



Case Report

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Arteriovenous Fistula at the Craniocervical Junction Found After Cervical Laminoplasty for Ossification of the Posterior Longitudinal Ligament

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Ossification of the posterior longitudinal ligament (OPLL) is common in East Asia. Arteriovenous fistula at the craniocervical junction (CCJ-AVF), in contrast, is rare. As OPLL occurs most often in the cervical region, these 2 conditions can coexist in the cervical spinal canal of a single patient. We report a case of CCJ-AVF found after cervical laminoplasty (CLP) for OPLL. A 68-year-old man experienced progressive myelopathy due to cervical OPLL. Magnetic resonance imaging (MRI) revealed a high-intensity area inside the spinal cord. CLP was performed and his symptoms immediately improved. Three months after CLP, however, myelopathy recurred. MRI revealed an exacerbated and enlarged high-intensity area inside the cord from the medulla oblongata to the C4/5 level with a flow void around the cord. Left vertebral artery angiography revealed CCJ-AVF with ascending and descending draining veins. Direct surgery was performed to interrupt shunt flow into the draining veins. The patient's symptoms improved to a limited degree. In this case, increased pressure inside the spinal canal due to OPLL might have decreased the shunt flow of the CCJ-AVF. Thus, the venous congestion induced by CCJ-AVF might have been exacerbated after the pressure was removed by CLP. Magnetic resonance angiography screening could help detect concurrent CCJ-AVF and OPLL.



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INTRODUCTION

Ossification of the posterior longitudinal ligament (OPLL) is a common disease in East Asia including Japan.^{1,2} OPLL sometimes causes stenosis of the cervical spinal canal. Some OPLL patients develop progressive myelopathy and/or radiculopathy with interactive effects of spinal canal stenosis and cervical instability. Decompression and/or fusion surgeries are often per-

formed for OPLL cases with myelopathy/radiculopathy.^{3,4}

Arteriovenous fistula at the craniocervical junction (CCJ-AVF), in contrast, is rare⁵⁻⁷ and seldom encountered by spinal surgeons. CCJ-AVF causes subarachnoid hemorrhage (SAH) or, less frequently, venous congestion.⁸⁻¹⁰ Most patients with venous congestion due to CCJ-AVF develop myelopathy. CCJ-AVF and OPLL are distinct diseases, yet both can develop in the cervical canal and cause similar symptoms. It is possible for

CCJ-AVF and OPLL to coexist in a single patient, in which case the similarity of their symptoms would make it difficult to detect both, as all symptoms would initially tend to be attributed to whichever had been discovered first. To our knowledge, there is no previous report on the coexistence of CCJ-AVF and OPLL. In this article, we report a case of CCJ-AVF found after cervical laminoplasty (CLP) for OPLL.

CASE REPORT

A 68-year-old man presented with progressive numbness of his left sole. One week later, he developed urinary retention and severe leg cramps with consequent gait disturbance. Magnetic resonance imaging (MRI) of the head and lumbar spine revealed no anomalies. When his symptoms had persisted for more than a month, he was referred to our hospital.

1. Physical Findings

In addition to numbness of the left sole, the patient reported decreased position sense of his left leg. He had no muscle weakness other than mild weakness of the gastrocnemius. The deep tendon reflexes of biceps, brachii, and triceps muscle were normal. He presented with bilateral increased patellar tendon reflex and spastic gait. His Japanese Orthopedic Association (JOA) score of lower extremity was 1 and JOA score of bladder function was 2.

2. Imaging Findings

There were no anomalies on MRI of the head or thoracic and lumbar spine. Cervical computed tomography (CT) revealed a mixed-type OPLL at the posterior aspect of the C2–6 vertebrae (Fig. 1A). Cervical T2-weighted imaging (T2-WI) revealed a high-intensity area inside the spinal cord (Fig. 1B). There were

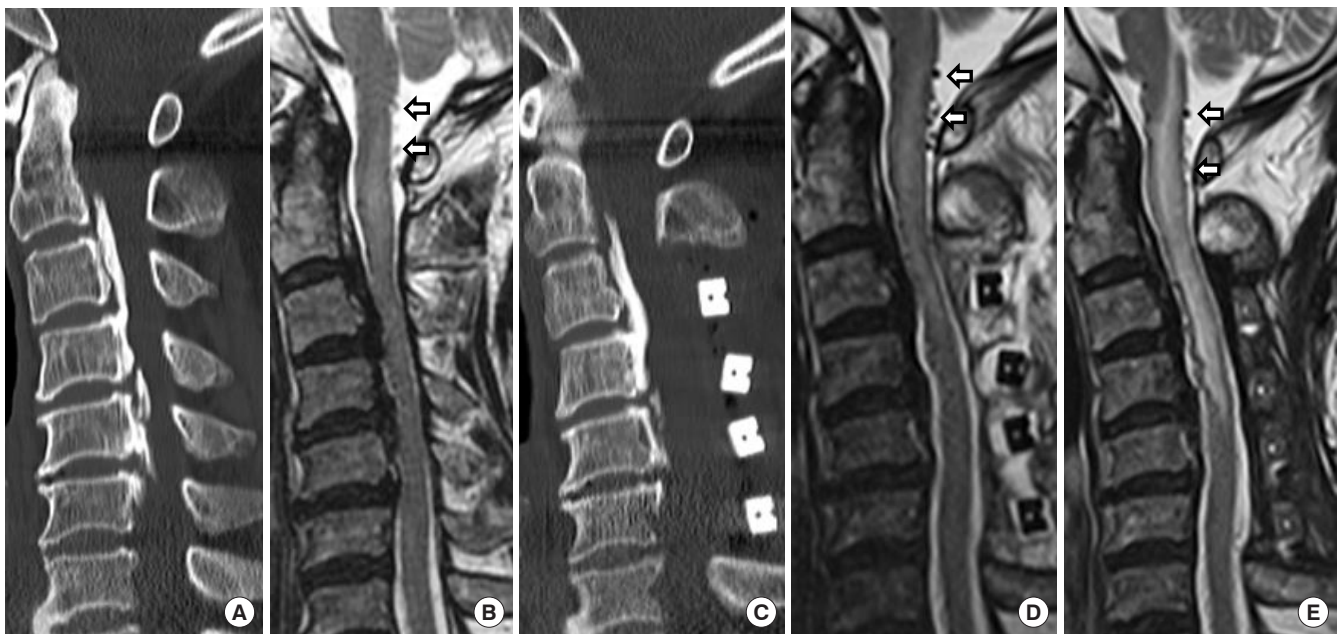


Fig. 1. (A) Sagittal view of cervical spine before cervical laminoplasty (CLP). Computed tomography (CT) revealed mixed-type ossification of the posterior longitudinal ligament (OPLL) on a posterior aspect of the vertebrae between the lower edge of C2 and the middle of C6. (B) Sagittal view of T2-weighted imaging (T2-WI) showed that the cervical spine was under high pressure due to spinal stenosis. A high-intensity area was observed in the cord between C1 and C3. Abnormal vessels (arrows) are visible behind the medulla oblongata, indicating coexistence of an arteriovenous fistula at craniocervical junction and OPLL. During treatment for OPLL, however, we did not notice these flow voids. (C) Sagittal view of cervical spine after CLP according to double door technique. CT imaging shows that double door CLP has been achieved. The cervical spinal canal was successfully enlarged. (D) Sagittal view of T2-WI at 2 months after CLP. T2-WI shows that the spinal cord is successfully decompressed at each level. However, the high-intensity area between C1 and C3 is exacerbated, and flow voids (arrows) can be seen behind the medulla oblongata. Although the flow void behind the medulla oblongata is visible in this image, we were not aware of it during treatment for OPLL. (E) Sagittal view of T2-WI at 3 months after CLP. The high-intensity area was exacerbated and enlarged from the medulla oblongata to C5. Flow voids (arrows) are visible at the anterior and posterior aspects of the spinal cord. It was at this time that we first noticed these vascular anomalies.

no instabilities of the cervical spine on roentgenokymography. We presumed that the patient's symptoms were caused by myelopathy due to OPLL.

3. Course of Treatment

CLP was performed between C3 and C6 according to the double door technique (Fig. 1C). We observed some dilated red vessels on the dura mater but presumed that these were normal epidural dilated veins due to congestion resulting from OPLL. CLP was achieved with no other problems or complications (blood loss: 50 mL). Postoperative T2-WI showed good decompression of the cervical spine with high intensity in the cord (C1–3) (Fig. 1D). The patient was able to walk immediately after CLP, although a subtly spastic gait and urinary retention remained. The postoperative course was uneventful and he was discharged from our hospital 10 days after CLP. His gait became increasingly stable and his urinary function improved during the 2 months after CLP. Two months after CLP, his JOA score of lower extremity was 4 and JOA score of bladder function was

4. Three months after CLP, however, the patient became unable to walk again due to exacerbated spastic gait, urinary retention also recurred, and his JOA score of lower extremity was 1 and JOA score of bladder function was 2. T2-WI revealed an exacerbated and enlarged high-intensity area inside the cord from the medulla oblongata to the C4/5 level with a flow void around the cord (Fig. 1E). Retrospectively, the flow void could be detected on MRI shortly after CLP (Fig. 1D). We performed angiography to detect any vascular lesions.

4. Angiographic Findings

Left vertebral artery angiography revealed a vascular tangle formed of tiny vessels at the C1 level and draining into the intradural vein with ascending and descending draining veins (Fig. 2A, B). We diagnosed this as a CCJ-AVF. The anterior spinal artery (ASA) was not involved in the CCJ-AVF (Fig. 2A). Selective angiography at the C1 level revealed that the CCJ-AVF was located on the dura mater and was fed by the C1 radiculomeningeal artery which drained into the intradural ascending

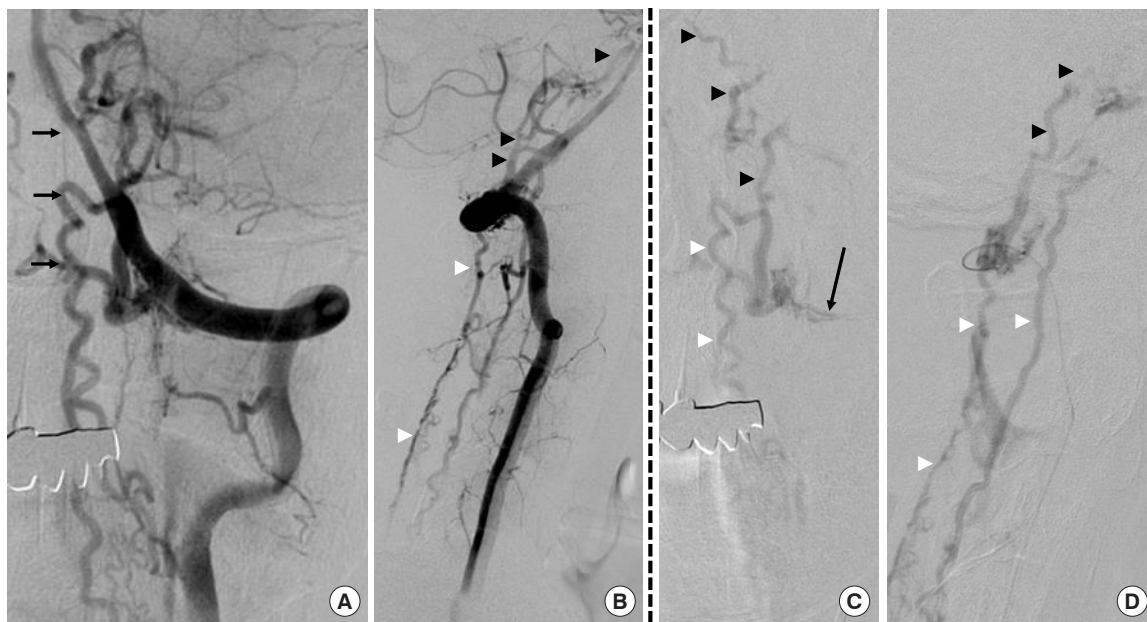


Fig. 2. (A) Anteroposterior (AP) view of an angiography of the left vertebral artery after cervical laminoplasty (CLP) showing a vascular tangle at the C1 level with ascending and descending draining veins. Arrows indicate the anterior spinal artery (ASA). This angiography reveals that the ASA was not involved in the CCJ-AVF. (B) Lateral view of angiography of left vertebral artery after CLP. Shunt flow drained through the arteriovenous fistula at the craniocervical junction (CCJ-AVF) into ascending draining veins (black arrowheads) and descending draining veins (white arrowheads). (C) AP view of selective angiography of C1 radiculomeningeal artery. A black arrow indicates the C1 radiculomeningeal artery. Shunt flow drained through the CCJ-AVF into ascending draining veins (black arrowheads) and descending draining veins (white arrowheads). Selective angiographies of other levels did not detect any anomalies. (D) Lateral view of selective angiography of C1 radiculomeningeal artery. Shunt flow drained through the CCJ-AVF into ascending draining veins (black arrowheads) and descending draining veins (white arrowheads).

and descending draining veins. The draining veins were involved in the anterior and posterior spinal veins, respectively. The descending draining veins seemed to run partly into the extradural space at the C3 level (Fig. 2C, D). No other radiculomeningeal arteries fed the CCJ-AVF. We concluded that the secondary myelopathy developing 3 months after CLP had been caused by this CCJ-AVF.

5. Course of Treatment After Secondary Myelopathy

We performed a direct surgery to interrupt the shunt flow into the draining veins. Lateral suboccipital craniotomy without C1 laminectomy was performed in the prone position. Red dilated vessels were immediately visible in our view of the medullary cistern (Fig. 3A). A vascular tangle was observed on the dura mater proximal to these veins (Fig. 3B). Presuming that the vascular tangle was a shunt point, we interrupted the draining vein to stop the shunt flow (Fig. 3C). Indocyanine green

fluorescence after this procedure confirmed that there was no flow to the draining veins. The red dilated vessels were then normalized (Fig. 3D). After surgery, left vertebral artery angiography showed no abnormal blood flow (Fig. 4A, B). T2-WI revealed a decreased high-intensity area inside the cord. After surgery, the patient was able to walk with a walking stick, although his gait remained somewhat spastic. His JOA score of lower extremity was 2 and JOA score of bladder function was 3 shortly after the surgery and has remained stable at 2 for the last 2 years.

DISCUSSION

OPLL is a very common disorder in Japan. Many clinical studies of OPLL have been reported from Japan and other East Asian countries, along with a few from Europe and North America. A survey of Japan, Korea, the USA, and Germany has reported

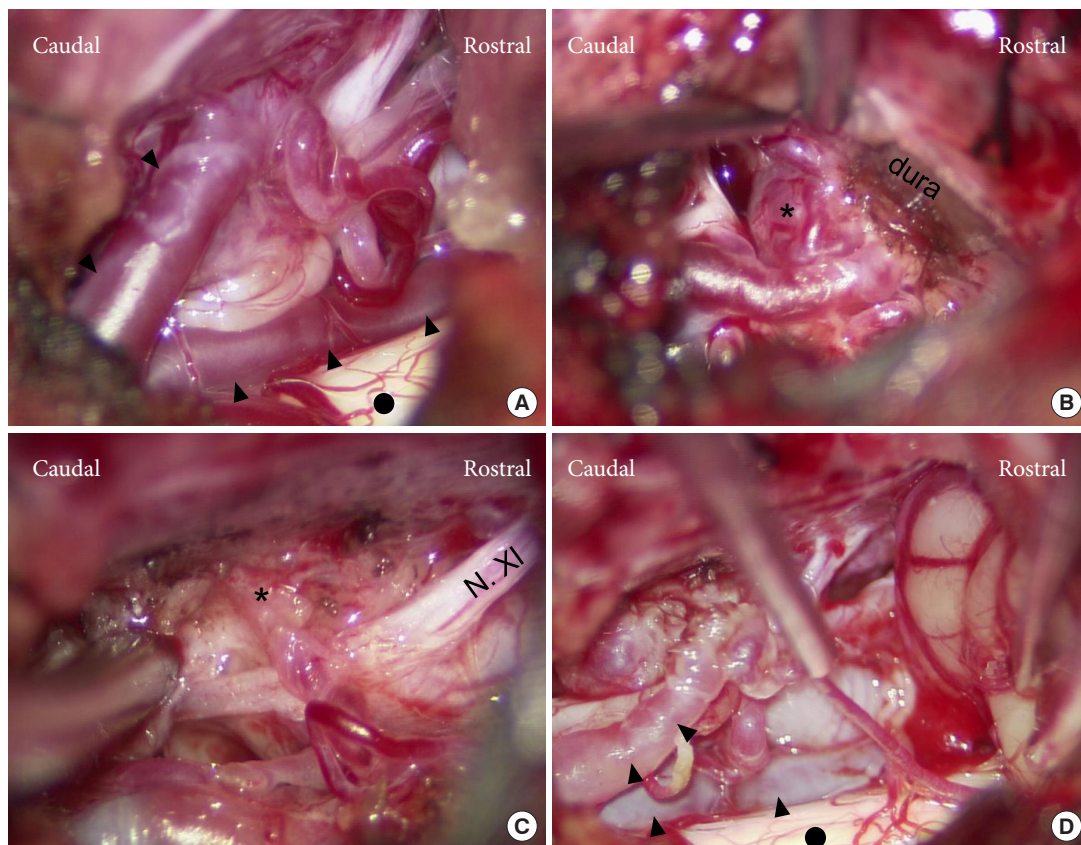


Fig. 3. (A) View of the left medullary cistern. We observed dilated veins (arrowheads) in this space immediately after exposure of the left medullary cistern. A circle indicates the medulla oblongata. (B) A vascular tangle (asterisk) was visible on the dura at the border between the occipital bone and C1. (C) Draining veins were interrupted by ablation, after which the vascular tangle faded away (asterisk). (D) Draining veins (arrowheads) subsequently lost their redness and dilation. A circle indicates the medulla oblongata.

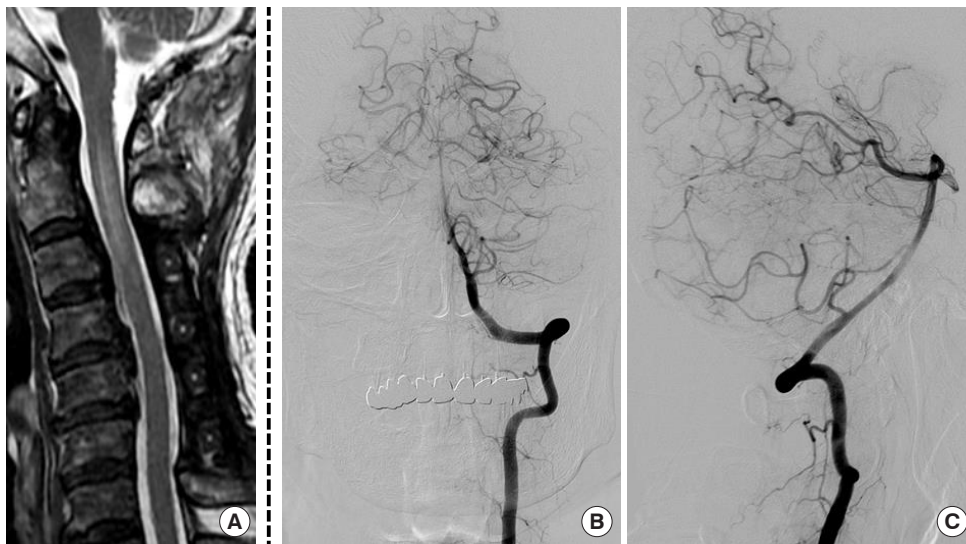


Fig. 4. Images after direct surgery for arteriovenous fistula at craniocervical junction. (A) Sagittal view of T2-weighted imaging of cervical spine. The high-intensity area in the spine decreased in size after the surgery. (B) Anteroposterior view of angiography of the left vertebral artery. There were no anomalies. (C) Lateral view of angiography of the left vertebral artery. There were no anomalies.

that the prevalence of OPLL in Japan is about 3%, whereas that in North America and Germany is 0.1%.¹¹ Most asymptomatic patients with OPLL are treated conservatively.^{12,13} Symptomatic patients can be candidates for surgery if they have some difficulty in daily living.^{3,4} In contrast, CCJ-AVF is a very rare vascular malformation. The incidences of dural AVF in Japan and Finland have been reported as 0.29 and 0.51, respectively, per 100,000 person-years. CCJ-AVFs are found in 1%–2% of patients with intracranial or spinal AVFs.^{5–7} Therefore, the prevalence of CCJ-AVF is estimated to be 0.000003%–0.00001%. These 2 disorders thus have vastly different prevalence.

CCJ-AVF is frequently associated with SAH and intramedullary hemorrhage and more rarely associated with venous congestion. Many studies have reported SAH as a symptom of CCJ-AVF.^{8–10} Historically, it has been recognized that an ascending draining route into an intracranial vein causes SAH whereas a descending draining route into the spinal canal causes venous congestion in the spinal cord.^{9,10,14,15} In contrast, Hiramatsu et al.⁸ recently reported that inclusion of the ASA as a feeder, aneurysmal dilatation of the feeder, exclusion of the external carotid artery as a feeder, and the presence of an arteriovenous shunt on the spinal nerve roots are all significantly associated with hemorrhagic presentation, while the presence of an ascending intradural draining vein is not significantly associated with hemorrhagic presentation. In this case, the ASA was not involved in the CCJ-AVF, which was fed only by the C1 radiculomeningeal artery. The AV shunt point was located on the dura

mater, although there were ascending draining routes into the intracranial veins. Therefore, we assume that this case had a low risk for SAH. Rather, the patient developed myelopathy due to venous congestion.

There are 2 previous case reports about CCJ-AVF presenting with SAH in which magnetic resonance angiography (MRA) or 3-dimensional CT angiography was not able to detect vascular anomalies.^{15,16} The authors of those reports hypothesized that this situation was caused by embolism of AVFs, spasm of feeders, or increased intracranial pressure. In this case, we were not able to detect any vascular anomalies without a clear flow void when OPLL was first detected, although a retrospective examination of the magnetic resonance images taken immediately after CLP revealed abnormal vessels at the posterior aspect of the medulla oblongata. There is a report about anatomical characteristics of subpial anterior spinal vein and subarachnoid posterior spinal vein.¹⁷ They presumed that the enlargement of the veins at the anterior aspect of spinal cord may remain undetected on T2-WI due to the anatomical characteristics. We presume that we were not able to detect vascular anomalies primarily because the high pressure in the spinal canal due to OPLL decreased the shunt flow of CCJ-AVF, and because the dilated veins at the anterior aspect of spinal cord were hard to be detected originally. Knowing that AVF often occurs after surgery or trauma, we considered the possibility that our CLP procedure had caused the AVF. In this case, CLP was performed between C3 and C6, and the AVF occurred at the CCJ. T2-WI af-

ter CLP retrospectively revealed abnormal vessels behind the medulla oblongata. During CLP, we observed some red dilated vessels on the dura mater of the spinal cord, although we were not able to recognize them as anomalies at that time. After CLP, selective angiography of the C1 radiculomeningeal artery revealed that some of the intradural descending draining veins ran partly into the extradural space at the C3 level. We consider that the red vessels seen during CLP might have been these extradural draining veins. Therefore, we are convinced that the CCJ-AVF was present prior to CLP.

In this case, CLP improved the myelopathy caused by OPLL but also led to decompression of the spinal canal, which may have increased the blood flow of the CCJ-AVF. The CCJ-AVF then exacerbated the patient's venous congestion over the 3 months following the CLP, causing myelopathy to worsen once again. There is a report about asymptomatic spinal dural AVE.¹⁸ They assumed that symptomatic venous congestion due to venous hypertension might be induced by the obstruction of alternate venous outflow routes like radicular veins from arterialized perimedullary veins. They also pointed out a possibility that increased blood flow of AVF might obstruct alternate venous outflow with acceleration of fibrosis or thrombosis of radicular veins. In our case, the increased blood flow by CLP might cause obstruction of alternate venous outflow, leading to venous congestion.

We treated the OPLL before the CCJ-AVF because we did not notice the CCJ-AVF before we performed the CLP. Even if we had noticed the CCJ-AVF prior to CLP, however, we would still have performed CLP first in this case because OPLL was causing severe compression of the cord leading to myelopathy. Prior to CLP, the OPLL was contributing much more to the patient's myelopathy than the CCJ-AVF was. If the OPLL had been less severe such that the CCJ-AVF had contributed more to the patient's myelopathy, and if the CCJ-AVF had been easily detectable on MRI, we may have opted to treat the CCJ-AVF first. In our opinion, whichever disorder is contributing more to the myelopathy should be treated first.

Many spinal surgeons are not familiar with vascular anomalies. MRI is typically performed before and after spinal surgeries, but MRA might be more effective to avoid overlooking vascular anomalies.

CONCLUSION

We experienced a case of CCJ-AVF found after CLP for OPLL. The 2 conditions had coexisted in the patient's spinal canal. High

pressure in the spinal canal due to OPLL may have decreased shunt flow of the CCJ-AVF. Imaging modalities such as MRA might improve detection of CCJ-AVF and other unexpected vascular anomalies, enabling us to prevent vascular events after cervical spinal surgeries.

CONFLICT OF INTEREST

The authors have nothing to disclose.

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We had a presentation at the 34th Annual Meeting of Neurospinal Society of Japan.

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