Although atlantoaxial rotatory subluxation is a well-known entity that affects the pediatric population, combined atlantoaxial and occipitoatlantal subluxation is a rare disease. We describe a new case in a teenage girl.

Case Report

History. This 17-year-old girl was admitted to our department with a history of torticollis. She presented with severe neck pain, having experienced typical spontaneous torticollis a few days before admission and received analgesic drug therapy for 3 days. After that, she underwent an unspecified chiropractic manipulation that she refused to describe.

Examination. At clinical examination, her head was slightly rotated to the left and tilted on the right (Fig. 1), but there was no real torticollis. Results of the neurological examination were otherwise normal.

Plain x-ray films (Fig. 2) revealed an abnormal location of the lateral mass of C-1 and the transverse process. A three-dimensional (3-D) computerized tomography (CT) study (Fig. 3) revealed double axial subluxation, with C-1 turned to the left and C-2 to the right, which perfectly explained the clinical presentation. Magnetic resonance imaging demonstrated compromise of the spinal canal and normal transverse ligament. The results of vertebral angiography were normal.

Operation. An initial attempt to reduce the double subluxation was made after induction of a general anesthetic to produce pharmacological paralysis. However, fluoroscopically guided cranial traction did not produce any mobilization of C-1 or C-2. Thus, we concluded that extensive lesions of the ligament must have occurred that caused double instability of occiput (Oc)–C1 and C1–2. An open operative approach was required.

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Key Words • atlantoaxial rotatory subluxation • occipitoatlantal subluxation • three-dimensional computerized tomography

Fig. 1. Photograph showing the appearance of combined atlantoaxial and occipitoatlantal subluxation in our patient at presentation. The head is slightly rotated on the left side and tilted on the right.
Occiput–C2 double subluxation

approach was undertaken in which reduction was performed aided by fluoroscopic guidance under direct visualization. Reduction was accomplished using cranial traction and rotation of C-1 with bone forceps. An Oc–C2 arthrodesis was performed in which tricortical iliac bone graft and stainless CCD rods and hooks (Sofamor Danek, Rang du Fliers, France) were used.

Postoperative Course. The postoperative course was uneventful. Evaluation of plain x-ray films revealed that satisfactory reduction had been accomplished (Fig. 4).

Discussion

A literature review in which the Medline database was searched identified four other cases of combined atlantoaxial occipitatoatlantal subluxation.1,2,5,6 The first case was described by Washington.6 The oldest patient was a 27-year-old woman.6 At clinical examination, patients with only atlantoaxial rotatory subluxation present with torticolis. This has been well described by Fielding and Hawkins,4 who called it the “cock robin position.” In patients with combined atlantoaxial occipitatoatlantal subluxation, the head is gently tilted and slightly rotated (Fig. 1). Rotation of the head to the opposite side is impossible and is limited and painful to the same side (the left side in our case). In all the cases, results of neurological examination were normal.1,2,4,6

In our case, the exact pathophysiological mechanism of the condition is unknown, but it was probably caused by a violent motion in rotation and traction that caused a “compensatory” contralateral subluxing of Oc–C1 onto the initial C1–2 subluxation, as Clark and colleagues2 have described. In the literature, three cases of subluxation occurred posttrauma and one was spontaneous, but the exact pathophysiological mechanisms were never adequately described.

The findings in our report are the first to be obtained using 3-D CT scanning. This technique elucidated the 3-D anatomical location of each vertebra and demonstrated compromise of the spinal canal. Magnetic resonance imaging confirmed the spinal cord compression and demonstrated the anatomical integrity of the transverse ligament on axial T2-weighted images, as previously described by Dickman, et al.3 The vertebral arteries were normal in our case; this feature was not described in the other cases.1,2,4,6

Two types of therapy could be proposed: cervical traction, with reduction and halo cast, or an operative procedure in which an Oc–C2 arthrodesis is performed.

After we failed to achieve reduction in our patient by using fluoroscopic guidance, we decided to perform an Oc–C2 arthrodesis in which CCD rods and hooks were

Fig. 2. Left: Plain x-ray film, lateral view, obtained in the cervical spine demonstrating rotation of C-1, with unusual visibility of transverse process. Right: Plain x-ray film, anteroposterior view (open mouth) of the cervical spine demonstrating left rotation of C-1 lateral masses.

Fig. 3. Three-dimensional CT studies. Left: Inferior view revealing double axial rotation of C-1 and C-2. Note the impairment of the spinal canal and the midline position of the head. Right: Anterior view.
used. This allowed us to make a rigid construction with strong bone graft compression and to treat the double cranio-cervical instability.

In the literature, only one case, in 1959, was treated by a procedure in which external immobilization was used. In one other case, a C1–2 arthrodesis was performed and the patient was placed in a halo vest. The other two patients were treated by undergoing Oc–C2 arthrodesis.

References


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