in the corticospinal tracts, extending into cervical segments of the spinal cord. No necrotic lesions conforming to territories of vascular supply were observed in the medulla oblongata (Fig. 3 right). The fibrous tissue shell of the odontoid process retained its pyramidal shape after enucleation (Fig. 4 left). The shell varied in width from 3.8 to 4.1 mm (Fig. 4 center), and contained a bone fragment (Fig. 4 right, arrow) at its tip. Foci of hyalinization and calcification were present in the wall, and those, together with its marked thickness, kept the shell from collapsing. Inside the shell, occupying the space from which bone had been removed, was a core filled with organizing blood vessels and clots. Soft tissue sections below the level of the odontoid process showed focal hyalinized nodules and mild perivascular inflammation consistent with rheumatoid arthritis.

Discussion

Rheumatoid arthritis may involve the cervical spine in 86% of patients with the disease. Horizontal subluxation of C-1 on C-2 has been reported in 25% of patients, and vertical subluxation of C-2 in 7% of patients.

Anteroposterior subluxation of C-1 on C-2 commonly compresses the cord at the level of C-1. However, vertical ascension of the odontoid process through the foramen magnum into the posterior fossa
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FIG. 4. Autopsy specimen. × 2.5. Left: The fibrous sheath around the odontoid process is shown. Center: Longitudinal section showing the thickness of the wall and the cavity resulting from operative removal of bone. Right: Microscopic section showing a thick fibrous sheath and a residual spicule of bone at the tip (arrow).

sufficient to compress the brain stem is rare. Vertical subluxation results from 1) destruction of the transverse atlantal, apical, and alar ligaments of the atlas and odontoid, and 2) erosive changes from the rheumatoid process, with accelerated bone resorption in the basilar processes of the skull, the occipital condyles, and the lateral masses of the atlas.1,4

With vertical subluxation of the odontoid, neurological symptoms may be produced by direct compression of the brain stem or by ischemia from compression of the vertebral arteries, anterior spinal arteries, or small perforating vessels of the brain stem and spinal cord.5,4 In the absence of subluxation of the odontoid, the patient with rheumatoid arthritis may develop neurological symptoms due to dural rheumatoid nodules or rheumatoid pachymeningitis.2,7

Nakano, et al.8 reported 32 patients with cervical myelopathy associated with rheumatoid arthritis. Two of the patients had vertical subluxation of C-2, seven had cranial nerve abnormalities, and six with medullary and pontine dysfunction had transient ischemic attacks believed to be caused by intermittent compression of the vertebral artery. Obstruction of the anterior spinal artery or alteration in microvasculature of the spinal cord by anteroposterior compressive forces was suggested as the mechanism of the myelopathy.

Davidson, et al.2 reported a patient with longstanding symptoms of bulbar compression who underwent transoral resection of the odontoid process. The patient enjoyed initial improvement with a halo cast and posterior fusion, but subsequently had recurrence of her symptoms. At operation, a mass of inflammatory tissue surrounding the tip of the upwardly displaced odontoid was seen to be impinging on the anterior surface of the medulla oblongata. Removal of the inflammatory tissue led to an improvement in symptoms, and the authors hypothesized that the symptoms had been caused by direct brain-stem compression from the odontoid and surrounding synovial tissue.

Mayer, et al.7 reported a patient with vertical subluxation of the dens and spinal cord and medullary compression who improved after posterior decompression. However, they did not suggest a specific mechanism for production of the neurological symptoms in their patient.

Our patient's neurological symptoms most likely represented direct compression of the medulla by the dens. Her history of a slowly progressive neurological deficit with an acute increase in that deficit after she had fallen (and was believed to have fractured the base of her skull and probably the odontoid) is suggestive of a compressive mechanism rather than a vascular etiology. The finding of an indented medulla at autopsy and the absence of pathological changes of the types associated with vascular insufficiency also support this mechanism. Eventual erosive changes in the base of the skull, resorption of the occipital condyles and lateral masses of the atlas, and destruction of ligaments from the rheumatoid process caused vertical ascent of the previously fractured dens through the foramen magnum.

Our case and that reported by Davis and Markley4 demonstrate that medullary compression by vertical or horizontal subluxation of the odontoid can be fatal in the patient with rheumatoid arthritis. Recognition of progressive neurological deficit is often difficult in patients with rheumatoid arthritis because the patients are inactive and the joints of their extremities are atrophic and often immobile. However, it is essential to recognize that deficit so that appropriate decompressive or stabilization procedures may be carried out.

The development of brain-stem symptoms, espe-
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cially dysphagia, hoarseness, and respiratory difficulty, is ominous, because it may indicate vertical subluxation of the dens with brain-stem compression. If such vertical subluxation is demonstrated, the transoral approach with removal of the odontoid, as advocated by Greenberg, et al., is recommended, because it permits decompression of the brain stem. As demonstrated in our patient, it is necessary to remove the bone and the overlying synovial tissue and fibrous capsule, since the capsule itself may continue to act as a rigid mass pressing upon the brain stem.

References

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