

## Case Report

# Spontaneous anterior arch fracture of the atlas after occipitocervical posterior decompression and fusion in a patient with athetoid cerebral palsy: A case report

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## ABSTRACT

**Background:** Treatment of cervical myelopathy for athetoid cerebral palsy (CP) is sometimes difficult because of the strong muscle tonus and involuntary movements.

**Case presentation:** A 61-year-old male who had a history of athetoid CP presented to our hospital with severe myelopathy. Six months after the primary posterior decompression and fusion (PDF) surgery from C1 to T1, he developed a recurrence of myelopathy with severe neck pain. Radiographic findings showed bilateral C1 lateral mass screw breakage. After the revision surgery of occipitocervical PDF, his neck pain and myelopathy improved. However, he had a recurrence of neck pain with crepitation without any injury 10 days after the surgery. CT showed an anterior arch fracture of the atlas.

**Conclusions:** The possibility of spontaneous anterior arch fracture of the atlas should be considered even after occipitocervical fusion surgery when the patient has a history of CP and multiple risk factors of this fracture.

## 1. Introduction

It is widely recognized that athetoid cerebral palsy (CP) can induce cervical spondylosis through involuntary movements with strong muscle tonus, frequently causing cervical myelopathy at a relatively young age [1,2]. Conservative treatment like collar immobilization is usually not effective because of the involuntary movements and surgical treatment is often required [3]. However, the treatment of cervical myelopathy for athetoid CP is sometimes difficult because of the involuntary movements with strong muscle tonus [4]. We report a rare case of spontaneous anterior arch fracture of the atlas after occipitocervical posterior decompression and fusion in a patient with athetoid CP.

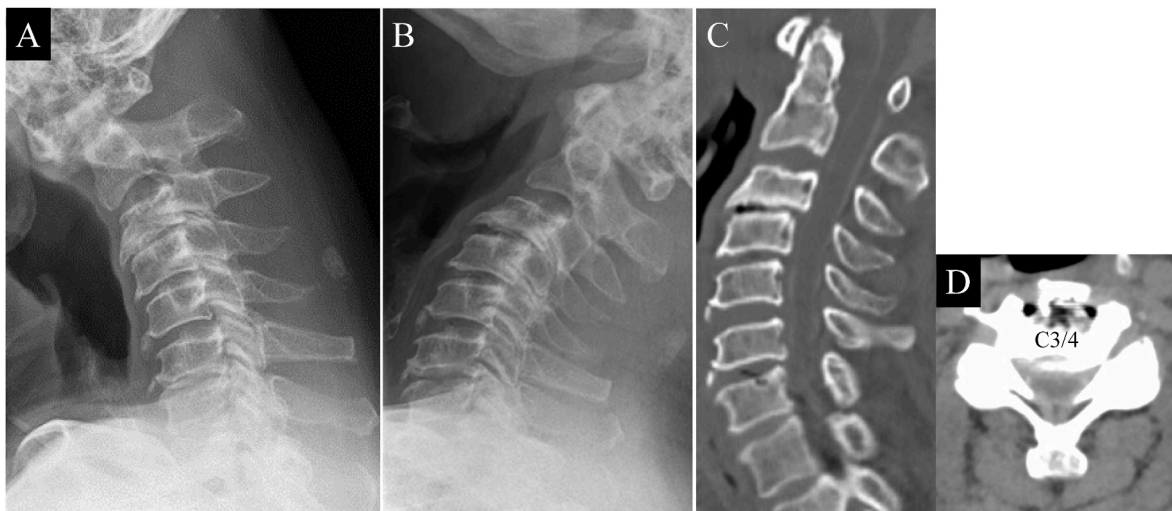
## 2. Case Presentation

A 61-year-old male presented to our hospital with progressive numbness of the upper extremities, worsening of bilateral finger clumsiness, and gait disturbance 3 months before. He had a history of athetoid CP with athetoid intensity classification grade 3 [5]. Neurological finding revealed severe myelopathy, with a Japanese Orthopaedic Association (JOA) score for cervical myelopathy of 4 points out of 17. Radiographic finding showed atlantoaxial subluxation (Fig. 1AB). Computed Tomography (CT) after myelogram showed spinal cord compression at the mainly C3/4 in addition to C1/2 and C4/5 levels with dynamic instability (Fig. 1CD). We diagnosed him with cervical myelopathy complicated with athetoid CP. Posterior decompression and fusion (PDF) with C3 to C6 laminoplasty was performed from C1 to T1 (Fig. 2A). We didn't perform C1 laminectomy because the cause of

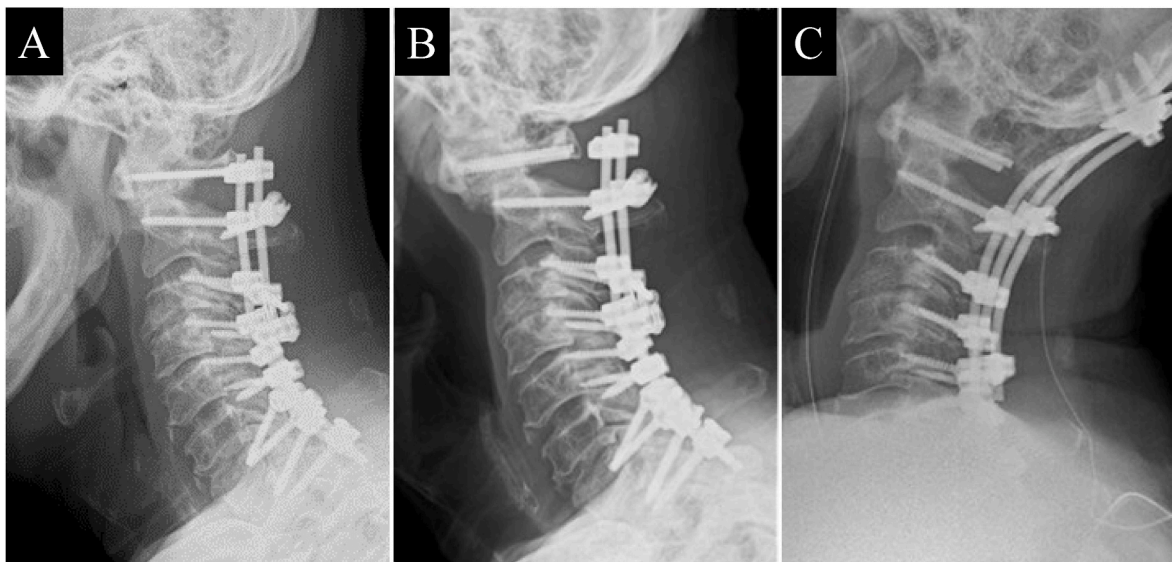
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**Fig. 1.** Imaging findings at the initial visit A. Lateral X-ray lateral image at flexion. The atlantodental interval is 5mm. B. Lateral X-ray image at extension. C. CT after myelogram sagittal reconstruction image. Spinal cord compression was observed at C1/2, C3/4, and C4/5 levels. D. CT after myelogram axial image at C3/4. Severe spinal cord compression was observed.



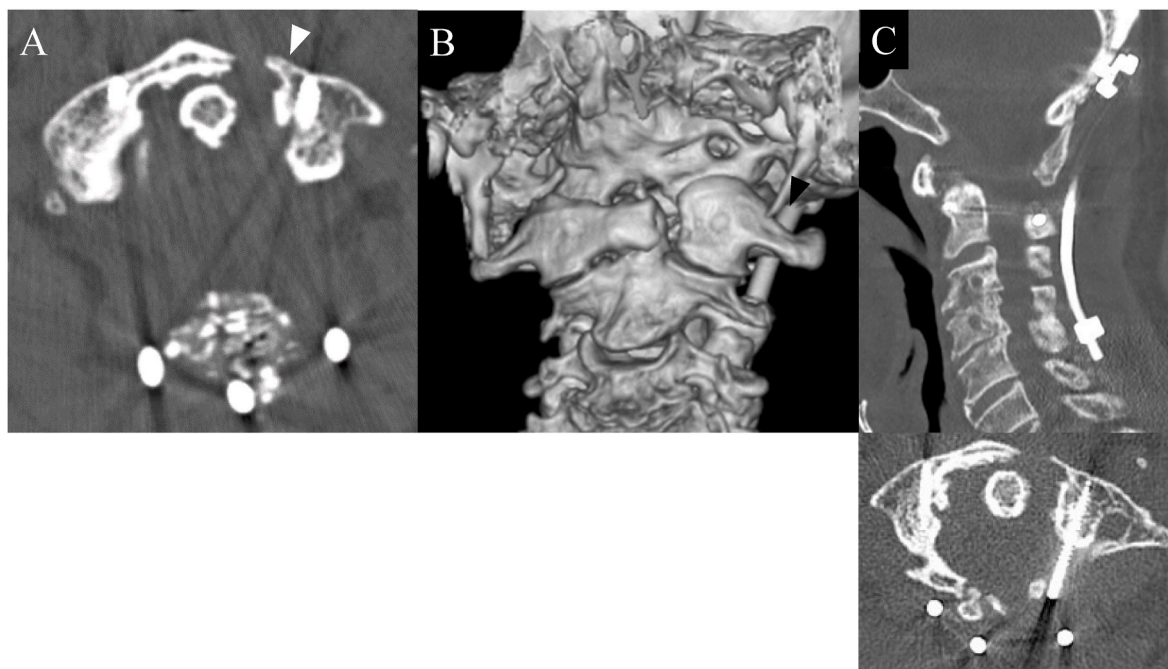
**Fig. 2.** A. Lateral X-ray image after the primary surgery. B. Lateral X-ray image 6 months after the primary surgery. Bilateral C1 lateral mass screw breakage was observed. C. Lateral X-ray image after the revision surgery.

myelopathy was mainly C3/4 and only C1-2 fusion was sufficient to improve his myelopathy. In addition, injection of botulin bilaterally to the sternomastoid muscles to reduce the muscle tonus 2 weeks after surgery. As the result, his myelopathy was significantly improved after surgery, and his JOA score improved to 10.5 points (recovery rate: 50 %). However, 6 months after the surgery, he developed a recurrence of myelopathy with severe neck pain and his JOA score declined to 8.5 points. Radiographic findings showed bilateral C1 screw breakage. He was diagnosed with recurrence of myelopathy due to atlantoaxial subluxation (Fig. 2B). After re-injection of botulin bilaterally to the sternomastoid muscles to reduce the muscle tonus, a revision surgery of occipitocervical PDF (O-T1) with C1 laminectomy was performed (Fig. 2C). His neck pain and myelopathy improved immediately after surgery. However, on the 10th postoperative day, his neck pain recurred with crepitation without any injury. CT showed an anterior arch fracture of the atlas (Fig. 3AB). He was diagnosed with a spontaneous anterior arch fracture of the atlas and insufficient control of his involuntary movements. Thus, we added the halo-vest immobilization for 3 months.

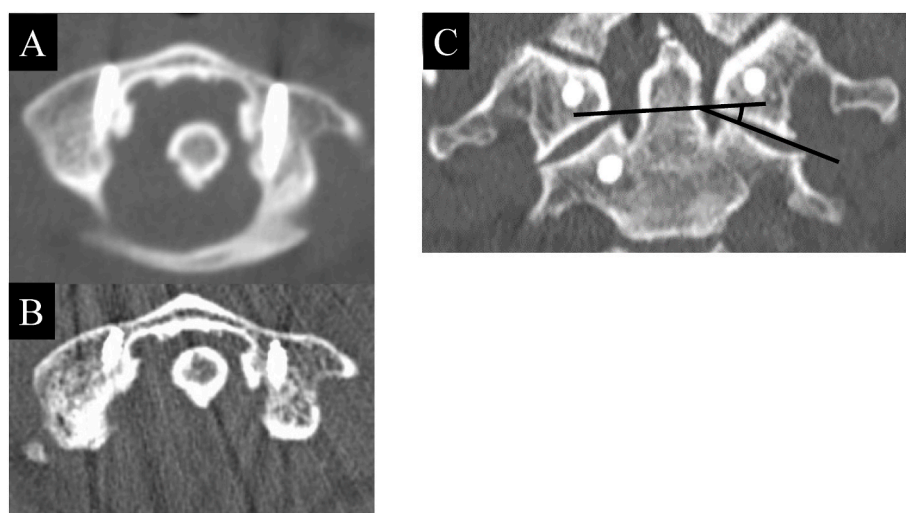
After the halo-vest immobilization, his neck pain gradually improved. The improvement of neck pain and myelopathy was maintained until the final follow-up one year after surgery when CT showed the progression of occipitocervical bony fusion from the grafted iliac bone with  $\beta$ -TCP (Fig. 3C).

### 3. Discussion

Recent reports indicate that spontaneous anterior arch fracture of the atlas is occasionally observed following C1 laminectomy without fusion, with the incidence of up to 14.2 % [6,7]. However, to our knowledge, this is the first case of spontaneous anterior arch fracture of the atlas after occipitocervical fusion surgery with C1 laminectomy. Regarding pathogenesis of the fracture, we initially consider the history of athetoid CP. Although PDF is a feasible procedure to treat cervical myelopathy associated with CP, the incidence of instrument failure after PDF surgery in patients with athetoid CP is relatively high [4]. In addition, in the present case, the fracture occurred unusually short duration after the



**Fig. 3.** A. CT axial image 10 days after the revision surgery. B. 3D CT image 10 days after the revision surgery. C. CT sagittal reconstruction image and axial image at C1 fracture site 1 year after surgery.



**Fig. 4.** A. CT axial image just before the revision surgery. B. CT axial image 1 week after the revision surgery (3 days prior to C1 anterior arch fracture). C. CT coronal reconstruction image at the C1/2 facet joint. The C1/2 inferior facet angle (IFA) was 27.0°.

surgery compared with the past report. Fortunately, we have a CT axial image just before the revision surgery and just after the revision surgery 3 days prior to the C1 anterior arch fracture (Fig. 4AB). However, even though we checked the CT axial image retrospectively, we could not detect any findings and could not predict this fracture. As the reason for the unusually short duration of this fracture, we speculate that strong muscle tonus and involuntary movements caused by athetoid CP may produce a continuous load on the atlas, considering that the C1 lateral mass screws broke. As the secondary pathogenesis of the fracture, Shimizu et al. reported a large inferior facet angle (IFA) at C1/2 greater than 23.0° and the presence of subaxial ankylosis as risk factors for anterior arch fracture of the atlas following C1 laminectomy without fusion [7]. In the present case, the IFA of the patient was 27.0° (Fig. 4C) and the subaxial vertebrae were fused during the primary PDF surgery. The existence of multiple risk factors in addition to the history of

athetoid CP may result in the spontaneous anterior arch fracture of the atlas following C1 laminectomy with occipitocervical fusion surgery.

For the treatment of anterior arch fracture of the atlas following C1 laminectomy without fusion, Shimizu et al. reported conservative treatment with collar immobilization was successful for 8 out of 10 patients, while occipitocervical fusion surgery was required for the remaining two patients [7]. In the present case, we added Halo-vest immobilization for the treatment of anterior arch fracture of the atlas to bear the vertical pressure for 3 months because the control of the patient's involuntary movements was insufficient. As a result, the patient's neck pain was relieved and occipitocervical bony fusion with grafted iliac bone progressed. The prognosis for this type of fracture would be favorable even if the fusion of the anterior arch of the atlas was not achieved, as long as occipitocervical bony fusion was achieved. In addition, as a point of reflection in the present case, the bone grafting

was insufficient at the primary surgery of PDF from C1 to T1 which may have contributed to this fracture. Appropriate bone grafting and reliable bony fusion are important for the success of atlantoaxial fusion surgery.

#### 4. Conclusion

We have encountered an extremely rare case of a spontaneous anterior arch fracture of the atlas following occipitocervical posterior decompression and fusion. The possibility of spontaneous anterior arch fracture of the atlas should be considered even after occipitocervical fusion surgery when the patient has increased involuntary movements with strong muscle tone, such as in cases of CP, and with multiple risk factors, including an increased inferior facet angle at C1/2 and subaxial ankylosis.

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#### Ethical statement

Written informed consent was obtained from the patient for the publication of this case report and accompanying images.

#### Declaration of competing interest

The authors declare that they have no conflicts of interests.

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