Case 1

History and examination

A 45-year-old male patient was admitted for diffuse neck pain developed from 3 years ago. There was no history of special trauma or rheumatoid arthritis. In the physical examination, abnormal findings were not detected, and in the neurologic examination, motor weakness or sensory changes were not detected. In cervical spine X-ray and computed tomography (CT), the atlanto-dental interval (ADI) was observed to be increased by 10 mm at the neutral and flexion position, the protrusion of the tip of odontoid to the McRae line just above, which was the finding suspicious to be BI, and thus various radiologic criteria diagnosing BI were measured (Fig. 1A). The odontoid tip was located below the Chamberlain line, the apex of odontoid tip was located above the McGregor line by 3.5 mm, which did not meet the criteria of BI. Nonetheless, in the Clark station, the anterior ring of the atlas was located in the station II, and it was 32 mm by the Redlund-Johnell criterion, 11 mm by the Ranawat criterion, and findings satisfying the criteria of BI were observed. In cervical spine magnetic resonance imaging, on T2 weighted images, high signal changes of the spinal cord at the C1-2 level was not shown, but behind the spinal cord, compression finding was detected. In CT angiogram, the findings of the high riding vertebral artery, abnormal course of vertebral artery, and narrow C2 pedicle were not observed. Evaluat-
ing the findings, we diagnosed this patient as idiopathic AAI associated with BI, and surgery was performed to correct the two pathologies simultaneously.

**Operation**

Under the intraoperative electrophysiologic monitoring system (IOM), intentional sacrifice of bilateral C2 ganglions were performed to obtain enough space for C1 screw entry point and bone fusion surface. Subsequently, by the application of Harms and Melcher technique, polyaxial screws were inserted to the bilateral C1 lateral masses and the C2 pedicles. The C1-2 joint was slightly distracted using a distractor, and the articular cartilages were removed widely using a curette and a high speed burr. The C1-2 joint was slightly distracted carefully under IOM to avoid overdistraction and subsequent stretching injury of the spinal cord and 8 mm iliac bone autograft were inserted bilaterally as joint spacers to maintain the vertical reduction, subsequently interlaminar fusion was performed using autograft iliac bone blocks and cables (Fig. 1C).

**Postoperative course**

After surgery, special neurological defects or occipital neuralgia caused by the resection of C2 ganglion were not seen and troublesome neck pain became tolerable immediately after surgery. In postsurgical cervical spine X-ray and CT, the Clark station, the Redlund-Johnell criterion and the Ranawat criterion were station I, 39 mm, and 15 mm, respectively, and they were restored to normal ranges (Fig. 1B, D, E).

**Case 2**

**History and examination**

A 69-year-old female patient was brought to emergency room because of sudden stuporous mentality. There was severe brain swelling with hemorrhagic infarction of the left cerebellar hemisphere on brain CT. In cervical spine X-ray, ADI was 9 mm and AAI findings were shown together with cervical kyphotic deformity. The tip of odontoid process was protruded above the McRae line by 5 mm. The odontoid tip was located above the Chamberlain line by 8 mm, and the apex of odontoid tip was located above the McGregor line by 11 mm. In the Clark station, the anterior ring of the atlas was located in the station II, it was 32 mm by the Redlund-Johnell criterion, 10 mm by the Ranawat criterion, and except the Redlund-Johnell criterion, the findings corresponding to the criteria of BI were evident (Fig. 2A, C, E).

**Operation**

Concerning the injury of cervicomedullary junction due to the neck flexion position during the emergency decompression of posterior inferior cerebellar artery infarction, correction of the high cervical deformity and decompression were performed first. To prevent hardware failures caused by C1 laminectomy for cervicomedullary decompression, kyphosis correction and poor bone quality, except the fusion extension using C3 and 4 lateral mass screws, surgery for AAI and BI was performed by the method identical to the case 1. After the surgery for the cervical spinal area, as planned, suboccipital decompression and extra-ventricular drainage were performed sequentially.

**Postoperative course**

After surgery, the mentality of patient was recovered slowly, and distinct motor weakness findings were not shown. In post-surgical cervical spine X-ray, the Clark station, the Redlund-Johnell criterion, and the Ranawat criterion were station I, 41 mm, and 17 mm, respectively, and they were restored to normal ranges (Fig. 2B, D, F).

**DISCUSSION**

In patients with anomalies including instability in the craniovertebral junction, because of the proprietary characteristic of the area such as the presence of the occipital bone, clivus, foramen magnum, upper cervical vertebrae, their associated liga-
During the past three decades, particularly in the past 10 years, there have been significant improvements in atlantoaxial fusion techniques, and various methods of atlantoaxial fixation have been described in the literature. Due to such development, in BI, posterior only surgery was attempted. Goel et al. have reported that in selected case of BI, posterior only surgery was feasible by intraoperative cervical traction followed by the distraction of C1-2 facetal joint, and subsequently, firm lateral mass fixation. Aryan et al. have reported that for cases with significant instability or vertical settling, allograft spacers were inserted to the C1-2 joint space. In addition, Jian et al. have reported that BI with AAI was treated by the direct posterior distraction technique between occipit and C2 pedicles.

After the report of a novel technique using the polyaxial screws and rods C1 lateral mass and C2 pedicle described by Harms and Melcher, the direct manipulation of C1 and C2 using the instrumentation as the reduction device became feasible. Similar to the 2 cases reported by us, in BI associated with AAI, although the ventral spinal cord compression was not severe, after placing the instrumentation, the C1-2 facetal joint was released, and autograft iliac bone spacers were inserted by distracting C1 and C2 screw. Cervical traction was not necessary preoperatively or even during the operation, because intraoperative C1-2 reduction was possible using C1-2 segmental screw construct. The procedure has several advantages; 1) vertical reduction of BI can eliminate the necessity of additional posterior decompressive procedure such as C1 laminectomy and posterior fossa decompression, 2) increase of the fusion bed using the facet graft could enhance fusion rate, 3) reduction of the risk of trans-oral surgery, 4) minimization of cervical traction prior to surgery or during surgery, and 5) preservation of occipito-atlantal motion avoiding occipito-cervical fusion.

C2 root resection is necessary to expose enough space of C1-2 facet joint and perform the entire operation under direct vision. Ames et al. have reported that there was no cerebrospinal fluid leakage and only one case of neuropathic pain postoperatively after sacrificing the C2 roots in 102 patients, however, routine sacrifice of C2 roots is controversial and long term follow up is mandatory to analyze the clinical effect of C2 nerve root resection.

Direct distraction of C1-2 facet joint can be used to reduce basilar invagination and to expose room for bone graft insertion within the joint cavity, which could provide remarkable stability to the region by itself. Exposed C1-2 facet joint surface provides a wide space for bone fusion. However, in cases of more severe ventral compression combined with irreducible BI, this procedure must be warranted by further investigation and transoral decompression might be necessary before or after posterior surgery.

Considering the spinal cord damage caused by overdistraction, the IOM should be observed carefully during the distraction performed after the facet release, and it is thought that the size of graft should be determined by considering the level of height restoration in advance. In addition, to prevent injury of spinal cord and medulla oblongata caused by the posterior shifting of the odontoid process, distraction should be performed while fixing C2 screws with a screwdriver.

**CONCLUSION**

We report experiences of AAI cases without severe ventral compression and associated with BI. The stabilization of AAI and the vertical reduction of BI were performed simultaneously by performing distraction of C1-2 facet joint with C1-2 screws, and inserting autograft iliac bone as facet joint spacers. These
cases show the importance to determine the presence or absence of vertical instability and reducibility in cases with BI. Stabilization and fusion in a reduced and anatomical position can reduce the level of fixation, result in sound bone fusion using interfacetal bone graft and maintain the occipitoatlantal joint motion.

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