

Posterior Fixation for Atlantoaxial Subluxation in a Case with Complex Anomaly of Persistent First Intersegmental Artery and Assimilation in the C1 Vertebra

Daisuke UMEBAYASHI,¹ Masahito HARA,¹ Yasuhiro NAKAJIMA,¹
Yusuke NISHIMURA,¹ and Toshihiko WAKABAYASHI¹

¹Department of Neurosurgery, Nagoya University, Nagoya, Aichi

Abstract

We report a very rare case of atlantoaxial subluxation (AAS) with persistent first intersegmental artery (PFIA) and assimilation in the atlas (C1) vertebra. This case demonstrates the difficulty of deciding on a surgical strategy for complex anomalies. A 63-year-old man presented with gait disturbance, neck pain, and severe dysesthesia in his left arm. Past history included a whiplash injury. Dynamic X-ray studies demonstrated an irreducible AAS and assimilation of C1. This subluxation was slightly deteriorated in an extended position. A three-dimensional computed tomography angiography (3DCTA) indicated that the PFIA was located on the left side. We performed a C1 posterior arch resection and C1 lateral mass–axis pedicle screw (C1LM–C2PS) fixation using the modified technique of skewering the occipital condyle and C1 lateral mass. The patient had no postoperative morbidity and his symptoms disappeared immediately after operation. Complex anomalies cause difficulty in determining surgical strategy although several surgical methods for simple craniovertebral junction anomaly have been reported. To avoid significant morbidities associated with vertebral artery injury, surgical strategies for these complex conditions are discussed. The modified technique of a C1 lateral mass screw penetrating the occipital condyle is a viable treatment option.

Key words: assimilation, atlantoaxial subluxation, persistent first intersegmental artery, posterior fixation

Introduction

Surgical management of atlantoaxial subluxation (AAS) depends on the symptoms and assessment of anomalies associated with craniovertebral junction (CVJ) including os odontoideum, ossiculum terminale, hypoplastic odontoid, assimilation of atlas (C1), ponticulus posterior, persistent first intersegmental artery (PFIA), and vertebral artery (VA) fenestration. If patients have these anomalies, surgical strategy would be more complex and difficult. Ignoring these variations when planning surgical intervention can lead to injury of the neurovascular structures and complication.

We encountered an AAS case with complex anomalies of PFIA and assimilation of C1. PFIA is an anomaly in which the VA courses below the C1 arch and enters the spinal canal without passing through the transverse foramen of C1.

Among posterior fixation methods between C1 and axis (C2), C1 lateral mass–C2 pedicle screw (C1LM–C2PS) fixation is more popular and rigid. The insertion point of the C1 lateral mass screw initially described by Goel and Laheri (1994) and Harms and Melcher (2001) is located at the inferior lateral mass.^{1,2)} However, it might be dangerous to insert the screw from the point of the inferior lateral mass if the VA courses along the caudal side of the C1 arch, such as PFIA. If there is enough space between the anomalous VA and the entry point to the C1 lateral mass, the lateral mass screw can be inserted by pre-determining the entry point, as described by Hong et al.³⁾ But assimilation of C1 has made it impossible to insert the screw from the entry point, reiterating what Hong has described. We also have to consider the factors such as foramen magnum stenosis, malalignment, and stiffness of the fixation method. There are no available surgical case reports of such a rare occurrence. In this report, we describe the process of making surgical strategies and the surgical technique for complex anomaly case.

Case Report

A 63-year-old man with a previous whiplash injury about 5 years ago, presented with gait disturbance, neck pain, and severe dysesthesia in his left arm since 2 years ago, and these symptoms progressed gradually. He had motor weakness in the left wrist extensor and handgrip. His Romberg sign was positive. He was then referred to our hospital.

I. Examination

X-ray revealed AAS and assimilation of C1, and the subluxation (which had not reduced) was slightly increased in the extended position (Fig. 1). The preoperative occipita (Oc)–C2 angle was -7.5° . Preoperative three-dimensional computed tomography angiography (3DCTA) revealed the PFIA on the left side (Fig. 2) and stenosis of the foramen magnum. Angiography and balloon occlusion test for VA were not performed because 3DCTA revealed enough information and we placed a primary emphasis on avoiding VA injury.

A magnetic resonance imaging (MRI) revealed myelomalacia in the upper cervical cord as a result of the compression of the posterior arch of C1 and the odontoid process (Fig. 3). As his symptoms were thought to be induced possibly by the upper cervical cord compression, which was strongly affected not only by irreducible AAS but also by foramen magnum stenosis, we planned a posterior arthrodesis with C1 laminectomy. Because we recognized irreducible AAS and foramen magnum stenosis, we avoided any traction method for the risk of cord compression.

II. Operation

The operation was performed with the patient in the prone position with his neck neutral, and the C1 posterior arch and C2 lamina were fully exposed, preserving the semispinalis cervicis muscle through a standard midline incision. We performed subperiosteal dissection to avoid

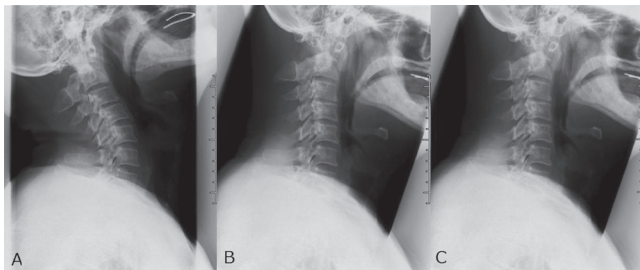


Fig. 1 An X-ray demonstrating atlantoaxial subluxation (AAS) and assimilation of C1. The subluxation was slightly increased in the extended position (A), and it was not reduced even in the neutral (B) and flexed positions (C).

bleeding from perivertebral venous plexus when both sides of C1–C2 articular spaces were exposed. However, since the condition was well developed, bleeding did occur. We managed to control bleeding, but didn't expose the inside of C1–C2 articular spaces completely due to the intermittent rebleeding. Thereafter, inferior part of occipital bone and C1 posterior arch were removed en bloc as a result of untiring drilling of these bones. It enabled us to get excellent operating fields and view so that we did not have to cut the C2 nerve root. Under direct examination of the C1 lateral mass and the C2 pedicle, C1LM–C2PS were inserted using a lateral X-ray. On the left side, where the PFIA was located, we intended to skewer these structures to avoid injuring the VA and penetrating the hypoglossal canal because of complete fusion between the occipital condyle and the C1 lateral mass, and also due to the limitations of the surgical trajectory. Fortunately, PFIA was able to be mobilized inferiorly on the left side, and we could insert the screws from the point of inferior lateral mass. On the right side, routine insertion of the C1 lateral mass screw was carried out. C2 pedicle screws were successfully settled by insertion from points higher than usual because of the relatively high location of the VA. Reduction was achieved by drawing the C1 posterior arch and applying compression force bilaterally between the C1 lateral mass screw and the C2 pedicle screws so that the articular surfaces became parallel. Local bone autografts were placed around the screw heads on the lamina surface and the space between C1 and C2 lamina which was decorticated as fusion bed.



Fig. 2 Preoperative three-dimensional computed tomography angiography (3DCTA) demonstrates the persistent first intersegmental artery (PFIA) on the left side, which courses abnormally below the arch of atlas.

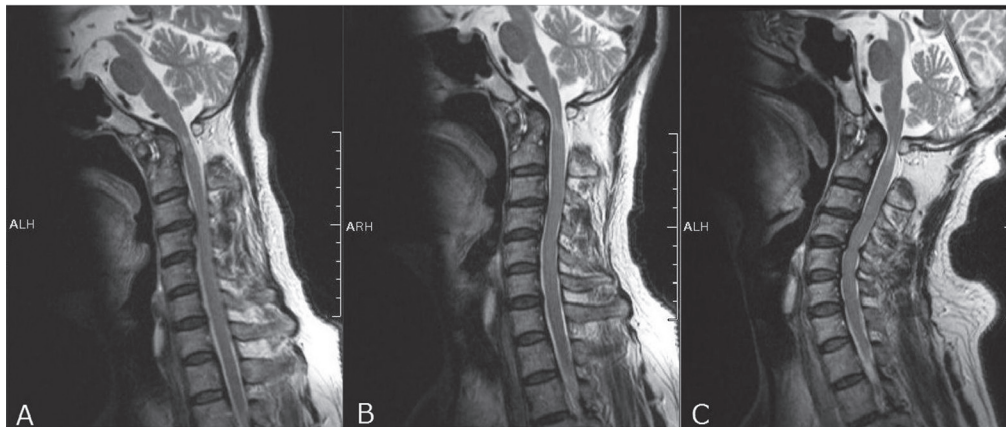


Fig. 3 A T₂-weighted sagittal magnetic resonance imaging (MRI) revealed myelomalacia in the upper cervical cord. The odontoid process compressed the ventral cervical cord in flexed (A) and neutral (B) positions, and caused cervical cord impingement in extended cord position (C). The atlantoaxial distance is slightly increased in the extended position.

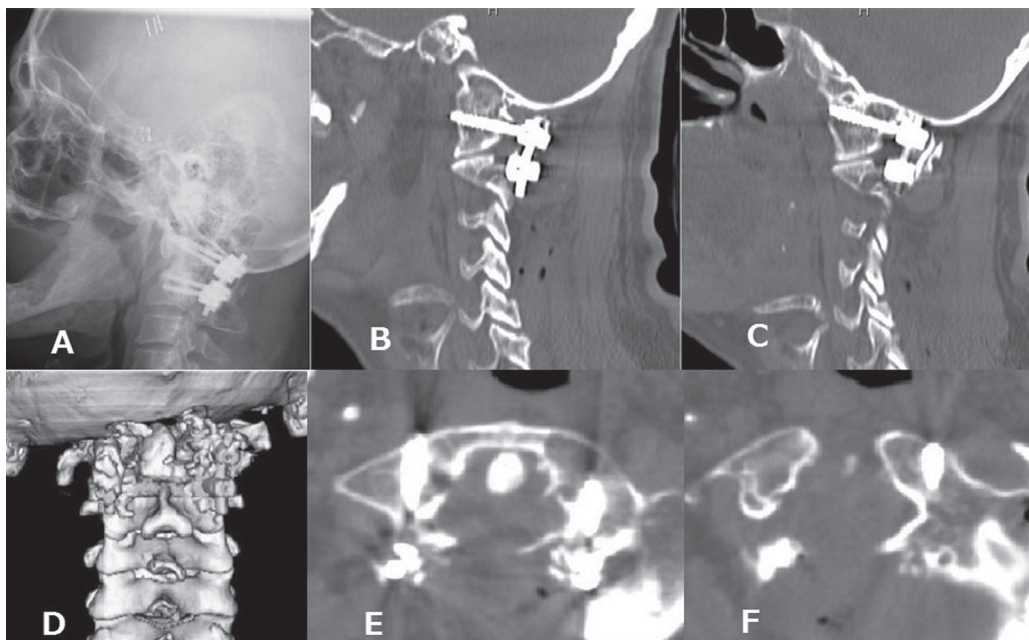


Fig. 4 An X-ray revealing a sufficient reduction between C1 and C2 (A). A three-dimensional computed tomography angiography (3DCTA) shows C1–C2 rod construct (D). Axial and sagittally reconstructed computed tomography (CT) reveals the C1 screw inserted into the lateral mass on the right side (B, E) and penetrating the occipital condyle on the left side (C, F).

III. Postoperative course

The patient's postoperative course was uneventful and his symptoms disappeared. A postoperative CT scan revealed that all the screws were properly placed (Fig. 4). The postoperative Oc–C2 angle improved to 5.5°. The alteration in the Oc–C2 angle was 13°.

Discussion

Several anatomical variations of the CVJ have been reported, including os odontoideum, ossiculum terminale, hypoplastic odontoid, assimilation of C1, ponticulus posterior, PFIA, and VA fenestration. Yamazaki et al. reported that of the 41 AAS patients without Down's syndrome, 5 had PFIA.¹⁷ They also encountered congenital osseous anomalies at the CVJ.

The incidence of PFIA at the CVJ in disease-free patients is reported to be approximately 0.60–0.67%.^{8,10}

However, Hong et al. reported an incidence of 4.7% in patients who underwent 3DCTA for a purpose other than the evaluation of VA disease.⁴ The C2 segmental type of the VA is often associated with assimilation of C1, which is described as failure of segmentation and a rearrangement of the embryonic sclerotome.^{5,10} The incidence is 75% among patients with PFIA.¹⁰ From this it can be assessed that the incidence of a complex anomaly of PFIA and assimilation of C1 at the CVJ is approximately 0.45% in disease-free patients.¹⁰ In these 0.45% patients, the number of patients who have an indication of surgical treatment for symptomatic AAS is quite small though AAS tends to complicate CVJ anomaly. We report a case of irreducible AAS accompanied by a PFIA and assimilation of C1. To the best of our knowledge, this is the first surgical case report addressing these complex anomalies.

In our case, irreducible AAS accompanied by foramen

magnum stenosis with assimilation of C1 may have induced the patient's symptoms. The upper cervical spinal cord was compressed by the ventral and dorsal bony pathology. Sagittal T₂-weighted imaging demonstrated ventral cervical cord compression in flexion and cervical cord impingement in extension. A number of studies corroborate that performing posterior decompression alone for ventral bony pathology, with or without Chiari type I malformation, may lead to neurological deterioration.¹⁵⁾ We thus decided to perform not only foramen magnum decompression by carrying out a C1 posterior arch resection, but also posterior fixation for this irreducible AAS. The necessity of a C1 laminectomy in this case should be further discussed. Assimilation of C1 is frequently associated with stenosis of the foramen magnum,¹²⁾ which was also present in this case. If C1 posterior arch resection was not performed and a horizontal reduction between C1 and C2 was not achieved, the spinal cord would be impinged. In a case of AAS, the proper surgical method should be selected depending on these conditions. If the AAS is reducible some fixation method is necessary, whereas if irreducible arthrodesis might be required in some situations. If assimilation of C1 with foramen magnum stenosis exists, as it did in our case, postoperative neurological deterioration may occur from spinal cord compression by the ventral bony pathology even if the instability is mild.

Among posterior fixation methods between C1 and C2, C1LM–C2PS fixation is more popular and rigid. The insertion point of the C1 lateral mass screw is located at the inferior lateral mass,^{1,2)} but it might be dangerous to insert the screw at this point in PFIA case because the VA courses along caudal side of the C1 arch, in other words, on the inferior lateral mass. Many spinal surgeons in that case choose alternative techniques to avoid VA injury, such as C1–C2 transarticular screw fixation, fixation between the occiput and C2, and others. If there is enough space between the anomalous VA and the entry point to the C1 lateral mass, the lateral mass screw can be inserted by predetermining the entry point, as described by Hong et al.³⁾ There is an option of inserting C2 translamina screw instead of C2 pedicle screw in the case of a narrow pedicle. Fortunately, C2 pedicles were wide enough to insert pedicle screws in this case.

We were able to perform the C1LM–C2PS fixation using a polyaxial screw–rod construct. As the screw trajectory for the patient's occipital bone and the entry point was limited, the C1 lateral mass screw was directed toward the occipital condyle on the left side, where these structures had fused. A 28-mm-long screw was used to penetrate the occipital condyle through the lateral mass.¹³⁾ It was reported that the mean stiffness of the condyle screw constructs are slightly greater than that of standard occipital plate fixation, although the difference is not statistically significant.¹⁴⁾ The occipital condyle–C2 fixation technique makes the occipital screw length longer

and the rod length shorter. The longer screws potentiate the pullout force and also provide better tolerance for higher screw loads. The shorter length of the rod with the condyle screws allows for less rod bending, which may slightly decrease the range of motion. A modified C1 lateral mass screw penetrating the occipital condyle, as in our case, may provide stronger fixation and better alignment by reliable reduction.

Further, we placed local bone autografts, but it was not into the lateral C1–C2 joints. It would be better for osseous fusion to insert autografts into the facet joint. However, we could not carry out these procedures due to the high occurrence of rebleeding in case it was carried out. It is necessary to observe carefully for the presence of pseudarthrosis in the future, and consider reconstructive surgery if needed.

The CVJ diseases include several anomalous conditions such as VA anomaly, bony pathology, and malalignment. Preoperative circumspet studies of these conditions are crucial for surgical planning. Recently, the usefulness of 3DCTA for evaluating VA anomalies at the CVJ has been described.¹⁶⁾ In addition, we must also pay great heed to subluxation between C1 and C2 in not only the horizontal direction, but also the vertical direction because vertical dislocation is often missed.

It may be important to improve the alignment of the upper cervical spine which influences the subaxial alignment.⁶⁾ In this case, the patient's preoperative cervical spine was straight, but became lordotic after the operation. We measured the alignment in this case by the Oc–C2 angle in neutral position. As the preoperative Oc–C2 angle was -7.5° , some degree of reduction of the alignment was deemed necessary because the mean angle in male subjects should approximately be 14.6° .⁹⁾ Toyama and Koyanagi warned that straight, kyphotic, and swan neck deformities occurred after insufficient reduction of C1–C2 angle, and recommended that the optimum postoperative C1–C2 angle should be 20° .¹¹⁾ Nevertheless, overreduction of the C1–C2 angle induces a loss of lordosis and kyphotic deformity in the lower cervical spine, resulting in a possibility of myelopathy with progressive subaxial subluxation.⁶⁾ The inappropriate Oc–C2 angle also has a considerable impact on dyspnea and dysphagia.⁷⁾ Sometimes, this may lead to a serious condition. Kato et al. recommended that the optimum postoperative C1–C2 angle should be 20° (preoperative C1–C2 angle) in patients with a preoperative C1–C2 angle of $< 0^\circ$.⁶⁾ In our case, Oc–C2 angle was improved to 5.5° after the operation, and the alteration in the Oc–C2 angle was 13° . These observations were found to be acceptable.

In conclusion, the possibility of VA and osseous anomalies should be considered when operating on patients with AAS. Great care must be taken not to injure the VA and to reduce the spinal alignment to some extent. Several of the methods for the fixation of posterior arthrodesis should be better mastered. As PFIA is frequently associated with

assimilation of C1, a modified C1–C2 screw fixation, as was carried out in this case, is a useful method.

Conflicts of Interest Disclosure

We have no conflicts of interest to disclose.

References

- 1) Goel A, Laheri V: Plate and screw fixation for atlanto-axial subluxation. *Acta Neurochir (Wien)* 129: 47–53, 1994
- 2) Harms J, Melcher RP: Posterior C1–C2 fusion with polyaxial screw and rod fixation. *Spine* 26: 2467–2471, 2001
- 3) Hong JT, Jang WY, Kim IS, Yang SH, Sung JH, Son BC, Lee SW: Posterior C1 stabilization using superior lateral mass as an entry point in a case with vertebral artery anomaly: technical case report. *Neurosurgery* 68: 246–249; discussion 249, 2011
- 4) Hong JT, Lee SW, Son BC, Sung JH, Yang SH, Kim IS, Park CK: Analysis of anatomical variations of bone and vascular structures around the posterior atlantal arch using three-dimensional computed tomography angiography. *J Neurosurg Spine* 8: 230–236, 2008
- 5) Hotta S, Morita A, Seichi A, Kirino T: Aberrant vertebral artery course in a case of Chiari malformation and occipitoatlantal assimilation. Case report. *J Neurosurg Spine* 3: 246–248, 2005
- 6) Kato Y, Itoh T, Kanaya K, Kubota M, Ito S: Relation between atlantoaxial (C1/2) and cervical alignment (C2–C7) angles with Magerl and Brooks techniques for atlantoaxial subluxation in rheumatoid arthritis. *J Orthop Sci* 11: 347–352, 2006
- 7) Miyata M, Neo M, Fujibayashi S, Ito H, Takemoto M, Nakamura T: O–C2 angle as a predictor of dyspnea and/or dysphagia after occipitocervical fusion. *Spine* 34: 184–188, 2009
- 8) Sato K, Watanabe T, Yoshimoto T, Kameyama M: Magnetic resonance imaging of C2 segmental type of vertebral artery. *Surg Neurol* 41: 45–51, 1994
- 9) Sherekar SK, Yadav YR, Basoor AS, Baghel A, Adam N: Clinical implications of alignment of upper and lower cervical spine. *Neurol India* 54: 264–267, 2006
- 10) Tokuda K, Miyasaka K, Abe H, Abe S, Takei H, Sugimoto S, Tsuru M: Anomalous atlantoaxial portions of vertebral and posterior inferior cerebellar arteries. *Neuroradiology* 27: 410–413, 1985
- 11) Toyama Y, Koyanagi T. [The optimum position of fusion in atlanto-axial arthrodesis.] *Seikeigeka* 45: 495–502, 1994 (Japanese)
- 12) Tubbs RS, Lancaster JR, Mortazavi MM, Shoja MM, Chern JJ, Loukas M, Cohen-Gadol AA: Morphometry of the outlet of the foramen magnum in crania with atlantooccipital fusion. *J Neurosurg Spine* 15: 55–59, 2011
- 13) Uribe JS, Ramos E, Vale F: Feasibility of occipital condyle screw placement for occipitocervical fixation: a cadaveric study and description of a novel technique. *J Spinal Disord Tech* 21: 540–546, 2008
- 14) Uribe JS, Ramos E, Youssef AS, Levine N, Turner AW, Johnson WM, Vale FL: Craniovertebral fixation with occipital condyle screws: biomechanical analysis of a novel technique. *Spine* 35: 931–938, 2010
- 15) Wang S, Wang C, Yan M, Zhou H, Jiang L: Syringomyelia with irreducible atlantoaxial dislocation, basilar invagination and Chiari I malformation. *Eur Spine J* 19: 361–366, 2010
- 16) Yamazaki M, Koda M, Aramomi MA, Hashimoto M, Masaki Y, Okawa A: Anomalous vertebral artery at the extraosseous and intraosseous regions of the craniovertebral junction: analysis by three-dimensional computed tomography angiography. *Spine* 30: 2452–2457, 2005
- 17) Yamazaki M, Okawa A, Hashimoto M, Aiba A, Someya Y, Koda M: Abnormal course of the vertebral artery at the craniovertebral junction in patients with Down syndrome visualized by three-dimensional CT angiography. *Neuroradiology* 50: 485–490, 2008

Address reprint requests to: Daisuke Umabayashi, MD, Department of Neurosurgery, Nagoya University School of Medicine, 65 Tsurumai-cho, Showa-ku, Nagoya, Aichi 466-8550, Japan. e-mail: umebayad@gmail.com