

Case Report

A Morphologically Atypical Case of Atlantoaxial Rotatory Subluxation

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A rare case of atlantoaxial rotatory subluxation occurred after pediatric cervical spine surgery performed to remove a dumbbell-shaped meningioma at the level of the C1/C2 vertebrae. This case is classified as a post-surgical atlantoaxial rotatory subluxation, but has a very rare morphology that has not previously been reported. Although there are several reports about post-surgical atlantoaxial rotatory subluxation, an important point of this case is that it might be directly related to the spinal cord surgery in C1/C2 level. On day 6 after surgery, the patient presented with the Cock Robin position, and a computed tomography scan revealed a normal type of atlantoaxial rotatory subluxation. Manual reduction was performed followed by external fixation with a neck collar. About 7 months after the first surgery, the subluxation became severe, irreducible, and assumed an atypical form where the anterior tubercle of C1 migrated to a cranial position, and the posterior tubercle of C1 and the occipital bone leaned in a caudal direction. The pathogenic process suggested deformity of the occipital condyle and bilateral C2 superior facets with atlantooccipital subluxation. A second operation for reduction and fixation was performed, and the subluxation was stabilized by posterior fixation. We encountered an unusual case of a refractory subluxation that was associated with an atypical deformity of the upper spine. The case was successfully managed by posterior fixation.

Key Words : Atlantoaxial rotatory subluxation · Atlantooccipital dislocation · Facet deformity.

INTRODUCTION

Atlantoaxial rotatory subluxation (AARS) is an infrequent condition that most commonly occurs in children. Its causes are unknown, but risks include pediatric surgery, otopharyngeal inflammation, trauma, and extreme rotation of the head^{2,8)}. It is particularly common after surgeries requiring marked lateral rotation of the head and extension of the neck during positioning⁹⁾. Some researchers have reported that AARS is a result of increased laxity of the capsular structure and alar and transverse ligaments¹⁾. Following atlantoaxial subluxation, lateral facet joints are gradually worn away. As a result, the dens migrates in a vertical direction (vertical subluxation) and movement is restricted. Surgery-related AARS accounts for about 31% of non-traumatic AARS⁷⁾. Almost all cases of AARS occur within 1 week of surgery, not immediately after surgery.

About 68% of non-traumatic AARS occur in patients aged less than 12 years⁹⁾. In children, the ligaments and joint capsules are sufficiently elastic to allow hypermobility without disruption, and C1-2, in particular, are shallower and more horizon-

tally oriented⁸⁾. Adding to these conditions, the disproportion between the size of the child's head and a hypermobile cervical spine with minimal nuchal muscle may predispose the atlantoaxial complex to AARS.

AARS is often relatively easy to reduce by using a closed reduction method and conservative therapies such as traction and orthoses. However, some cases recur and thus require strict conservative treatment and surgical therapy because recurrent cases differ from the initial cases that were intractable to treatment. Patients with long-standing subluxations are more likely to experience recurrence and to require surgery⁸⁾.

We encountered an unusual case of a recurrent subluxation that led to an atypical deformity in the upper spine. We believe it contains important information about AARS because of its rarity and the experience of successful management.

CASE REPORT

Past history and present illness

A 6-year-old girl presented with neck pain and ataxia. She

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had undergone surgery for maxillary gum and subcutaneous tumors that were pathologically diagnosed as neurofibromatosis (NF). Magnetic resonance imaging (MRI) revealed bilateral internal auditory canal tumors, a right optic nerve sheath tumor, and an extramedullary spinal tumor on the right side of the occipital bone at the C1-2 level (Fig. 1). She was diagnosed with NF 2, and referred to our hospital.

Examination

Neurological examination on admission showed no abnormalities except slight ataxia. She didn't have café au lait macules or other signs characteristic of NF 1. Preoperative radiographic images revealed no abnormal deformity of the cervical spine.

First operation

We scheduled a surgery for total removal of the spinal tumor, which was thought to be the cause of her neck pain and ataxia. The operation was performed under general anesthesia with the patient prone with her neck in an anteflex position. A skin incision of about 7 cm was made, and the nuchal fascia was exposed on the right side. The right upper part of the C2 spinous process was cut, and the oblique capitis inferior muscle and the rectus capitis posterior major muscle were retracted laterally. The trapezius and rectus capitis posterior minor were partially dissected from the occipital bone. The C2 lamina, C1 posterior arch, and the occipital bone were exposed only on the right side. A C1 hemilaminectomy was performed, and the upper parts of the C2 lamina and occipital bone were partially removed. The tumor was visible on the dura and at the right side of the spinal canal. Gross total removal of the tumor was achieved. Closure was performed according to standard protocols. Histological examination revealed a grade 1 meningothelial meningioma, as determined according to the World Health Organization classification criteria. Reports of dumbbell-shaped meningiomas in NF patients are very rare. The tumor was difficult to distinguish from a Schwannoma, which is the most common spinal tumor associated with NF, and this distinction will be discussed elsewhere.

Postoperative course

The postoperative course was uneventful, and no neurological deficit was observed. But on day 6 after surgery, the patient experienced severe neck pain and assumed the Cock Robin posture. Computed tomography (CT) scan revealed an AARS. We sub-

sequently performed a manual reduction followed by external fixation with a neck collar. However, the AARS was refractory and intractable to treatment. About 7 months after the first surgery, it became severe and irreducible. The girl complained of neck pain, and she could not flex her neck. This was an atypical form of subluxation in which the anterior tubercle of C1 had migrated to a cranial position, and the posterior tubercle of C1 had migrated caudally with the occipital bone also leaning in a caudal direction (Fig. 2). The pathogenic process suggested deformity of the occipital condyle and C2 facet; the Fielding classification was type 1 because there was no anterior displacement of the C1 vertebra (Fig. 3, 4).

Several measurements were made using the sagittal view of the cervical spine on the CT images. The atlantodental interval (ADI) was 1.94, the Powers ratio 1.26, the basion-axial interval (BAI) 1.16, and the basion-dens interval (BDI) 2.27. There was no basilar invagination because the Chamberlain and McGregor lines were normal. Cervical alignment was kyphotic with a C2-7 angle of -20.3° . An atlantooccipital dislocation was diagnosed because of the abnormal Powers ratio, but the BAI and BDI were normal. A 3-dimensional CT scan showed an anterior dislocation of the bilateral occipital condyles from the superior articular facets of C1. The clivo-axial angle was 160° . A lateral inclination of $<20^\circ$ was observed.

It is important to diagnose and treat atlantooccipital dislocations as quickly as possible because neurologic injuries, without treatment, can be devastating³. According to the ADI score, there

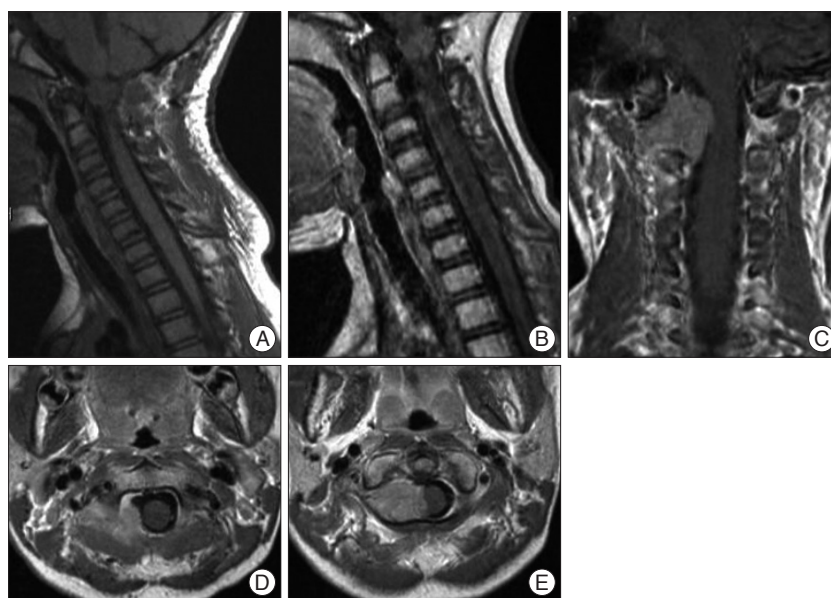


Fig. 1. Magnetic resonance imaging (MRI). A : Sagittal T1 weighted image. B : Sagittal T1 weighted image with gadolinium enhancement. C : Coronal T1 weighted image with gadolinium enhancement. D : Axial T1 weighted image with gadolinium enhancement at upper rim of C1 level. E : Axial T1 weighted image with gadolinium enhancement at lower rim of C1 level. The tumor seems to be intradural extramedullary tumor without dural tail sign, and its shape is semiovale. The tumor is iso signal intensity in T1 weighted image, and homogeneously enhanced with gadolinium (B–E). These images show a dumbbell shaped spinal tumor, which is intradural extramedullary tumor at right foramen of C2 root (C). It is located in the right side of the spinal canal at C1/2 level and spinal cord is deviated to left due to the compression of tumor (D and E).

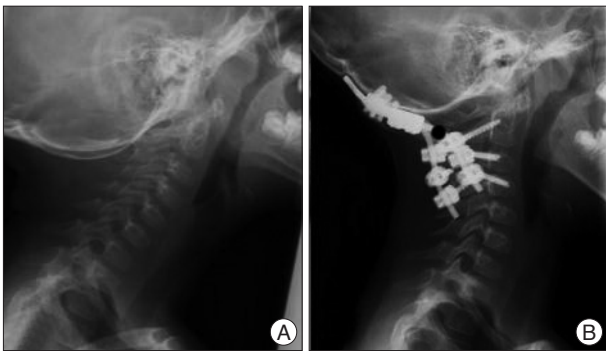


Fig. 2. A : Plain radiograph of the cervical spine shows an atypical form of subluxation in which the anterior tubercle of C1 had migrated to a cranial position, the posterior tubercle of C1 had migrated in a caudal direction, and the occipital bone began to lean caudally. B : The postoperative radiograph shows occipital plate C2-C3-C4 bilateral screw fixation and recovery of cervical lordosis.

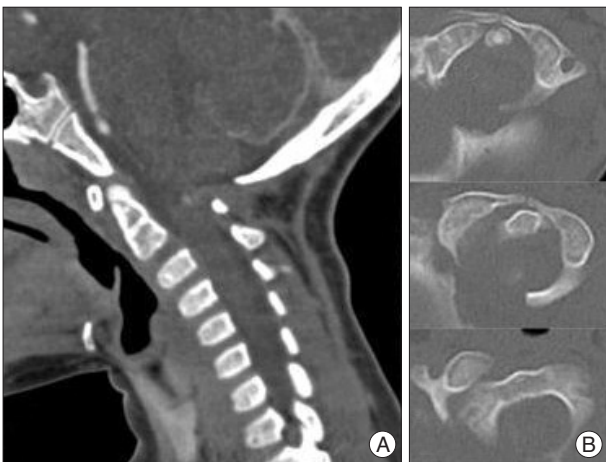


Fig. 3. Computed tomography images of the cervical spine. A : Sagittal image showing the kyphotic cervical alignment. B : Axial images showing that the Fielding classification was type 1.

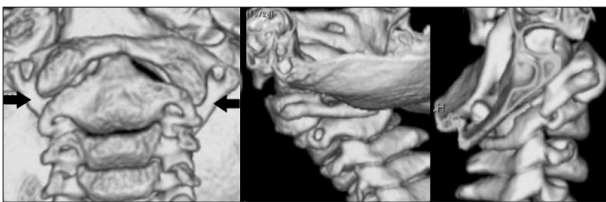


Fig. 4. Three-dimensional computed tomography images of the cervical spine show deformities of the C2 superior facet (arrow), and that the C1 vertebra was rotated to the left.

was no apparent AAS. However, the axial images revealed AARS.

Second operation

We performed a second surgery under navigation guided for reduction and occipital plate C2-C3-C4 bilateral screw fixation. Under general anesthesia, the patient was placed in a prone position and lateral radiography was performed. We tried to reduce C1 dislocation, and a partial reduction was achieved. The vertebral level was identified. The C4, C3, C2, and C1 (left half) laminae and spinous processes were exposed. At the right side

of C3, a pedicle screw was successfully inserted. At the left side of C3, the lateral mass was fragile and a spinous process screw was inserted. We also added a C4 lateral mass screw to the left side and a pedicle screw to the right side. At C2, an isthmic screw was inserted into the left side and a pedicle screw into the right side. The occipital plate was applied and fixed in place with 3 screws, measuring 6, 10, and 6 mm. Two rods were applied and fixed together. The compression force was applied between the C2 and C4 screws. The occipital bone was harvested as an autograft for Occiput-C3 fusion.

The subluxation was stabilized by posterior fixation (Fig. 5). After surgery, the clivo-axial angle improved from 160° to 125°. The postoperative course was uneventful, and the girl was discharged without complications. There has been no recurrence for the past 3 years.

DISCUSSION

Fielding and Hawkins classified AARS into 4 different types : Type I, rotatory fixation with no anterior displacement, with the odontoid acting as the pivot; Type II, rotatory fixation with anterior displacement of 3–5 mm with 1 lateral articular process acting as the pivot; Type III, rotatory fixation with an anterior displacement >5 mm; and Type IV, rotatory fixation with posterior displacement²⁾.

Rotational dislocation involving both lateral masses was first observed in patients with an intact transverse ligament. The Fielding type 1 classification is the benign type because the transverse ligament is intact. The transverse ligament inhibits an anterior shift of the atlas (C1) on the axis (C2)⁷⁾. In an intact structure, the alar ligament restricts over-rotation. Excessive rotation >56–65° will cause AARS. This rotation can usually be reduced easily and without delay⁴⁾. The right alar ligament prevents a left rotation and vice versa⁷⁾. Some authors have proposed a laxity of the atlantoaxial ligaments as the main cause of instability^{1,2,10)}; therefore, deficiencies of the alar ligament may induce AARS. Deficiencies of the transverse ligament can also occur due to this condition. Stability of the atlantooccipital and atlantoaxial segments is maintained by a complex ligamentous system. Atlantooccipital dislocation has been produced experimentally by severing the tectorial membrane and alar ligament, indicating that these 2 ligaments act as the “fulcrum” of the occipitocervical junction⁹⁾.

The more frequent the recurrence of AARS, the more severe the deformity. If signs of facet deformity are noted, or if the deformation lasts for more than 3 months, fixation surgery should be performed. Because a deformity of the superior C2 facet joint is frequently observed in patients with chronic AARS, it is a risk factor for recurrent dislocation. A lateral inclination of the atlas >20° indicates an irreducible subluxation⁴⁾. However, there have been many reports of conservative therapies. For example, Ishii et al.⁵⁾ reported a novel closed reduction method with Halo retraction for remodeling a facet deformity.

Neurological symptoms are uncommon in AARS, but occasionally sudden death occurs due to the forward movement of a rotation deformity that leads to breakage of the transverse ligament of the atlas⁵. With prolonged rotatory deformity, the heads of several patients tend to assume a forward and downward slouching position in addition to the typical Cock Robin position because of the facet deformity⁶.

AARS may be the starting gate of the facet deformity. The presence of a facet deformity is an important factor in refractory AARS. A facet deformity begins as an abnormality of the dens ligament, and leads to injury of the facet joint capsule. In a patient, the atlas changed from a horizontal to an acutely forward slant in relation to C2⁶. This drastic change occurred because of the anterior displacement of the ipsilateral C1 facet on C2, which gradually increased the bending movement and encouraged further flexion and translation of the head⁶.

Our case had AARS type 1 with an atlantooccipital dislocation. The alar ligament and tectorial membrane may have been injured, but the transverse ligament was probably intact due to the Fielding type 1 classification. Radiological studies of our case showed that the atlas changed from a horizontal to an acutely backward slant in relation to C2, which is contrary to Pang's case noted above⁶. In our case, posterior displacement of the C1 facet on C2 may have occurred in addition to a facet deformity of the posterior surface; there have been no other reports of such a case in the literature. This was not a vertical subluxation, and the atypical form is a deformity rather than a subluxation.

The key characteristic of the deformity in our case was the elevation of C1 relative to C2, which may have been caused by an inflammation injury of the anterior longitudinal ligament. Most surgeries associated with the cervical junction involve 2 columns, but some involve 3 columns and extend from the anterior to the posterior components. Dumbbell tumor surgery requires foraminal and paravertebral space surgery, and the tumor space enables this approach. As a result, it involved 3 columns. We approached the dumbbell tumor from the posterior direction, and so we manipulated the posterior component as well as both the posterior and anterior longitudinal ligaments. After surgery, nonbacterial inflammation is likely in these 3 columns, and this may be the mechanism responsible for the atypical deformity. The alar ligament might not have involved any postsurgical inflammation and infection because at the initial onset of AARS our patient's C-reactive protein level was not elevated, and

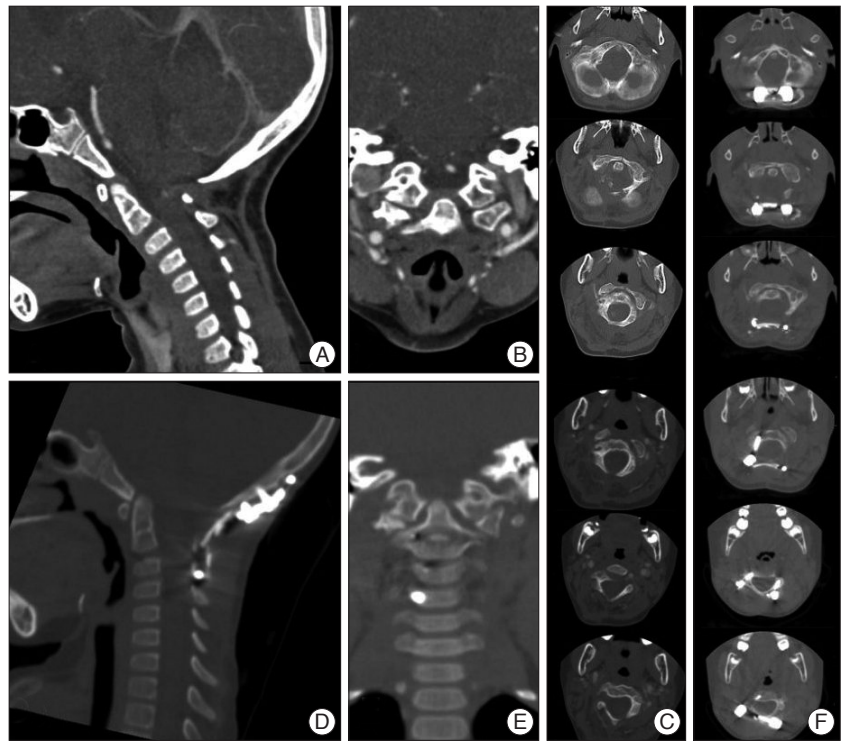


Fig. 5. A, B, and C : Pre-operative computed tomography images. D, E, and F : Postoperative computed tomography images of the cervical spine. Screws are inserted correctly and posterior fixation is achieved. Alignment is recovered.

there were no other signs of infection. This pathology was thus not related to an infection, and so we didn't use any additional antibiotics. However, a secondary non-bacterial inflammation may have extended to the facet joint capsules and the tectorial membrane and led to a facet deformity and AARS with occipitoaxial dislocation.

We had to perform an O-C1-C2 fixation for an atlantoaxial subluxation and AARS. For stabilization, we planned to use bilateral C3 pedicle screws for fixation. Theoretically, we didn't need C4 pedicle screws. However, we found that the force of the C3 pedicle screws was insufficient, and so we extended the fixation to the C4 level.

CONCLUSION

In conclusion, we report our experience of an unusual case of AARS. We hypothesize that the cause of this unusual form was the extensive approach used specifically for the removal of a dumbbell tumor. However, to our knowledge, there are no other reports of such cases to provide evidence for this hypothesis, and more cases of this unusual form of AARS are required before our findings can be confirmed.

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