www.jkns.or.kr http://dx.doi.org/10.3340/jkns.2012.51.3.177 Print ISSN 2005-3711 On-line ISSN 1598-7876

J Korean Neurosurg Soc 51 : 177-181, 2012

Technical Note

Intraoperative Vertebral Artery Angiography to Guide C1-2 Transarticular Screw Fixation in a Patient with Athetoid Cerebral Palsy

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We present a case of an athetoid cerebral palsy with quadriparesis caused by kyphotic deformity of the cervical spine, severe spinal stenosis at the cervicomedullary junction, and atlantoaxial instability. The patient improved after the first surgery, which included a C1 total laminectomy and C-arm guided righ side unilateral C1-2 transarticular screw fixation. C1-2 fixation was not performed on the other side because of an aberrant and dominant vertebral artery (VA). Eight months after the first operation, the patient required revision surgery for persistent neck pain and screw malposition. We used intraoperative VA angiography with simultaneous fluoroscopy for precise image guidance during bilateral C1-2 transarticular screw fixation. Intraoperative VA angiography allowed the accurate insertion of screws, and can therefore be used to avoid VA injury during C1-2 transarticular screw fixation in comorbid patients with atlantoaxial deformities.

Key Words : Atlantoaxial instability · Transarticular screw fixation · Vertebral artery · Intraoperative angiography · Athetoid cerebral palsy.

INTRODUCTION

C1-2 transarticular screw fixation is widely used to correct atlantoaxial instability because of its greater biomechanical stability and superior fixation of atlantoaxial rotation than wiring methods or Halifax clamps[4,7,8,17,22]. However, Madawi et al.[11] stated that about 20% of patients have an atlantoaxial anatomy that precludes safe screw insertion, such as an aberrant vertebral artery (VA), a small C2 isthmus, bone destruction, severe kyphosis, or obesity. Of these, an aberrant VA remains an absolute contraindication. Furthermore, VA injury during screw insertion remains a serious complication of C1-2 transarticular screw fixation. In previous studies, the risk of VA injury was 4.1-5% per patient or 2.2-2.7% per screw inserted[20].

Recent studies reported that novel, safer trajectories for atlantoaxial screw fixation to overcome a high-riding VA, narrow isthmus axis, or comorbid conditions could be determined based on image-guidance systems or aiming devices[9,13]. Several studies reported that use of computer-assisted systems allowed the safe and accurate insertion of pedicle screws and transarticular screws[8,15]. We describe the use of intraoperative VA angiography for safe and accurate C1-2 transarticular screw fixation in a patient with an aberrant VA due to severe kyphosis and instability.

MATERIALS AND METHODS

Clinical presentation

A 54-year-old female with athetoid movement and torticollis due to cerebral palsy presented with neck pain and was unable to stand unassisted after falling down a set of stairs. On neurologic examination, there was quadriparesis, and all extremities were grade 2 according to the Medical Research Council (MRC) muscle strength scale. The examination also revealed typical signs of myelopathy such as limb hyperreflexia and sensory loss. Initial T2-weighted MRI scans demonstrated a prominent kyphotic angulation deformity of the cervical spine and marked severe spinal stenosis at the cervicomedullary junction with signal change of the spinal cord. Cervical spine radiographs re-

• Received : April 14, 2011 • Revised : December 6, 2011 • Accepted : March 8, 2012
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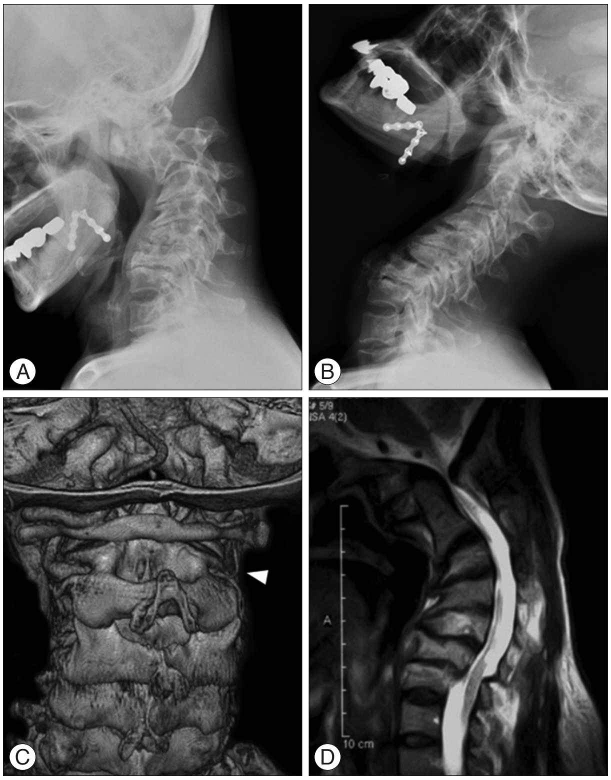

Fig. 1. Preoperative images obtained in a static 54-year-old female with torticollis. Lateral flexion (A) and extension (B) cervical radiographs showing atlantoaxial instability and severe kyphotic deformity. Initial 3D CT angiography shows hypoplastic right vertebral artery (C). Sagittal plane MRI (D) demonstrating marked spinal cord compression at cervicomedullary junction.

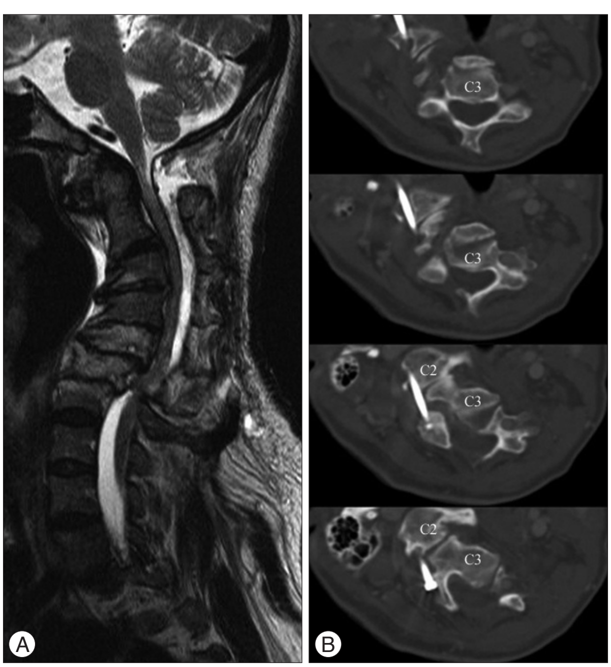

Fig. 2. Images obtained after 8 months of the initial operation. Follow-up sagittal plane MRI (A) showing enough decompression of stenosis at cervicomedullary junction. However, serial CT scans (B) reveal malposition of screw and encroachment of VA. VA : vertebral artery.

vealed that the atlantodental interval was 6 mm in flexion and 11 mm in extension, indicating atlantoaxial instability. 3D CT angiography showed hypoplastic right vertebral artery and MRI demonstrated marked spinal cord compression at cervicomedullary junction (Fig. 1).

During the initial operation, we performed a bilateral total C1 laminectomy and a right unilateral C1-2 transarticular screw fixation. The cervical vertebrae were so seriously distorted that only the unilateral side of the non-dominant VA was considered feasible to correct. Postoperative T2-weighted MRI demonstrated a fully decompressed spinal cord. The patient showed remarkable recovery after surgery; she began to walk with assistance and was free from neck pain. Her muscle power improved to grade 4 on the MRC scale and there were no other perioperative complications. Eight months after the initial operation, however, she reported severe neck pain again and the quadriparesis had worsened to grade 2 on the MRC scale. The follow-up MRI and CT scans revealed sufficient decompression of stenosis, however, the initial screw did not pass articular facet of atlantoaxial joint thoroughly but contacted at the edge of the facet (Fig. 2). The instability was aggravated due to the malposition of the screw, therefore, the patient required revision surgery to correct the neurologic deficit and C1-2 instability. We decided to perform bilateral screw fixation to overcome the intractable instability of the atlantoaxial joints, despite the risk of VA injury. We decided to use intraoperative VA angiography with simultaneous fluoroscopy for precise image guidance during C1-2 transarticular screw fixation in this patient with a distorted VA anatomy.

Operating technique

Under general anesthesia with fiberoptic intubation, the patient was carefully positioned prone on the operating table. Her skull was held in a translucent Mayfield three-point pin fixation device. Under direct lateral fluoroscopic vision, the patient's neck was flexed and the skull was translated posteriorly into a 'military tuck' position. To perform VA angiography, the radiologist stood on the right side of the operating table. In this configuration, the left popliteal region could be positioned to ensure that the proximal end of the introducer sheath was easily accessible. Access into the popliteal artery was obtained using an 18-gauge needle. Subsequently, a 0.035-inch steerable wire was inserted through the superficial femoral artery. A 7-French sheath was then positioned over the wire within the popliteal artery. The sheath was sutured in place at the skin entry site and the side port was connected to a system that delivered a pressurized infusion of heparinized saline. A 6-French guiding catheter was introduced far into the left cervical VA.

A midline posterior cervical incision was made and subperiosteal dissection was done from the occiput to C3. The cortical surface of the posterior arch of the atlas was clearly delineated with curettes and the pars of the axis was also exposed for screw insertion. The K-wire was positioned at the C2-3 facet joint through the percutaneous tunnel along the desired trajectory. The entry point was confirmed by an imaginary trajectory line seen on fluoroscopy based on a standard entry point. Usually, the articular surface of the C1-2 complex is flat and in a craniocaudal orientation with respect to the sagittal plane. In this case, the C1 inferior articular facets and the C2 superior articular facets had an anteroposterior orientation. Because the imaginary line from the C2-3 facet joint to the C1 anterior tubercle inclined toward the ventral side, we had to revise the trajectory of the K-wire to be more ventral than usual. Moreover, the atlas was deeply positioned in the working field of the operation because of severe kyphotic angulation of the atlantoaxial joint. There was not sufficient space to expose the posterior arch or lateral mass of the atlas.

Using a high-speed drill, we pierced the cortical bone of the C2 inferior facet to mark the K-wire entry point. During the operation, VA angiography was performed whenever we needed to view real-time images. The course of VA was visualized on lateral fluoroscopy when contrast was temporarily injected. The drill had a trajectory that was 10 degrees medial toward the anterior tubercle of the atlas. We advanced the drill above the loop of the VA seen on angiography (Fig. 3). After we confirmed the patency of the VA repeatedly, a 42-mm-length cortical screw was inserted. Immediate VA angiography showed no violation of the VA during cortical screw insertion. We performed contralateral screw fixation as described above.

Postoperative course

Postoperatively, the patient's weakness was improved to the extent that she was able to walk with assistance again. Her neck pain disappeared 6 months after the last surgery. Both screws inserted during the second operation using intraoperative VA angiography were positioned safely. The screw trajectories were assessed with postoperative CT and plain radiographs. There were no screw encroachments on the VA or spinal canal. The second procedure achieved bicortical purchase and provided satisfactory stability. Four years after the second operation, CT scans showed completely fused atlantoaxial joints (Fig. 4).

DISCUSSION

Surgery is usually performed to treat patients with cervical myelopathy complicated by athetoid cerebral palsy. We treated our patient by laminectomy and unilateral atlantoaxial screw fixation. The patient's symptoms initially improved after the operation, but became worse again when C1-2 instability developed. The malpositioned screw was finally displaced because of repetitive involuntary neck movement. It is considered very difficult for us to manipulate deeply positioned suboccipital area of the patient, and she also did not want to be immobilize in her neck motion even though she had repetitive neck tremor. Therefore, we intended to limit fusion to preserve a residual mobility of the head and cervical spine.

Some authors performed circumferential fusion or limited their fusion to either anterior or posterior elements[3,6,14]. Hirose and Kadoya[6] performed interbody fusion for patients with cervical spondylotic myelopathy secondary to athetoid involuntary movements. Although, the patients were preoperatively bedridden, all have been able to walk after surgery. The authors also revealed an excellent fusion up to four and a half years of follow-up. In contrast, others reported about a relapsing deterioration in long-term period, because spinal fusion increases strains on adjacent levels[1,5]. Azuma et al.[1] experienced late neurologic deterioration after combined anterior-posterior fusion for cervical spondylotic myelopathy in patients with cerebral palsy. Even though surgical results at the most improved period were

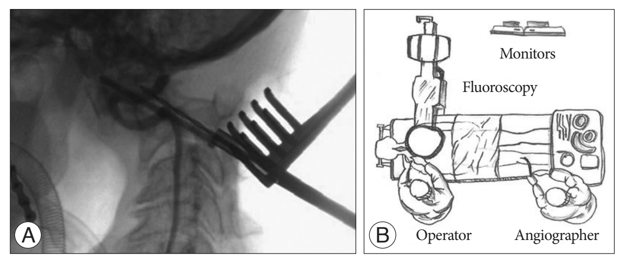

Fig. 3. Intraoperative lateral fluoroscopic image obtained during vertebral angiography and insertion of the drill (A). Illustration for layout of the operation room (B) suitable for transpopliteal intraoperative VA angiography with fluoroscopy during atlantoaxial screw fixation. VA : vertebral artery.

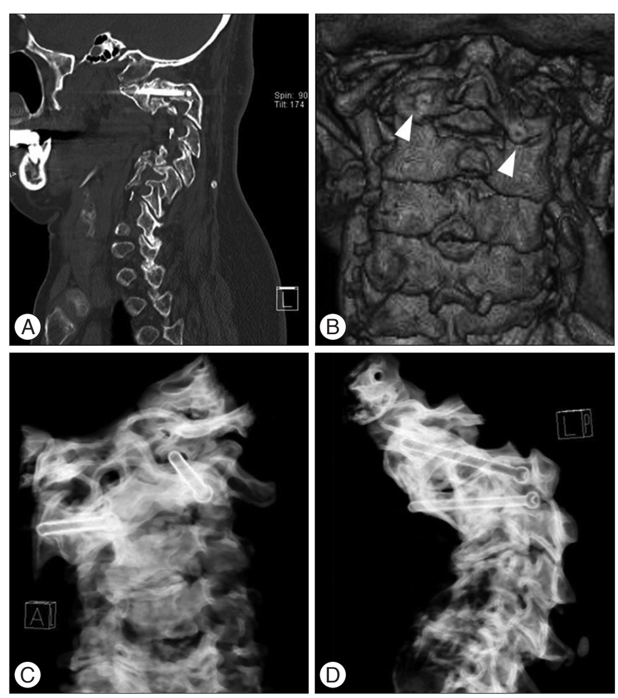

Fig. 4. Last follow-up three-dimensional CT scans obtained after 4 years of the second operation showing completely fused atlantoaxial joint (A). 3D CT scans (B) demonstrate head of atlantoaxial screws at inferior facet of C2 (white arrow) and right vertebral artery is not visible due to the violation. Individual screws on transparent 3D reconstruction (C and D) show extraordinary trajectory caused by spinal deformity but both of them thoroughly pass atlantoaxial joints.

good or excellent in all 10 patients. Six of them deteriorated from 8 to 13 years after surgery, largely because of progressive deformity. It seems that loss of cervical spine motion by fusion increased the shearing force by athetoid movements further. In 8.7-year-follow-up study of Haro et al.[5], 22 patients with cervical myelopathy caused by athetoid cerebral palsy underwent posterior fusion with anterior interbody fusion with internal fixation. The mean motion between C1 and C2 increased and atlantoaxial subluxation occurred postoperatively in five patients.

Because injury to the dominant VA can result in catastrophic complications. We only inserted the screw on one side with a hypoplastic VA to avoid dominant VA injury on the other side. Neither VA injury nor spinal cord injury occurred when this surgery was performed. We felt that fluoroscopy was not sufficient for guidance during C1-2 transarticular screw fixation to avoid VA injury or to ensure accurate insertion of the screw. We decided to use intraoperative VA angiography with simultaneous fluoroscopy for precise image guidance during C1-2 transarticular screw fixation. After this surgery, good bony fusion was obtained with satisfactory results. In patients with atlantoaxial instability complicating athetoid cerebral palsy, atlantoaxial screw fixation may be contraindicated due to complicated anatomy. However, intraoperative VA angiography for image guidance during screw insertion allows safe screw insertion, even in patients with atlantoaxial instability.

VA anomalies in comorbid patients

Yamazaki et al.[21] reported atlantoaxial instability in Down syndrome patients with craniovertebral junction (CVJ) anomalies. They later reported the presence of coexisting VA anomalies, such as intersegmentation, duplication, fenestration, and a tortuous course. They emphasized preoperative three-dimensional fine-cut CT or CT angiography to obtain detailed information about the VA morphology. Tokuda et al.[18] performed VA angiograms in 300 patients free from disease at the CVJ and found only three cases of VA anomalies. However, in CVJ disease cases, for example, Klippel-Feil syndrome, hypoplasia of the atlas, basilar invagination, or occipitalization of the atlas, the incidence of VA anomalies is increased[18].

In our study, the patient had structural abnormalities because of athetoid cerebral palsy. The structural abnormalities were caused by repetitive neck movements and seemed to accelerate the progression of cervical instability[3,19]. A larger pendulous involuntary motion of the neck and dystonic posture cause excessive flexion, extension and rotation, and increase cervical degeneration. We felt that conservative management options, such as medication, cervical orthosis, and traction, would not be effective in this patient because she could not maintain cervical immobilization[14]. We reasoned that surgical decompression and fusion would be effective treatments to avoid the progression of myelopathy in this patient.

In addition to extensive preoperative imaging studies, such as 3-dimensional CT with angiography and MRI, we performed intraoperative lateral fluoroscopy and VA angiography. The orientation of the articular facets of the C1-2 complex guides the trajectory of screw insertion. In our patient, about 1/3 of the atlas had slipped anteriorly and the articular facets of the C1-2 complex were disoriented. In contrast to normal individuals, the C1 inferior articular facet of our patient faced posteroinferiorly, and her C2 superior articular facet faced anterosuperiorly. Thus, the trajectory from the caudal edge of the C2 inferior facet to the C1 anterior tubercle had a more coronal inclination in the sagittal plane. The slipped atlas was embedded in a narrow space between the occiput and axis. Insertion of a C1 lateral mass screw therefore seemed impossible. The VA leaving the C2 transverse foramen did not course straight upward to the transverse foramen of C1, but rather, curved anteriorly along the line of kyphotic angulation. The loop formed before the VA course to the cranium superomedially was on the most critical point because it is the highest point the screw should pass above the VA at the level of atlantoaxial joint. The VA angiography was essential to avoid damage to the anomalous VA in our patient.

Avoidance of intraoperative complications

Transarticular screws allow stronger fixation of the atlantoaxial joint than other conventional posterior wiring methods. However, this technique may result in damage to the neural element or the VA. In particular, the course of the VA in the C2 vertebra is variable and complex. The VA also makes an acute lateral band just below the superior articular facet of the C2 vertebra[2]. If the height or width of the C2 vertebral isthmus is very narrow in patients with an aberrant VA or a large VA groove, measurement of the screw trajectory is more complicated.

Kawaguchi et al.[8] described the use of frameless stereotaxis to aid transarticular screw fixation. Frameless stereotaxis provides real-time anatomic information during surgery that can be used to gauge whether the screw can be inserted safely and what screw length is appropriate[9,15]. However, frameless stereotaxis has some disadvantages. Frameless stereotaxis systems are expensive and the procedure is time-consuming. In contrast to cranial procedures, it is harder to match bony landmarks with prepared images in spinal surgery. For example, a reference frame must be applied to the spinous process. Because the atlantoaxial joint is deeper and narrower than any other spinal segments, use of a frameless stereotaxis system requires resolution of more technical problems. Neo et al.[13] designed an aiming device for transarticular screw fixation. The tip of the device is placed in the pit on the isthmus and is fixed in a stable position. These authors obtained more successful screw trajectories in 10 patients. Their device is much less expensive and their procedure less time-consuming than frameless stereotaxis, and is applicable to all patients with a normal anatomy. However, 4 of 20 screws were not accurately placed using this technique. Moreover, it cannot be used to adjust patients with an anomalous VA and/or craniovertebral junction.

Unless a frameless stereotaxis system or an aiming device is available, intraoperative angiography with fluoroscopy is useful for determining the technical success of transarticular screw fixation. Schievink et al.[16] demonstrated that intraoperative spinal angiography is technically feasible, can be performed safely, and has adequate resolution for the surgical management of spinal arteriovenous malformations. They found intraoperative angiography useful not only to confirm whether important vessels were obliterated or not, but also to guide the operation. However, spinal angiography performed with a patient in a prone position may be limited by obstacles such as an inaccessible femoral artery. Leakage and bleeding from the femoral artery may be missed in cases because the catheter puncture site is invisible[10]. Furthermore, the femoral artery may be occluded by the patient's weight loaded on the sheath[7]. We recommend to use a transpopliteal route when a patient is in a prone position, as this allows observation of the catheter-puncture site during the operation and access to the popliteal artery is easy when the patient is in the prone position. On the other hand, hemostasis at the operative field might be annoying with heparinization during intraoperative angiography. We could maintain small amount of heparin by the continuous perfusion of high-concentration heparinized saline, following previously reported method of Mihara et al.[12] The author have suggested a continuous infusion of heparin at 3 mL/h, of a solution of 1 liter of saline containing 8000 units of heparin in an attempt to reduce the total dose of heparin to the patient during intraoperative angiography.

CONCLUSION

When planning surgical treatment of a patient with athetoid cerebral palsy, VA anomalies and cervical spinal deformity should be taken into consideration. Despite the risks of transarticular screw fixation in such patients, it is nevertheless a very effective method of achieving atlantoaxial stability. Intraoperative VA angiography may allow real-time assessment of the vascular anatomy and immediate correction of technical faults during the procedure. Therefore, it enables the accurate insertion of screws and avoidance of VA injury during atlantoaxial transarticular screw fixation in patients with an anomalous VA.

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