

ATLANTOAXIAL ROTATORY DISLOCATION WITH HYPOGLOSSAL NERVE PALSY IN A PATIENT WITH ANKYLOSING SPONDYLITIS

A CASE REPORT

BY JUI-TENG CHIEN, MD, ING-HO CHEN, MD, AND KUN-HUEI LIN, MD

Investigation performed at the Department of Orthopedics, Buddhist Tzuchi Dalin General Hospital, Chiayi, Taiwan

Atlantoaxial subluxation is a rare but well-recognized complication of ankylosing spondylitis¹⁻⁷. Anterior subluxation is most common, but vertical subluxation of the dens may also occur and may result in cervical myelopathy, lower cranial nerve palsy, or even sudden death due to cervicomedullary compression⁸⁻¹⁰. Tongue paralysis due to isolated hypoglossal nerve palsy is also rare¹¹⁻¹³. In addition to occurring in patients with rheumatoid arthritis^{14,15}, hypoglossal nerve palsy has been noted in association with a number of pathological conditions such as occipital condylar fracture, metastatic tumor in the skull base, and infection¹⁶. We report on a patient with ankylosing spondylitis who presented with life-threatening atlantoaxial rotatory dislocation and bilateral hypoglossal nerve palsy and was successfully treated with halo-dependent traction followed by occipitocervical arthrodesis. To the best of our knowledge, bilateral hypoglossal nerve palsy with tongue paralysis arising from atlantoaxial rotatory dislocation has never been reported in a patient with ankylosing spondylitis. The patient consented to have the data concerning the case submitted for publication.

Case Report

In October 2001, a forty-nine-year-old man with a thirty-year history of ankylosing spondylitis presented with severe torticollis, a chin-on-chest deformity, and a protruded tongue. These conditions had developed three months before admission and had progressed rapidly over the preceding four weeks. The patient reported no history of trauma to the head or neck in the preceding months.

Neurological examination revealed generalized hyperreflexia, sustained ankle clonus, and positive Babinski signs. Muscle power was decreased (grade 3 of 5) throughout both the upper and lower extremities. The bowel and bladder sphincters were not affected. In addition to the spinal deformity, the most striking finding was that the tongue protruded, was immobile despite voluntary efforts to move it, and was coated with thick layers of whitish secretions. No wasting or

fasciculation was detected. The patient was unable to speak, chew, or swallow and he continuously drooled saliva. Because of the tilt and rotation of the head, the face was so disfigured that the patient could not raise the eyelid of the right eye. Nutrition was provided with nasogastric feeding, and the airway was secured by constant suction.

A lateral plain radiograph showed severe anterior dislocation of the atlas on the axis that markedly compromised the spinal canal (Fig. 1-A). A sagittal reconstruction of a computed tomographic scan revealed marked anterior and vertical migration of the dens with substantial cervicomedullary compression (Fig. 1-B). A coronal computed tomographic reconstruction showed extensive erosion of the C1-C2 facet joints with marked rotatory subluxation (Fig. 1-C).

We decided to gradually correct the multiple-plane deformity. First, halo-dependent traction was used with 2 kg of weight applied anterolaterally to correct the rotational displacement. Three days later, 2 kg of vertical traction was added anteriorly to correct the chin-on-chest deformity while the anterolateral traction was decreased to 1.5 kg to maintain the corrected position. On the thirteenth day, the patient was able to retract the tongue completely and open the right eye, as the head tilt and rotation had been corrected. He could move the tongue freely and was able to speak again. Sixteen days following the commencement of traction, he underwent craniocervical arthrodesis from the occiput to C6 with the use of allografts, plates, and wires. Following reduction and surgery, the signs of cervical myelopathy (weakness and clonus) and hypoglossal nerve palsy (dysphagia, dysarthria, and dysphonia) all rapidly resolved. The patient was able to eat solid foods without choking and could articulate clearly. Two weeks after the craniocervical arthrodesis, lumbar osteotomies were performed to correct a thoracolumbar kyphosis. He continued to wear a body cast that incorporated the halo ring for a total of 4.5 months. At 2.5 years after surgery, he was able to walk with the aid of a walker, and radiographs confirmed a solid occipitocervical fusion without loss of correction (Fig. 2).



Fig. 1-A
Lateral radiograph demonstrating marked anterior dislocation of C1 on C2 and severe ankylosis of the subaxial cervical spine.

Discussion

The precise mechanism of the hypoglossal nerve palsy in our patient is not known. It may have been caused by either mechanical traction on or vascular insufficiency of the hypoglossal nerve. The hypoglossal nerve exits the skull base anterior to the occipital condyle. It then travels through the hypoglossal foramen, following a vertical course inferiorly just lateral to the jugular foramen to cross the anterior aspect of the lateral masses of C1-C2¹⁷. Macedo et al.¹⁵ reported two cases of bilateral hypoglossal nerve palsy due to vertical migration of the dens as a result of late-stage rheumatoid arthritis. They proposed that the hypoglossal nerves were directly compressed at the level of the C1 transverse process. Kenrick et al.¹³ contended that anterior subluxation of C1 could injure the hypoglossal nerves bilaterally as they exit from their canals while not affecting cranial nerves IX, X, and XI, which emerge from the more lateral jugular foramen. Shim et al.⁹ reported on a patient with ankylosing spondylitis who had vertical migration of the dens, and they concluded that vascular insufficiency was the main cause of the medullary ischemia and bulbar symptoms. In our patient, the signs and symptoms may have been due to mechanical traction on the hypoglossal nerves. When the head tilt, rotation, and chin-on-chest deformity first developed, the patient complained of a thick tongue with slurred speech. As the deformity progressed, the paralyzed tongue was gradually forced out of the mouth. Subsequently, the airway became markedly obstructed, placing the patient at risk for suffocation and aspira-

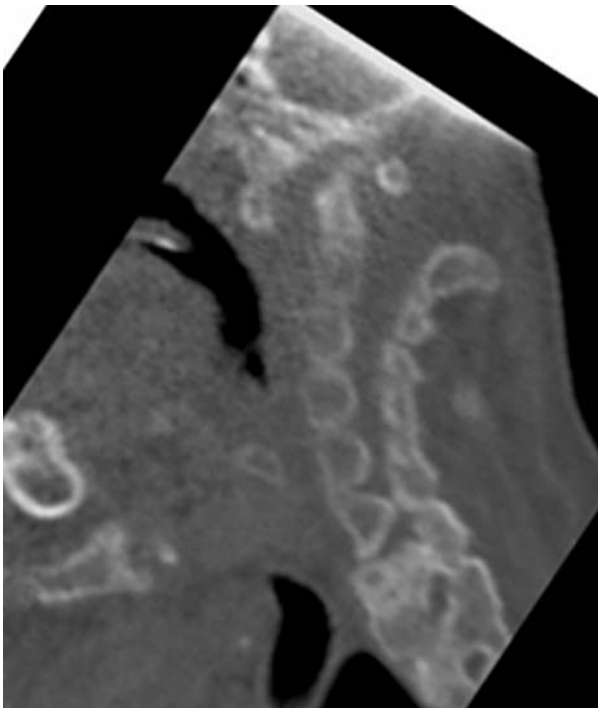


Fig. 1-B

Fig. 1-B Sagittal computed tomographic reconstruction demonstrating anterior subluxation and vertical migration of the dens with marked cervicomedullary compression. The anterior atlantodental interval measures 14 mm. **Fig. 1-C** Coronal computed tomographic reconstruction showing extensive erosion of the C1-C2 facet joints with marked rotatory subluxation.

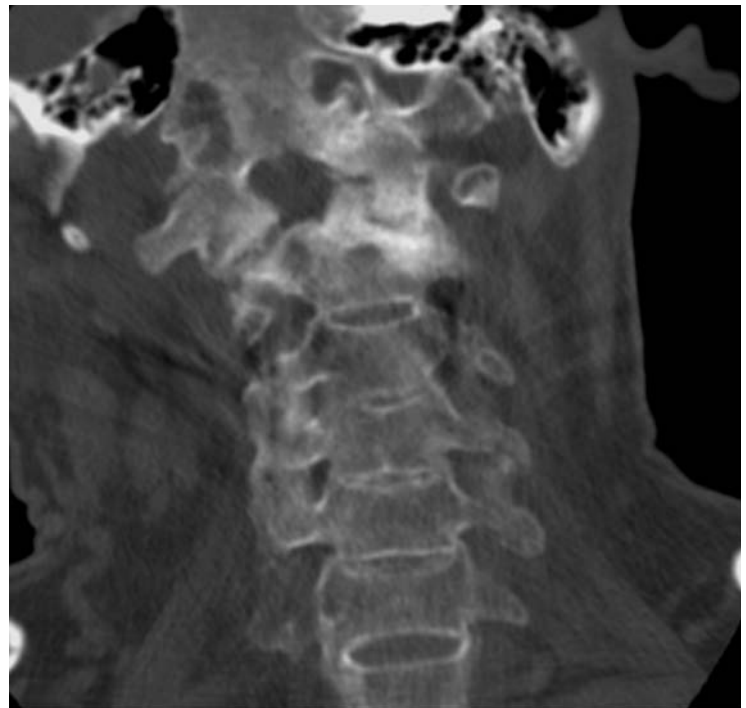


Fig. 1-C



Fig. 2
Lateral radiograph made 2.5 years postoperatively, demonstrating a solid occipitocervical fusion in satisfactory alignment.

tion. Soon after successful reduction was accomplished with halo traction, he was able to retract the tongue and the dysphagia and dysarthria improved dramatically, suggesting that the tension on the hypoglossal nerve had been relieved.

Graziano et al.^{18,19} and Mehdiian et al.²⁰ reported successful reduction of cervical deformity with halo traction in patients with rheumatoid arthritis and ankylosing spondylitis. Halo traction was used for our patient for several reasons. First, it made it possible to address the devastating deformity with readily available equipment. Second, judging from the disease course, the dislocation had been present for a relatively

short duration (four weeks at the most), suggesting that the deformity was still supple. Third, reduction in the presence of marked atlantoaxial rotatory dislocation can be risky²¹ and can be hampered by difficulty with intubation²². Accordingly, the rotational deformity of the head was first corrected with the anterolateral traction, after which the vertical traction was applied to correct the anterior and upward displacement of the dens. During the application of the traction, the patient was monitored closely with serial neurological examinations to avoid additional cranial nerve or cord injury²³. The weight and the vector force of the traction were adjusted frequently according to the extent of the reduction and the patient's tolerance. We placed emphasis on the duration and vector force of the traction rather than simply increasing longitudinal traction to extend and derotate the neck.

In summary, gradual correction with halo-dependent traction followed by occipitocervical arthrodesis was a safe and effective method for treating this life-threatening atlantoaxial rotatory dislocation. ■

Jui-Teng Chien, MD

Kun-Huei Lin, MD

Department of Orthopedics, Buddhist Tzuchi Dalin General Hospital, 2, Min-Sheng Road, Dalin Town, Chiayi 622, Taiwan. E-mail address for J.-T. Chien: jtchien@mail2000.com.tw

Ing-Ho Chen, MD

Department of Orthopedics, Buddhist Tzuchi Medical Center, 707, Section 3, Chung-Yang Road, Hualien 970, Taiwan

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