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CASE REPORT

Nontraumatic posterior atlantooccipital dislocation associated with atlantoaxial instability

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Abstract
Introduction Nontraumatic posterior atlantooccipital dislocation has only been rarely reported. In the current study, the authors reported an extremely rare case of nontraumatic posterior atlantooccipital dislocation associated with atlantoaxial instability.
Materials and methods A 47-year-old female was referred with a history of neck pain for 5 years. The patient had no history of trauma. The axial rotation of range of motion of the cervical spine was severely restricted. Posterior atlantooccipital dislocation with atlantoaxial instability was confirmed through conventional radiography, computed tomography and magnetic resonance imaging. We performed realignment of the dislocation and posterior occipitocervical (C0–C2) fusion. After the surgery, the patient's symptoms improved significantly and she manifested neurological improvement.
Conclusion To our knowledge, this lesion has not been reported previously. Anomalies of upper cervical spine may have induced this instability.

Keywords Nontraumatic · Atlantooccipital joint · Atlantoaxial joint · Dislocation

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Introduction

Combined nontraumatic atlantooccipital and atlantoaxial instability resulting from any cause is extremely rare. This disorder is often related to Down's syndrome [1], rheumatoid arthritis [2], tumor of the atlas [3], infection in the head or neck [4], or torticollis [5]. We present here a new case of posterior atlantooccipital dislocation associated with atlantoaxial instability who did not suffer from the above-mentioned diseases. We explain our treatment and management for this rare case.

Case report

A 47-year-old female patient, who had no history of trauma, rheumatoid arthritis, psoriasis, tumor, infection in the neck and torticollis, complained of considerable pain and stiffness in the neck for 5 years. The axial rotation of the range of motion of her cervical spine was severely restricted, and she did not move her head position spontaneously, since it induced deterioration of pain. She did not describe any improvement of symptoms in the flexion position, and experienced deterioration in the extension position. Neurological examination revealed diminished sensation to light touch bilaterally in the upper and lower extremities in all dermatomes. Her muscle strength was Grade 4 in all major muscle groups in the bilateral upper and lower extremities. She demonstrated hyperreflexia and spasticity bilaterally in the upper and lower extremities. Her sphincter function was normal. A positive Hoffman's sign and inverted radial reflex were noted bilaterally. Babinski test revealed down-going toes. She could walk without assistive devices, but her gait was shuffling. Dynamic lateral cervical radiographs showed instability of

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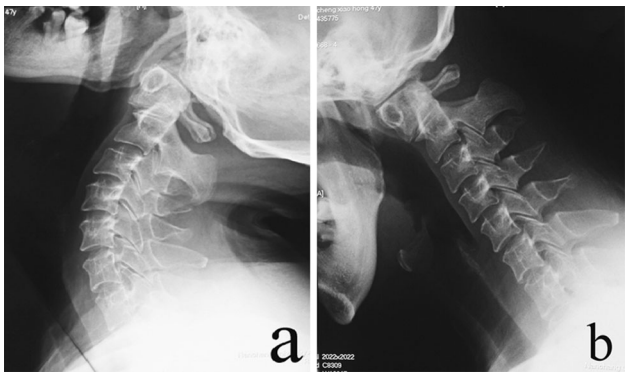

Fig. 1 The cervical spine motion is shown in extension (**a**) and flexion (**b**) on lateral radiographs. A combined atlantooccipital and atlantoaxial articulation instability is shown

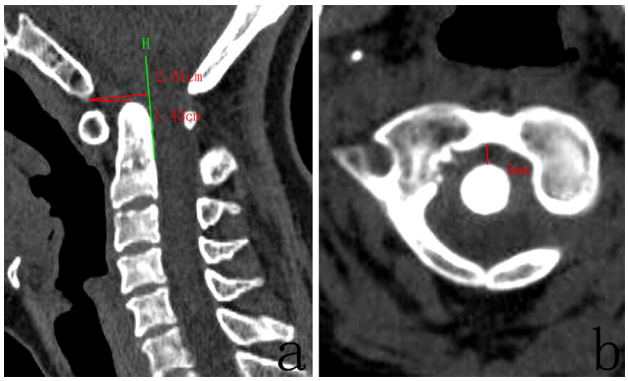

Fig. 2 Sagittal CT reconstruction **a** demonstrating exceeding of Harris Rule of Twelve (the basion-dens interval = 14 mm, basion-axial interval = 20 mm). Axial CT reconstruction **b** demonstrating dislocation of atlantoaxial (AAI = 5 mm)

atlantooccipital joint with posterior dislocation in extension and reduction in flexion, and instability of atlantoaxial joint with increased anterior atlantodental interval (AAI) in flexion than in extension (Fig. 1a, b). Sagittal and axial reconstruction views of CT images further confirmed a combined atlantooccipital and atlantoaxial dislocation (Fig. 2a, b). A coronal view on a computed tomography (CT) reconstruction image showed a loss of angle of the bilateral atlantooccipital joint, and a sagittal reconstruction view of CT images also demonstrated a flatness of the atlantooccipital joint (Fig. 3a–c). Spinal cord compression and signs of myelopathy due to chronic instability were detected on MR images (Fig. 4a, b). Surgical intervention was indicated because of the spinal cord compression and signs of myelopathy due to chronic instability. We put the patient in the prone position under general anesthesia, and examined her under an image intensifier to get a reduction between the occipital bone and the axis by Mayfield skull holder (Fig. 5a, b). A posterior fusion was performed from the occipital bone to C2 using the occipitocervical fixation system [6] (made in China, a combination of C2 pedicle screws and plates) performed with a bone graft from the posterior iliac bone. Cervical radiographs 1 year after surgery showed the reduction of the atlantooccipital joint and stability of occipitoatlantoaxial complex (Fig. 6a–c). A three-dimensional reconstruction image on CT clearly demonstrated the fusion mass from the occipital bone to C2 (Fig. 7). A magnetic resonance image suggests that the damaged spinal cord was gradually restored (Fig. 8a, b). The patient got a significant relief of neck pain and improvement in motor function of the upper and lower extremities. The patient was able to move her head much better compared to before surgery, but her movement was still restricted.

Discussion

There are numerous reports of patients showing posterior atlantooccipital dislocation due to trauma [7, 8]. This disorder is often associated with fatal results. However,

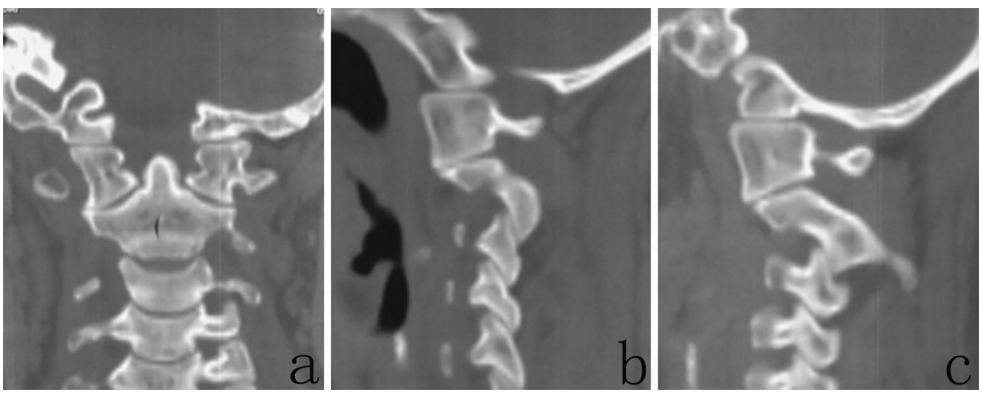

Fig. 3 Computed tomography on a coronal reconstruction view **a** showed a loss of slope of the atlantooccipital joint. That on sagittal reconstruction views (**b** Rt, **c** Lt) also showed a flatness of the atlantooccipital joint on both sides

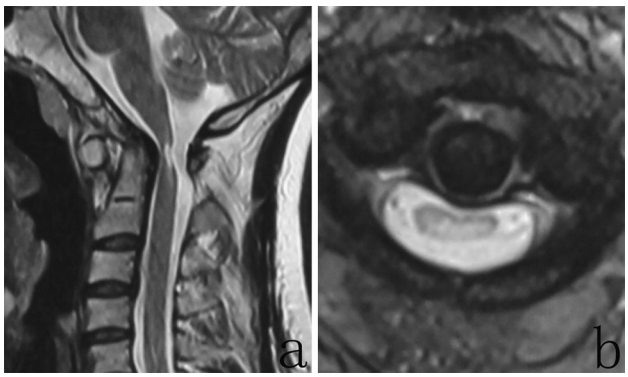

Fig. 4 Magnetic resonance image before surgery, sagittal T2 (**a**), and axial T2 (**b**)-weighted images of the cervical spine showing spinal cord compression and intramedullary cord signal abnormality at the level of the atlantooccipitoaxial region

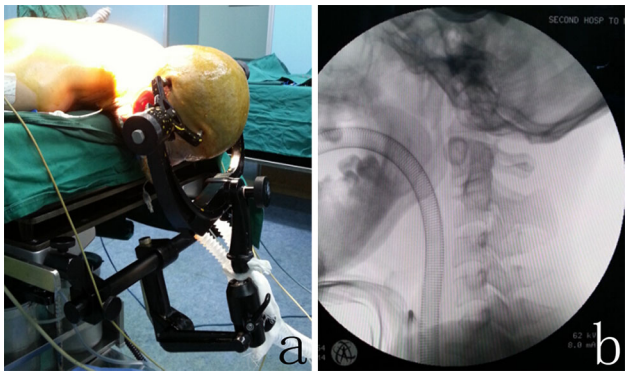

Fig. 5 The patient is shown in the prone position following Mayfield skull holder (**a**). Perioperative sign of reduction between the occipital bone and the axis during the positioning (**b**)

nontraumatic posterior atlantooccipital joint dislocation has only been rarely reported. Abumi et al. [9] reported a case of nontraumatic rotatory posterior atlantooccipital joint subluxation which was presumed to have derived from articular tropism. Takechi et al. [10] reported a case of nontraumatic atlantooccipital joint dislocation. And they proposed that flatness of the bilateral atlantooccipital joint may induce this instability. To our knowledge, to date, the nontraumatic posterior atlantooccipital dislocation associated with atlantoaxial instability has not been reported in the literatures published.

The etiologies of nontraumatic occipitoatlantoaxial complex instability are still unclear. Wiesel et al. [11] proposed that a high cervical fusion produces more stress at the atlantooccipital articulation which gradually causes the ligaments to stretch, resulting in hypermobility. But there was no hypermobility or dislocation of atlantooccipital joint in 150 cases of Klippel–Feil syndrome reviewed by Georgopoulos et al. [12] According to previously reported studies, atlantooccipital instability related to Down's syndrome [1], rheumatoid arthritis [2], or tumor of the atlas [3] is always associated with atlantoaxial instability. Washington [4] reported a nontraumatic atlantooccipital and atlantoaxial dislocation in an 11-year-old girl due to pharyngitis, and they agreed that an inflammatory process of the joint and its periarticular structures associated with the instability. But the female in our study was excluded from the above-mentioned diseases.

It is worth noting that in our case in the upper cervical spine exist osseous anomalies, same as the case of Takechi et al. [10]. The anterior arch of the atlas was hypertrophic and the bilateral atlantooccipital joint was flat but ball-and-socket shaped, which could prevent dislocation of the occipital condyle forward or backward. We agreed that anomalies of upper cervical spine such as flatness of the bilateral atlantooccipital joint may account for this instability, which also explained why most atlantooccipital dislocations occur in children whose occipital condyles are very small and the atlantooccipital joint relatively horizontal. We speculated that abnormal movement of atlantooccipital joint would increase load of the atlantoaxial

Fig. 6 Lateral cervical spine radiography in neutral (**a**), flexion (**b**) and extension (**c**) demonstrated no instability of the atlantooccipitoaxial region 1 year after the operation

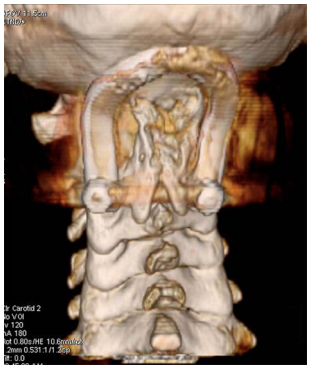

Fig. 7 Computed tomography on a three-dimensional reconstruction view demonstrating a good fusion mass from the occipital bone to C2

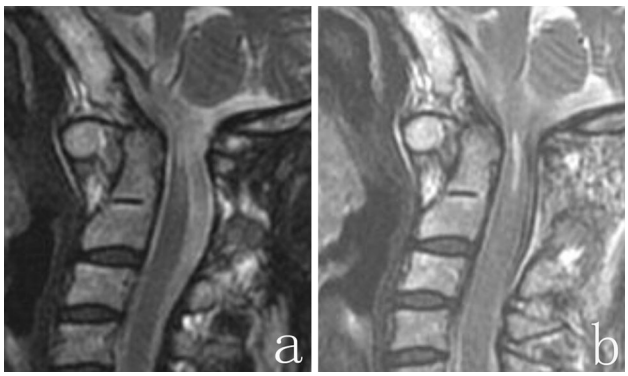

Fig. 8 MRI showed a restoration of the damaged spinal cord 1 year after operation (**a**) compared to 1 week after operation (**b**)

joint, resulting in laxity of atlas transverse ligament and causing atlantoaxial instability.

It is universally agreed that instability of the occipitocervical region should be stabilized surgically. The method of choice in atlantooccipital dislocation operative treatment is posterior occipitocervical fusion [13]. Abumi et al. [9] applied posterior atlantooccipital fusion using wiring with a bone graft. Takechi et al. [10] applied posterior atlantooccipital fusion using the Axon system (Synthes) performed with a bone graft from the posterior iliac bone. There are also other options being described, Ogihara et al. [14] proved the efficacy of the halo vest in preoperative reduction. This should be considered, if the definitive procedure is postponed due to the severe general health status of the patient. Occipital–cervical fusion (C0–C2) using the occipitocervical fixation system was chosen in the present case since the patient showed restricted axial rotation of the neck before surgery. After the surgery, MR images found that the lesion of spinal cord presented obvious improvement. Meanwhile, the patient's pain of cervical vertebrae improved significantly and muscle strength restored to normal degree.

Conflict of interest None of the authors has any potential conflict of interest.

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