Intramedullary spinal cord abscess associated with right-to-left shunt via right superior vena cava draining into left atrium

A case report

Satoshi Hirose, MD, PhD, Naohiro Sudo, MD, Masahiro Okada, MD, PhD, Naotoshi Natori, MD, Takayoshi Akimoto, MD, PhD, Makoto Hara, MD, PhD, Hideto Nakajima, MD, PhD

Abstract

Rationale: Intramedullary spinal cord abscess (ISCA) is a rare but treatable bacterial infection of the central nervous system, and the etiology in no less than 40% of the cases is cryptogenic. Although a few cases of ISCA in individuals with a right-to-left shunt (RL shunt) have been reported, only few arguments focused on the association between RL shunt and ISCA have been provoked. The right superior vena cava (RSVC) draining into the left atrium (LA) is an uncommon systemic venous anomaly that results in an RL shunt, and this anomaly causes several types of neurological complication such as stroke or brain abscess. We report the first case of ISCA associated with RSVC-LA RL shunt.

Patient concerns: A 36-year-old man developed progressive paraparesis, dysuria, and spontaneous pain in the lumbar region and lower extremities. Spinal magnetic resonance imaging revealed an intramedullary lesion extended from Th12 to L2 with ring-shaped gadolinium enhancement. Cerebrospinal fluid (CSF) study exhibited a marked pleocytosis, and CSF culture grew *Streptococcus intermedius*. Cardiovascular computed tomography angiography identified RSVC-LA RL shunt, which caused transient acute cardiac syndrome due to air embolus.

Diagnoses: The patient was diagnosed with ISCA associated with an RSVC-LA RL shunt.

Interventions: The patient was treated with a combination of intravenous administration of meropenem and vancomycin in a daily dose of 6 and 2.5 g, respectively, followed by intravenous administration of ampicillin in a daily dose of 750 mg. The intravenous antibiotic therapy was continued for 37 days.

Outcomes: A favorable neurological outcome was obtained by the intravenous antibiotic therapy, and recurrence of infection was prevented by continuous oral antibiotic therapy for 18 months.

Lessons: With a literature review of ISCA associated with RL shunt, we insist that screening for RSVC-LA is beneficial to patients who are diagnosed with cryptogenic ISCA as its identification leads to appropriate preventive therapy.

Abbreviations: CSF = cerebrospinal fluid, ISCA = intramedullary spinal cord abscess, LA = left atrium, MRI = magnetic resonance imaging, RL shunt = right-to-left shunt, RSVC = right superior vena cava.

Keywords: abscess, central nervous system infection, right-to-left shunt, spinal cord, superior vena cava

1. Introduction

Intramedullary spinal cord abscess (ISCA) is a rare bacterial infection of the central nervous system (CNS) that can lead to sensory, motor, and/or autonomic dysfunction and is sometimes fatal. Although the etiology is cryptogenic in 40% of the cases, rare cases of ISCA in individuals with a right-to-left shunt (RL shunt) have also been reported. The right superior vena cava (RSVC) draining into the left atrium (LA) is an uncommon systemic venous anomaly that results in an RL shunt. This RSVC-LA RL shunt is typically identified in the neonatal period or childhood because of hypoxemia; however, hypoxemia, stroke, myocardial infarction, and brain abscess are seen in completely asymptomatic adult patients with normal growth and development. Here, we describe the first case of ISCA with an RSVC-LA RL shunt.
2. Case presentation

A 36-year-old Japanese man was admitted to our hospital with progressive paraparesis, dysuria, and spontaneous pain in the lumbar region and lower extremities that occurred 7 days before admission. He had a medical history of an endoscopic polypectomy performed for colonic polyps. His history was otherwise unremarkable. A neurological examination at admission revealed that he was alert and well oriented, and did not exhibit aphasia or cranial nerve involvement. However, extremely impaired perceptions to pinprick and vibration below the L2 level were noted, along with severe weakness of the lower extremities, predominantly in the distal regions. Tendon reflexes of the lower extremities were lost, and Babinski sign was absent in both sides. Nuchal rigidity was not present. He was unable to micturate. There was no fever or any other signs of systemic infection, and otorhinolaryngologic and dental evaluation did not reveal any signs of infection. Spinal magnetic resonance imaging (MRI) (Fig. 1) revealed a high-intensity intramedullary lesion on T2-weighted imaging that extended from the Th12 to L2 vertebral levels (Fig. 1A), which appeared ring-shaped with gadolinium enhancement (Fig. 1B and C). Additionally, swelling of the spinal cord around the lesion was evident (Fig. 1A–C). The lesion exhibited high intensity on diffusion-weighted imaging (Fig. 1D) but low intensity on the apparent diffusion coefficient map. An enhanced whole-body computed tomography (CT) was unremarkable. A spinal tap yielded yellow and turbid cerebrospinal fluid (CSF) with a markedly high white blood cell count (6400/µL, polymorphonuclear cells 100%) and total protein level (357 mg/dL), along with low glucose (23 mg/dL; blood glucose 104 mg/dL). CSF was negative for both herpes simplex virus DNA and varicella zoster virus DNA upon high-sensitive polymerase chain reaction. Further, even though CSF culture grew *Streptococcus intermedius*, two sets of blood cultures remained sterile. Hematological analysis showed slightly elevated white blood cell count (10,900 cell/µL), normal C-reactive protein level, and no antibodies in the serum against aquaporin-4 or other systemic autoimmune diseases, including antinuclear, anti-double-stranded DNA, antiphospholipid, anti-Sjögren syndrome antigen A, anti-Sjögren syndrome antigen B antibodies, and anti-neutrophil cytoplasmic antibody. Urinalysis was normal.

Based on the above, he was diagnosed with ISCA and treated with a combination of intravenous administration of meropenem and vancomycin in a daily dose of 6 and 2.5 g, respectively, from the day of admission. Antibiotics were descaledated to ampicillin in a daily dose of 750 mg on day 9 based on antibiotic sensitivity of *S. intermedius* identified in the CSF. On day 14 after admission, he developed sudden onset chest tightness and decreased oxygen saturation with ST-segment elevations in leads II, III, and augmented vector foot on 12-lead electrocardiogram (ECG). A CT angiography under the pulmonary embolism protocol was performed with contrast medium injected through an intravenous catheter in the right forearm. Surprisingly, contrast was detected in the superior vena cava, the LA, the left ventricle (LV), and the aorta, but not in the pulmonary arterial system (Fig. 2A and B), which indicated the presence of an RL shunt. Additionally, air was detected in the LV (Fig. 2C), along with the following systemic venous anomalies (Fig. 2A and B), viz., a persistent left superior vena cava (PLSVC) that drained into the right atrium via the coronary sinus, the RSVC draining into the LA, and a defective brachiocephalic vein. The course of the inferior vena cava was normal. After a few hours, chest tightness disappeared, oxygen saturation increased, and a repeat ECG revealed no ST-segment elevations; hence, these symptoms were attributed to a transient acute cardiac syndrome caused by an air embolus during intravenous catheter placement in the right forearm that had migrated to the LA via drainage from the RSVC and subsequently reached the coronary arteries.

Follow-up CSF studies on days 4, 14, 25, 37, and 50 showed improvement in pleocytosis and decreased glucose, and MRI acquired on day 35 revealed resolution of the intramedullary lesion and no swelling of the spinal cord. No adverse event occurred during the intravenous antibiotic therapy, and the intravenous antibiotic therapy was discontinued on day 37. Continuous oral administration of ampicillin in a daily dose of 750 mg was then started to prevent recurrence of infection. The patient showed gradual improvement, was able to walk independently, and could return to work by 5 months after rehabilitation. At 18 months after disease onset, his muscle strength was normal, but urinary retention and rectal dysfunction persisted. During this follow-up period, no infectious diseases or exacerbation of neurological symptoms was seen. The patient provided written informed consent for publication of this case report.

3. Discussion

We describe the case of a 36-year-old man who developed progressive paraparesis, dysuria, and spontaneous pain in the lumbar region and lower extremities, and was diagnosed with ISCA associated with an RSV-LA RL shunt. Antibiotic therapy successfully improved his neurological symptoms, and a favorable outcome was obtained, although urinary retention and rectal dysfunction remained. Recurrence of infection and neurological symptoms was prevented by continuous oral antibiotic therapy for 18 months.

ISCA is a rare bacterial infection of the CNS that leads to sensory, motor, and/or autonomic dysfunction, and may sometimes be fatal. A previous review of 54 cases of ISCA has reported that the cause of infection was cryptogenic in 39% of the patients, followed by congenital dermal sinus infection in 28%, sepsis in 26%, extension from contiguous lesions (e.g., discitis, trauma, or surgery) in 6%, and infectious endocarditis in 1%. In our case, none of these common causes of ISCA was seen; rather, an anomalous RSV-LA RL shunt was identified. ISCA in individuals with an RL shunt has been reported in three case reports in the English-language literature (Table 1)—one...
case with pulmonary arteriovenous fistula (PAVF) and two with patent foramen ovale (PFO). Although prevention therapy and neurological outcomes vary across these cases with ISCA and RL shunt (Table 1), a relatively favorable outcome was obtained using intravenous antibiotic and continuous oral antibiotic therapy in our patient.

A mechanism by which ISCA occurred in a case with PFO was speculated by Higuchi et al. as follows: the presence of an RL shunt allows pathogens to bypass pulmonary defenses and enter the arterial circulation easily, which leads to high risk of ISCA as well as brain abscess. Brain abscess is a more common CNS infection than ISCA in the presence of an RL shunt, such as a PFO, or cyanotic cardiac disease, or PAVF. Although less frequent, cases of brain abscess associated with RSV-LA have also been reported. Bacteria are usually intercepted by phagocytosis in the pulmonary capillary vessels, but the RL shunt allows the bacterial mass to circumvent this, thereby permitting cerebral embolization of the bacterial mass and formation of a brain abscess. Interestingly, brain abscesses have been reported to occur without bloodstream infection in the presence of an RL shunt. Based on the aforementioned discussion, we suggest a mechanism by which ISCA could have occurred in our patient with the RSV-LA RL shunt is as follows: a bacterial mass of S. intermedius, which is normal oral flora in humans, initially invaded capillary vessels from the oral cavity, migrated to the aorta via the RSV-LA RL shunt, and finally reached the capillary vessels in the intramedullary spinal cord where embolization of the bacterial mass led to formation of the abscess, as with the cases of brain abscess associated with RSV-LA.

Anomalous systemic venous drainage into the LA is an uncommon cause of RL shunt and, if present, is most commonly due to the PLSVC draining into the LA. Thus, the RSV-LA is extremely rare in the absence of other cardiac abnormalities. Since its first identification in 1956, there have been approximately 60 reported cases of RSV-LA, and most patients are diagnosed in the neonatal period or childhood because the shunt leads to hypoxemia and cyanosis. However, in adult patients, even after complete asymptomatic normal growth and development, this systemic venous anomaly can cause hypoxemia, myocardial infarction, stroke or transient ischemic attack, and brain abscess, with the need for surgical correction of this systemic venous anomaly varying among individual cases.

### Table 1

| Case no. | Author | Age (yr) | Sex | Symptoms (mRS at peak) | Location | Pathogens | RL shunt | Treatment | Prevention | Follow-up (mo) | Outcome (mRS) | IV C = inferior vena cava, LA = left atrium, MRC = Medical Research Council’s scale, mRS = modified Rankin Scale, N.A. = not available, PAVF = pulmonary arteriovenous fistula, PFO = patent foramen ovale, RL shunt = right-left shunt, RSVV = right superior vena cava. |
|----------|--------|---------|-----|------------------------|----------|-----------|----------|------------|-------------|----------------|----------------|-----------------------------------------------|
| 1        | David et al 1997 | 27 M | M | Fever, tetraplegia, and sensory deficit (5) | C5 | Haemophilus aphrophilus and Actinomyces meyeri | PAVF | Drainage of abscess and antibiotics | Fistula embolization | N.A. | “Satisfactory improvement” | 3 | 3 |
| 2        | Higuchi et al 2011 | 51 M | M | Fever, tetraplegia, ischemia, constipation, dysphagia, and hiccup (5) | Entire spinal cord and medulla oblongata | Streptococcus viridans | PFO | Antibiotics and corticosteroids | None | 3 | Muscle weakness (able to walk with parallel bars) | 4–5 |
| 3        | Teterov et al 2011 | 59 M | F | Tetraplegia and sensory deficit (5) | C3–C7 | S viridans | PFO | Antibiotics, corticosteroids, and surgical removal of abscess | Intervventional cardiology on PFO, IV C filtering | N.A. | Muscle weakness (MRC 3 in right side and MRC 1 in left side) | 16 | 1 |
| 4        | Our case | 36 M | M | Paraplegia, urinary retention, and fecal incontinence (6) | Th12–L2 | Streptococcus intermedius | RSVC to LA | Antibiotics | Continuous oral antibiotics | N.A. | Urinary retention and fecal incontinence | 1 | 1 |

IVC = inferior vena cava, LA = left atrium, MRC = Medical Research Council’s scale, mRS = modified Rankin Scale, N.A. = not available, PAVF = pulmonary arteriovenous fistula, PFO = patent foramen ovale, RL shunt = right-left shunt, RSVV = right superior vena cava.
To the best of our knowledge, only 6 detailed case reports of neurological complications secondary to an R SVC-LA RL shunt have been published (Table 2) in the English-language literature: three cases of brain abscesses\(^{6,12,13}\) and three cases of cerebrovascular disorder\(^{7,11,13}\). Our article is the first case description of ISCA in a patient with an R SVC-LA RL shunt. Three cases were female and the median age was 46.5 (range, 34–65) years. Past history of myocardial infarction was reported in two cases\(^{6,17}\) and our patient similarly developed a transient acute cardiac syndrome caused by an air embolus, which led to identification of his R SVC-LA RL shunt. Past history of brain abscess was reported in one case\(^{18}\). Remarkably, hypoxemia due to R SVC-LA RL shunt was seen in three cases despite their normal growth and development\(^{7,12,13}\). Unsurprisingly, symptoms, treatments, and outcomes vary among the cases\(^{6,7,12,11,13,17}\) according to their neurological complications and affected regions. To prevent further complications due to R SVC-LA RL shunt, surgical intervention was performed in three cases\(^{7,12,13}\) and oral antithrombotic therapy was undergone in one case.\(^{17}\) While our patient continues to take oral antibiotics to prevent recurrence of the abscess or systemic infection, surgical intervention was deemed unnecessary due to the absence of hypoxemia or heart failure. We also instructed him that an intravenous catheter must not be placed in his right arm to avoid a risk of accidental complications caused by air embolus.

In summary, the R SVC-LA draining into the LA is a rare venous anomaly that can potentially lead to not only hypoxemia and cardiac events but also result in neurological complications due to the R SVC-LA RL shunt. This case identifies RL shunt as an important etiologic agent of ISCA, and hence, screening for R SVC-LA RL shunt may be beneficial to patients with cryptogenic ISCA as its identification can lead to appropriate preventive therapy.

### References

1. Iwasaki M, Yano S, Aoyama T, et al. Acute onset intramedullary spinal cord abscess with spinal artery occlusion: a case report and review. Eur Spine J 2011;20 Suppl 2:5294–301.
2. David C, Brasme L, Peruzzi P, et al. Intramedullary abscess of the spinal cord in a patient with a right-to-left shunt: case report. Clin Infect Dis Off Publ Infect Diseases Soc Am 1997;24:89–90.
3. Higuchi K, Ishihara H, Okuda S, et al. A 51-year-old man with intramedullary spinal cord abscess having a patent foramen ovale. BMJ Case Rep 2011;2011:bcr1120103512.
4. Terterov S, Tadjha A, Khalessi AA, et al. Intramedullary abscess of the spinal cord in the setting of patent foramen ovale. World Neurosurg 2011;76:361.e1361.e311–361.e14.
5. Chowdhury UK, Anderson RH, George N, et al. A review of the surgical management of anomalous connection of the right superior caval vein to the morphologically left atrium and bialtral drainage of right superior caval vein. World J Pediatr Congenit Heart Surg 2020;11:466–44.
6. Schick EC Jr., Lekakis J, Rothendler JA, et al. Persistent left superior vena cava and right superior vena cava drainage into the left atrium without arterial hypoxemia. J Am Coll Cardiol 1985;5:374–8.
7. Sadek H, Gilkeson RC, Hoit BD, et al. Images in cardiovascular medicine. Case of anomalous right superior vena cava. Circulation 2006;114:e532–3.
8. Baggett C, Sken SJ, Gantts DS, et al. Isolated right superior vena cava drainage into the left atrium diagnosed noninvasively in the peripartum period. Tex Heart Inst J 2009;36:611–4.
9. Sadahiro H, Nomura S, Inamura A, et al. Brain abscess associated with patent foramen ovale. Acta Neurochir 2014;156:1971–6.
10. Lumbiganon P, Chaikittipanyo A. Antibiotics for brain abscesses in people with cyanotic congenital heart disease. Cochrane Database Syst Rev 2013;2013:Cd004469.
11. Gao LY, Xu GR, Dai TJ. Precision diagnosis and therapy of a case of brain abscesses associated with asymptomatic pulmonary arteriovenous fistulas. BMC Infect Dis 2020;20:370.
12. Leys D, Manouvrier J, Dupard T, et al. Right superior vena cava draining into the left atrium with left superior vena cava draining into the right atrium. Brit Med J (Clin Res Ed) 1986;293:855.
[13] Hong SN, Nayar A, Srichai MB, et al. A case of an anomalous superior vena cava with anomalous pulmonary veins—when two wrongs do not make a right. Echocardiography 2011;28:E39–41.
[14] Issa E, Salloum T, Tokajian S. From normal flora to brain abscesses: a review of Streptococcus intermedius. Front Microbiol 2020;11:826.
[15] Clark C, MacDonald L. Right-sided superior vena cava draining into the left atrium in a patient with persistent left-sided superior vena cava emptying into the right atrium diagnosed by echocardiography. Proc (Bayl Univ Med Cent) 2015;28:363–6.
[16] Wood PH. Diseases of the Heart and Circulation. 2nd ed. Philadelphia, PA: Lippincott; 1956:457.
[17] Karavassilis ME, Hajj-Coll M, Keenan NG. Multiple thromboembolic events associated with bilateral superior vena cava and anomalous drainage into the left atrium. BMJ Case Rep 2021;14: e237401.