

Os Odontoideum With Cerebellar Infarction

A Case Report

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Study Design. A case report.

Objectives. To report the case of a child with os odontoideum associated with cerebellar infarction and to discuss the correlation between atlantoaxial instability with os odontoideum and vertebrobasilar artery insufficiency.

Summary of Background Data. Knowledge of the influence of atlantoaxial instability on vertebrobasilar artery insufficiency remains limited despite the publication of several reports.

Methods. A 5-year-old boy with ataxic gait disturbance was hospitalized in the pediatric ward. Magnetic resonance imaging revealed multiple cerebellar infarctions, and cerebral angiogram showed occlusions of several branches of the basilar artery and a winding of the left vertebral artery. Stress lateral radiographs of the cervical spine showed atlantoaxial instability with os odontoideum. Posterior C1–C2 transarticular screw fixation with iliac bone graft was applied to obtain firm stability and fusion.

Results. There was no damage to the vertebral arteries or spinal nerves in the perioperative period. Solid union of the grafted bone and rigid stability of the atlantoaxial joint were seen on lateral flexion–extension radiographs 1 year after the operation. There has been no sign of recurrent arterial insufficiency, and the patient has been free from cerebellar dysfunction to date.

Conclusions. Atlantoaxial instability may cause insufficiency of the vertebral artery as well as spinal cord injury. More attention should be paid to the possible relation between atlantoaxial instability and vertebrobasilar artery insufficiency. [Key words: os odontoideum, cerebellar infarction, child] *Spine* 2000;25:1178–1181

Os odontoideum with atlantoaxial instability may cause cervical cord injury. There have been several reports about vertebral artery insufficiency due to this instability. Here a rare case of os odontoideum with cerebellar infarction is described.

■ Case Report

A 5-year-old boy with mental retardation developed an ataxic gait disturbance. He touched the back of his head repeatedly and seemed to experience head pain. He also vomited several times. A history of head or neck trauma was unclear. He was admitted to the pediatric department with a diagnosis of acute encephalitis or cerebellar

ataxia at admission. Magnetic resonance imaging (MRI), however, revealed that there were areas of multiple infarctions in the left cerebellum, the left thalamus, and part of the right cerebellum. Echocardiogram did not show any embolus. Cerebral angiogram showed occlusions of several branches of the basilar artery and a winding of the left vertebral artery at C1–C2 (Figure 1). After receiving antithrombotic therapy for the multiple cerebellar infarctions, he gradually recovered the ability to walk.

Forty days after the onset, he visited the clinic for further examination of the cervical spine, which was suspected to be a possible cause of the child cerebellar infarction. Routine radiographs of the neck showed an atlantoaxial subluxation with os odontoideum. Lateral films taken with active flexion and extension showed 10 mm of translation between the two positions (Figure 2). The space available for cord (SAC) was 11 mm at both flexion and extension. Magnetic resonance imaging in neutral position showed neither direct cord compression at C1–C2 nor narrowing of the spinal canal (Figure 3). The patient did not have trisomy 21.

Internal fixation and fusion was selected to secure the reduced position between C1 and C2 and to omit excessive postoperative immobilization. Posterior C1–C2 transarticular screw fixation was performed under lateral fluoroscopic guidance with titanium cannulated cancellous screws with a diameter of 2.7 mm (Figure 4). Autologous iliac bone was placed between the two laminae and tied with a braided polyester suture. There was no damage to the vertebral arteries or to the spinal nerves during the operation. A simple soft collar was used for 3 months. Although there was a subcutaneous infection a few weeks after the operation, rigid stability and solid union of the grafted bone were seen on lateral flexion–extension radiographs 1 year after the operation. There has been no sign of recurrent arterial insufficiency, and the patient has been free from cerebellar dysfunction to date.

■ Discussion

Although the etiology of os odontoideum is controversial, previous trauma to the neck has been considered one of the most probable causes.³ Os odontoideum presenting as atlantoaxial instability may cause symptomatic cord compression; thus, posterior fusion for stabilization is recommended in some cases. Indications for surgical fusion include persistent neck pain, chronic or acute myelopathy, vertebrobasilar artery insufficiency, and evi-

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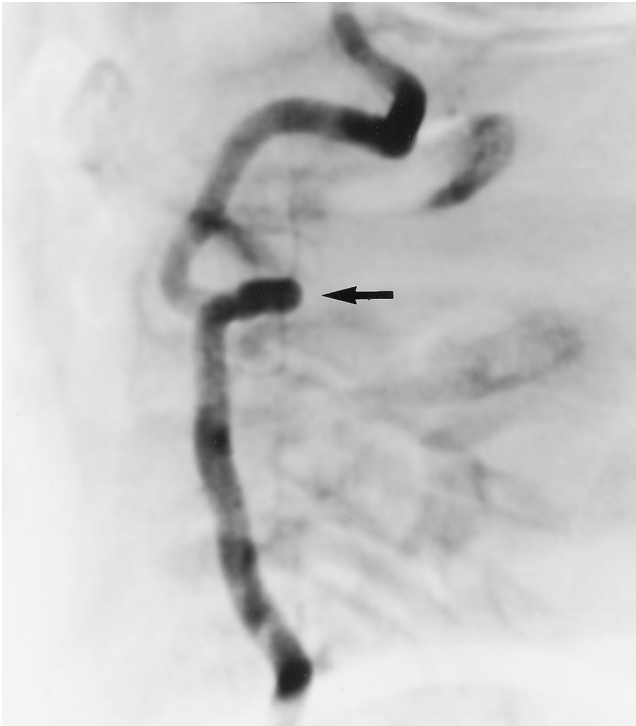


Figure 1. Lateral view of left vertebral angiogram, subtraction radiograph film, demonstrating winding of the left vertebral artery (arrow).

dence of progressive subluxation.⁵ Roentgenographically, the presence of translation instability of 10 mm or more between the atlas and the axis is considered an indication for fusion if the spinal cord symptoms are progressive.² Spierings et al¹⁰ measured the distance between the posterior border of the body of the axis and the posterior arch of the atlas on flexion of the neck (D_{\min}). They suggested that the patients whose D_{\min} is less than 13 mm are at risk of developing permanent or progressive cord signs. Although they encountered four patients with back lifting symptoms, they made no mention of arterial insufficiency. In children who had a reducible subluxation and adequate posterior elements, Smith et al⁹ recommended internal fixation with sublaminar wiring to decrease the need for more restrictive or prolonged postoperative immobilization; however, 6 of 13 patients in that study exhibited overreduction (posterior translation defined as greater than 2 mm) that could damage the cord by a “pincer” mechanism between the os fragment and the upper arch of the axis.

Pediatric ischemic stroke in the vertebrobasilar region is rare.⁷ Miyata et al⁷ have reviewed 49 reported cases of childhood vertebrobasilar artery occlusion. They summarized the causes of vertebrobasilar occlusion to be blunt head trauma, infection such as sepsis and bacterial



Figure 2. Lateral films of cervical spine with active flexion (A) and extension (B) clearly show atlantoaxial instability.



Figure 3. Sagittal view of T1-weighted MRI in neutral position shows no direct compression at C1–C2 nor narrowing of the spinal canal. A part of cerebellar infarctions can be seen.

endocarditis, congenital vascular anomalies, and os odontoideum. Other suspected causes they cited were congenital heart disease, arteritis, migraine, fibromuscular dysplasia, Duchenne muscular dystrophy, chiropractic manipulation, spontaneous neck rotation, upper extremity injury, and surgery.

Five cases of os odontoideum associated with pediatric cerebellar infarction have been reported.^{1,4,6–8} Cerebral angiogram showed narrowing or occlusion of the vertebral artery or the basilar artery in all cases. Singer et al⁸ proposed that repeated subluxation of the atlas on axis may produce kinking, compression, stretching, and tearing of the intima of the vertebral arteries leading to thromboembolic vertebrobasilar occlusions. Bhatnagar et al¹ speculated that many symptoms such as headache, hemiplegia, seizures, gait abnormality, apnea, vertigo, and loss of consciousness may be caused by localized cord compression or vertebral artery ischemia. In five cases, the operative procedure included posterior C1–C2 wiring with iliac bone graft in three and occiput–C3 fusion in one (bone graft was not mentioned).^{1,4,6,8} One patient underwent urgent suboccipital craniectomy to deal with a swollen cerebellum; further surgery was declined by the parents because of good neurologic recovery.⁷

In the case presented, MRI showed multiple cerebellar infarctions, and cerebral angiogram showed occlusion of the basilar artery and winding of the left vertebral artery at the level of the axis. Although the patient recovered

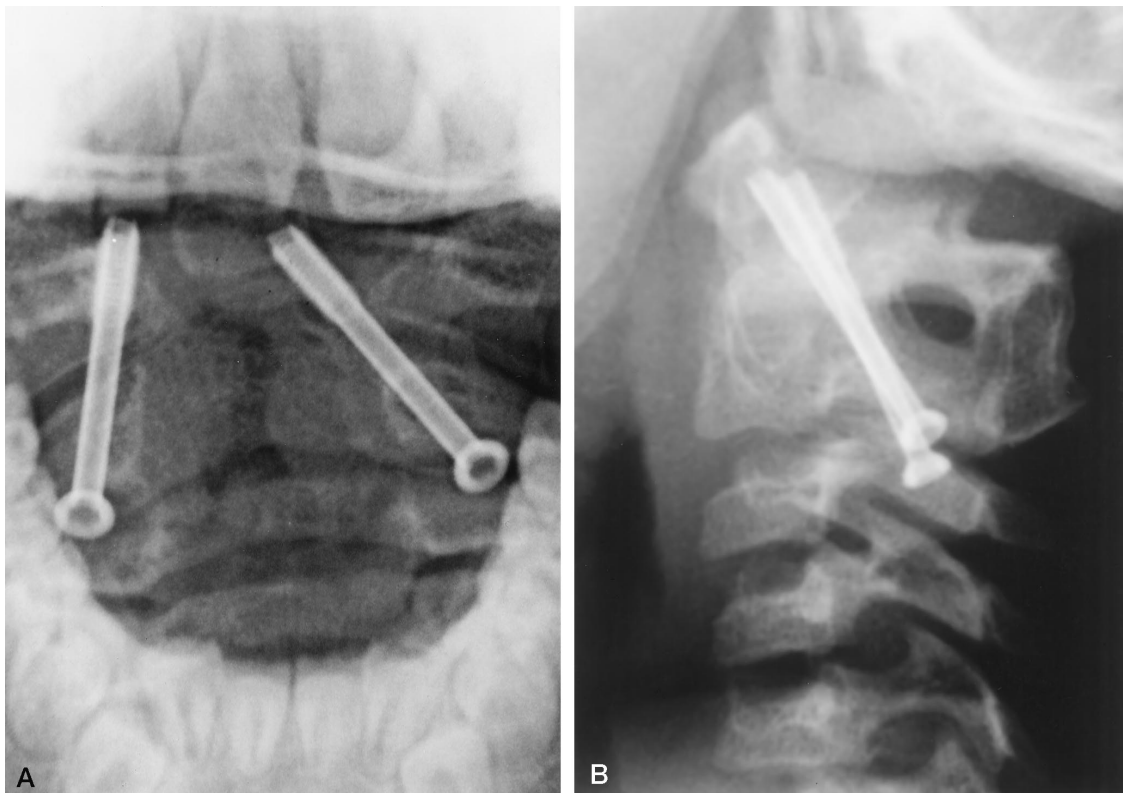


Figure 4. Postoperative anteroposterior (A) and lateral (B) radiographs of the cervical spine. Solid union of the grafted bone is seen in (B).

from the ataxia, the cause of the occlusion of the basilar artery remained unclear. In addition, atlantoaxial instability with os odontoideum was found, and the correlations of these findings were thought to be the cause of cerebellar infarction.

Transarticular screw fixation was performed to stabilize the atlas and axis with titanium cannulated cancellous screws with a diameter of 2.7 mm. This technique was performed successfully even in the child-sized atlas and axis. Initial stability was good, so strict postoperative immobilization was not necessary. A solid union of the grafted autologous iliac bone was obtained 1 year after the operation.

Atlantoaxial instability may cause mechanical stress to the vertebral artery as well as spinal cord injury. More attention should be paid to the possible relation between atlantoaxial instability and vertebrobasilar artery insufficiency.

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