Bilateral Vertebral Artery Occlusion Secondary to Atlantoaxial Dislocation with Os Odontoideum: Implication for Prophylactic Cervical Stabilization by Fusion

—Case Report—

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Abstract

A 27-year-old male presented with an unusual atlantoaxial dislocation associated with os odontoideum, after suffering sudden onset of significant neurological deterioration. Angiography demonstrated simultaneous occlusion of the bilateral vertebral arteries assumed responsible for the devastating neurological deficits. He initially underwent cervical immobilization with a Crutchfield tong for 2 months and subsequent posterior fusion using an iliac bone graft and wires. The neurological symptoms gradually subsided, and 5 years later good healing of the bone graft without instability and ample subarachnoid space around the spinal cord were radiologically confirmed. Early prophylactic stabilization of atlantoaxial dislocation due to os odontoideum is recommended to prevent a poor outcome. Careful angiographic evaluation of the vertebrobasilar circulation is important for management of patients with os odontoideum.

Key words: atlantoaxial dislocation, os odontoideum, posterior fusion, vertebral artery

Introduction

Os odontoideum is a separate odontoid process divided by a transverse gap from the body of the axis, which may be congenital or acquired,^{5,8,15} and is an infrequent cause of atlantoaxial dislocation.^{2,9,10,12-14,16} The pathogenesis of neurological manifestations associated with atlantoaxial dislocation may be mechanical due to myelopathy caused by direct compression of the upper spinal cord, or circulatory associated with ischemic accident caused by obstruction of the vertebral arteries stretched between the transverse foramina of the atlas and the axis.¹² Vertebrobasilar insufficiency associated with atlantoaxial dislocation is rare,^{6,12} while complete occlusion of the bilateral vertebral arteries visualized

by angiography is even more uncommon with only two such cases reported.^{10,16}

Here we describe a patient with bilateral vertebral artery occlusion secondary to atlantoaxial dislocation associated with os odontoideum.

Case Report

A 27-year-old male was admitted with complaints of nausea, vertigo and nuchal pain on March 12, 1986. He had no history of head or neck trauma. Four years previously investigation of vertigo and nuchal pain had identified atlantoaxial dislocation associated with os odontoideum. Surgery was planned for posterior fusion to achieve cervical stabilization for the dislocation. However, the symptoms spontaneously subsided with conservative management during hospitalization, and therefore he was observed as an outpatient.

On admission physical examination revealed a well-developed man with a short neck. He was alert with no neurological deficits until 12 hours later, when a variety of neurological symptoms gradually developed indicating sensorimotor and cranial nerve dysfunctions. Neurological examination disclosed right spastic hemiparesis, impaired pain and thermal sense on the left, impaired tactile and proprioceptive sense on the right, dysfunctions of the left Vth, VIth, IXth, Xth and XIIth cranial nerves, and horizontal nystagmus toward the left. This clinical picture of long-tract signs with cerebellar and cranial nerve abnormalities suggested the presence of multiple brainstem lesions involving the left side of the lower pons and medulla oblongata, and the right tegmentum of the pons.

A lateral roentgenogram of the upper cervical spine demonstrated extreme posterior displacement of the atlas in relation to the axis. The atlanto-odontoid distance was 2 mm and unchanged by flexion or

Fig. 1 Lateral roentgenograms of the upper cervical spine at the previous (*left*) and present hospitalization (*right*), showing no substantial changes in extreme posterior displacement of the atlas in relation to the axis.

extension of the neck. The backward shift of the atlas had remained grossly unchanged during the last 4 years (Fig. 1). Computed tomographic (CT) scans of the neck revealed a normal spinal canal at the level of the atlantoaxial junction with no evidence of cord compression. Brain CT scans demonstrated multiple infarcts involving the pons and cerebellum (Fig. 2).

Right vertebral angiograms demonstrated complete occlusion of the vertebral artery distal to the junction of the posterior inferior cerebellar artery (Fig. 3 *upper*). Left vertebral angiograms similarly showed complete occlusion of the vertebral artery after branching of the posterior inferior cerebellar artery. In addition, the extracranial portion of the vertebral artery between the transverse foramen of the axis and the foramen magnum appeared stretched and narrowed (Fig. 3 *middle*). These findings were in sharp contrast to those from the vertebral angiograms obtained during the previous hospitalization, in which neither stenosis nor obstruction of the vertebral artery was demonstrated, and the basilar artery was excellently visualized (Fig. 3 *lower*). Carotid angiograms revealed retrograde filling of the basilar artery through the bilateral posterior communicating arteries, suggesting that the blood supply to the brainstem was through the anterior circulation (Fig. 4). The angiographic findings and neurological signs suggested that the pathogenesis of neurological deterioration was circulatory insufficiency caused by occlusion of the bilateral vertebral arteries, rather than myelopathy by progressive atlantoaxial dislocation due to os odontoideum.

He underwent cervical immobilization using a Crutchfield tong for 2 months. On May 17, 1986 posterior fusion of the upper cervical spine (atlas, axis and C3) was carried out using a bone graft from

Fig. 2 Precontrast CT scans 1 month after onset, showing multiple infarcts involving the pons and cerebellum.

Fig. 3 Vertebral angiograms, anteroposterior (*left*) and lateral views (*right*). *upper*: Right vertebral angiograms, demonstrating complete occlusion of the vertebral artery distal to the junction of the posterior inferior cerebellar artery. *middle*: Left vertebral angiograms, showing the vertebral artery occluded after branching of the posterior inferior cerebellar artery. *lower*: Left vertebral angiograms 4 years previously, revealing patency of the vertebrobasilar circulation.

the iliac bone and a double stainless wire loop to stabilize the atlantoaxial junction and prevent further deterioration of the dislocation by bony fusion. After the neck was immobilized in the neutral position with a Halo-vest, he was placed in the prone position and the suboccipital cranium, posterior arch of the atlas, and laminae of the axis and the third cervical vertebra were exposed. These structures appeared normal except for the axis, whose lamina appeared hypoplastic and extremely dislocated anteriorly in relation to the atlas. In addition, the lamina of the axis moved back and forth with the respiration, suggesting instability between the atlas and the axis. Presumably, the congenital anomaly of the axis comprising the os odontoideum

Fig. 4 Right carotid angiogram, lateral view, demonstrating the basilar artery (*arrows*) visualized through the posterior communicating artery.

Fig. 5 Lateral plain roentgenogram (*left*) and T₁-weighted MR image (*right*) of the neck 5 years after surgery, revealing good healing of the bone graft and no evidence of cord compression regardless of the neck position.

and unstable hypoplastic lamina may cause the backward shift of the atlas, resulting in stretching of the vertebral artery between the transverse foramina of the atlas and the axis.

The postoperative course was uneventful. The Halo-vest was removed 3 months after surgery. Follow-up vertebral angiograms demonstrated no substantial changes. The neurological symptoms gradually subsided after rehabilitation therapy, and he finally became ambulant with motor functions graded as 4/5 right hemiparesis 5 years after discharge. Recent plain roentgenograms showed good healing of the bone graft without evidence of instability, and magnetic resonance (MR) images revealed ample subarachnoid space around the spinal cord both at flexion and extension, indicating no direct compression of the spinal cord (Fig. 5).

Discussion

The presence of os odontoideum creates a locus minoris resistentiae between the atlas and the axis, reduces the atlantoaxial stability, and facilitates dislocation when ligament support also fails.²⁾ Neurological symptoms due to atlantoaxial dislocation generally result from myelopathy due to direct compression of the upper cervical cord. Vertebrobasilar insufficiency due to intermittent obstruction of the vertebral artery caused by instability of the atlantoaxial joint is another pathogenesis for neurological manifestations.^{6,12)} However, complete occlusion of the vertebral artery secondary to nontraumatic atlantoaxial dislocation is extremely rare.

Singer *et al.*¹³⁾ reported the first case of angiographically verified occlusion of the left vertebral artery in 1975 at the level of the axis in a 6-year-old boy with os odontoideum. Only four cases with either unilateral or bilateral vertebral artery occlusion secondary to atlantoaxial dislocation have since been reported (Table 1).^{9,10,13,16)} All cases had os odontoideum and were juvenile males ranging from 6 to 18 years old. The common sites of obstruction are the level of the atlas and the axis where the vertebral artery runs through the transverse foramina, abruptly changes direction, and courses horizontally backwards in the vertebral groove. Two cases of unilateral occlusion were associated with vascular lesions, such as aneurysmal dilatation on the symmetrical portion of the patent contralateral vertebral artery. This may indicate that instability between the atlas and the axis results in constant stretching and vessel wall injury to the vertebral artery, which leads to aneurysmal dilatation. Minor head or neck injury was involved in three cases, but none were evaluated for atlantoaxial dislocation subsequent to the injury despite frequent episodes

of neurological symptoms presumably due to vertebrobasilar insufficiency.

In our case, extensive investigations including vertebral angiography were carried out for vertigo 4 years previously, but a devastating vascular accident could not be avoided despite careful observation. We postulate that angulation and stretching of the vertebral artery caused by the backward shift of the atlas with other concomitant congenital anomalies resulted in intimal injuries on the vessel wall, resulting in thrombus formation, and eventually complete occlusion of both arteries. Angiography must therefore be used to investigate the effect of atlantoaxial dislocation on the vertebral artery in patients with os odontoideum regardless of the presence or absence of clinical symptoms due to vertebrobasilar insufficiency.

The surgical management of atlantoaxial dislocation due to os odontoideum remains controversial.¹¹⁾ Atlantoaxial dislocation with os odontoideum may result in a devastating outcome,^{1,7,10,12,16)} but the true natural history of os odontoideum generally remains unknown.¹¹⁾ Spierings and Braakman¹⁴⁾ reported a series of 37 patients with os odontoideum, 20 treated conservatively and 17 surgically. They found no significant differences between the two modes of treatment in patients without cord compression signs. In contrast, Dyck²⁾ performed surgical management by stabilization and bone graft fusion in seven of eight patients with os odontoideum including children not demonstrating atlantoaxial instability. Surgical treatment can be justified by age-related increases in ligamentous laxity at the atlantoaxial junction and dislocation, resulting in a neurological crisis.²⁾ There are two main approaches to decompression and stabilization of the dislocated odontoid process: the anterior and posterior approaches.^{3,4,7,10,12)} In principle the approach is determined by the reducibility of the dislocation and the

Table 1 Vertebral artery occlusion associated with atlantoaxial dislocation

Case No.	Author (Year)	Age/Sex	Site of occlusion	Presence of collaterals	Associated vascular lesion	Presence of os odontoideum	Prior head or neck injury	Surgery
1	Singer <i>et al.</i> (1975) ¹³⁾	6/M	rt VA (C2)	yes	lt VA (C2) aneurysm, BA occlusion	yes	no	posterior fusion
2	Mitsumori <i>et al.</i> (1975) ¹⁰⁾	18/M	bil VA (C3)	yes	no	yes	yes	anterior fusion
3	Kurimoto <i>et al.</i> (1988) ⁹⁾	10/M	rt VA (C1)	no	lt VA (C2) aneurysm, BA occlusion	yes	yes	posterior fusion
4	Yamauchi and Ogawa (1991) ¹⁶⁾	17/M	bil VA (C2)	yes	no	yes	yes	no
5	Present case	27/M	bil VA (C1)	no	no	yes	no	posterior fusion

BA: basilar artery, C1: atlas, C2: axis, C3: the third cervical vertebra, VA: vertebral artery.

presence or absence of cord compression.

Three of the four previously reported cases of vertebral artery occlusion in association with os odontoideum were treated by surgery for stabilization of the dislocation: one by anterior fusion¹⁰⁾ and the remaining two by posterior fusion.^{9,13)} Mitsumori *et al.*¹⁰⁾ used the transoral anterior approach for their patient with a separate odontoid process shifted posteriorly and compressing the spinal cord, and blood flow to the brainstem maintained by collaterals from the ascending cervical artery. They thought that the posterior approach would not relieve cord compression due to the dislocated odontoid process, and would jeopardize collaterals to the brainstem. Conversely, Kurimoto *et al.*⁹⁾ used the posterior approach because there was no evidence of cord compression or collateral flow from cervical arteries.

In our case there was no evidence of spinal cord compression by the separate odontoid process. The os odontoideum, *per se*, should not cause myelopathy because it is held in place by the transatlantal, apical, and alar ligaments.²⁾ Therefore the odontoid process does not require removal by the direct anterior approach to achieve decompression for myelopathy, which was not present anyway in this patient. In addition, acute occlusion of bilateral vertebral arteries had occurred, but the blood supply to the brainstem was maintained by the anterior circulation through the posterior communicating arteries. There was no collateral blood flow to the brainstem through arteries in the neck such as ascending cervical arteries. Based on the absence of cord compression and collateral circulation to the brainstem *via* the cervical arteries, technical difficulties, morbidity and mortality, and overall success rate of stabilization by bony fusion, the posterior approach was superior to the anterior approach in our patient.

A high degree of atlantoaxial dislocation due to os odontoideum even without neurological manifestations requires prophylactic cervical stabilization. The need to perform angiography for early recognition and diagnosis of associated vertebral artery stenosis or occlusion is greatly emphasized. Angiography may also help decide whether anterior or posterior fusion is indicated for stabilization of atlantoaxial dislocation due to os odontoideum.

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