 doi: 10.2176/jns-nmc.2022-0066

NMC Case Report Journal 9, 209-212, 2022

Spinal Epidural Hematoma Due to Venous Congestion Caused by Nutcracker Syndrome

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Abstract

The causes of spinal epidural hematoma (SEH) have been attributed to coagulopathy, trauma, vascular anomalies, and so forth. The incidence of vascular anomalies shown by digital subtraction angiography has been reported to be 15%, and most cases have been reported to be spinal epidural arteriovenous fistulae. SEH has rarely been caused by venous congestion. We report a case of SEH in a 78-year-old male who presented to our emergency department with sudden-onset back pain, followed by complete paraplegia with bladder and rectal disturbance. Magnetic resonance imaging revealed a dorsally placed extradural hematoma extending from T10 to L1. An urgent laminectomy from T11 to L2 was performed. Computed tomography angiography (CTA) performed 1 week after the operation showed compression of the left renal vein between the aorta and superior mesenteric artery with dilation of the surrounding veins, including the spinal epidural venous plexus, at the same level as the hematoma. This was diagnosed as Nutcracker syndrome (NCS), which was consistent as a cause of SEH. The patient's symptoms gradually improved, and after 6 months, he regained normal strength in his lower extremities, but bladder and rectal disturbance remained and required intermittent self-catheterization. We chose conservative treatment for NCS, and SEH did not recur until the patient died of a cause unrelated to SEH or NCS. SEH could occur secondary to venous congestion including NCS. We emphasize the importance of investigating venous return to evaluate the etiology of SEH, which can be clearly visualized using CTA.

Keywords: spinal epidural hematoma, Nutcracker syndrome, CTA

Introduction

Spinal epidural hematoma (SEH) is a rare condition defined as an accumulation of blood in the epidural space leading to spinal cord compression, of which the pathophysiology often remains unclear. However, some SEHs are related to vascular anomalies, such as arteriovenous fistulae, vertebral hemangiomas, hypertension, and abnormal bleeding tendencies.[1-3] Vascular anomalies shown by digital subtraction angiography (DSA) have been reported at 15%, and most of them are spinal epidural arteriovenous fistulae.[4] Here we describe a case of SEH at the T10-L1 level causing severe paraplegia with bladder and rectal disturbance. The radiological findings showed venous congestion including the spinal epidural venous plexus at the T11-L2 level caused by Nutcracker syndrome (NCS), which is consistent with the cause of bleeding.

Case Report

A 78-year-old male presented with sudden-onset back pain, followed by complete paraplegia with bladder and rectal disturbance. He denied straining or coughing at the time of the onset. The patient looked thin, but he said that his weight had almost remained unchanged since he was 20 years old. Clinical examination indicated that he was underweight. He was 172 centimeters tall and weighed 53 kilograms, and his body mass index was 17.9, which

Received March 7, 2022; Accepted May 9, 2022

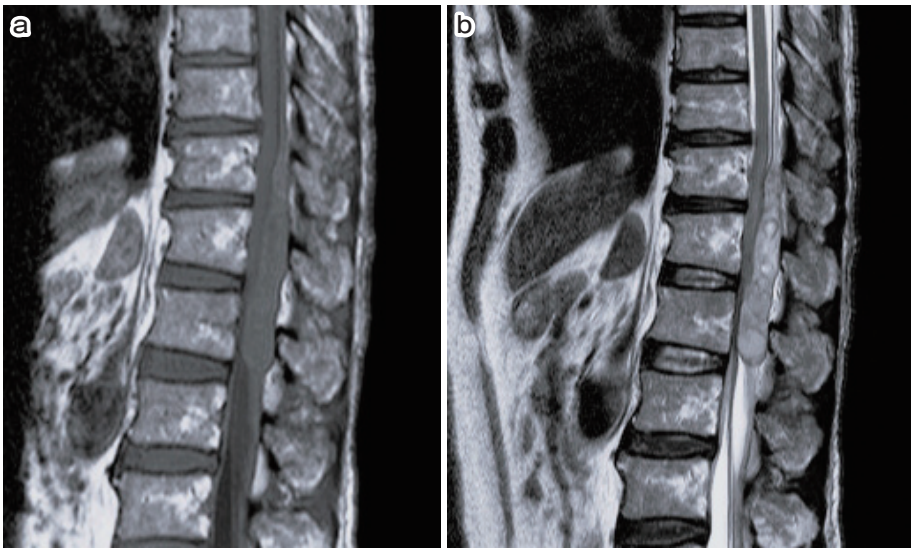

Fig. 1 a, b: Magnetic resonance imaging demonstrates a well-circumscribed posterior epidural mass at the T10-L2 level measuring 110 mm × 18 mm × 12 mm compressing the adjacent segment of the spinal cord. It elicits isointensity to the spinal cord on the T1-weighted image (Fig. 1a) and heterogeneous high intensity on the T2-weighted image, which is consistent with spinal epidural hematoma in the acute phase.

was classified as underweight.[5] Neurological examination revealed complete paraplegia, loss of tendon reflexes in both lower extremities, and impaired external sphincter reflexes. Preoperative magnetic resonance imaging (MRI) revealed a dorsally placed extradural mass extending from T10 to L1 (Fig. 1), diagnosed as SEH. An urgent laminectomy from T11 to L2 was performed. The hematoma was evacuated, except that which surrounded the left side of the epidural venous plexus, which was solid and was suspected as the bleeding source. The postoperative course was uneventful. Computed tomography angiography (CTA) performed 1 week after the operation showed left renal vein compression. The narrowed space between the superior mesenteric artery (SMA) and aorta and consequent dilation of the surrounding veins including the spinal epidural venous plexus from T11 to L2 (Fig. 2) was diagnosed as NCS, which was consistent as a cause of SEH. Urinalysis showed uric blood, but the patient's blood test showed a normal level of hemoglobin. Spinal DSA was performed with no significant findings. Considering that the patient's hematuria was mild, the radiologist decided to treat the NCS conservatively. We educated the patient to gain weight in order to increase visceral fat in expectation of increasing the angle between the SMA and aorta. His symptoms gradually improved, and after 6 months, he regained normal strength in his legs, but bladder and rectal disturbance remained and required intermittent self-catheterization. Repeated follow-up CTA showed that the congestion of veins including the epidural venous plexus remained unchanged. His body weight had remained unchanged until suffering from malignant lymphoma, and he gradually lost weight with exacerbation of the illness, but SEH did not recur until he died of malignant lymphoma 6 years after the operation. His malignant lymphoma was diagnosed 6 months before his death and was unrelated to NCS.

Discussion

The patient in this report experienced sudden-onset back pain followed by paraplegia. MRI showed SEH at the T10-L1 level. CTA after urgent laminectomy showed venous congestion caused by NCS, and the congestion extended to the spinal epidural venous plexus, which is consistent with a cause of SEH.

NCS is a rare clinical entity characterized by a group of clinical manifestations caused by compression of the left renal vein with a narrowing space between the SMA and aorta. The symptoms of NCS reported previously are macro and microhematuria, proteinuria, and flank pain. There are no clinical criteria for diagnosis; therefore, the diagnosis is made after excluding other causes and confirmed by Doppler ultrasonography or CTA.[6] In this case, venous hypertension caused by NCS increased blood flow into the collateral hemiazygos vein and superior lumbar vein, leading to enlargement of the epidural venous plexus. CTA is useful to diagnose the accurate relation between the SMA and left renal vein as well as the congested veins. DSA is the gold standard for diagnosing spinal epidural arteriovenous fistulae, which is the main cause of SEH due to vascular malformation. However, less information was obtained from DSA compared with CTA in this case.

The venous congestion caused by NCS is thought to be the cause of SEH here. There have been no reports stating

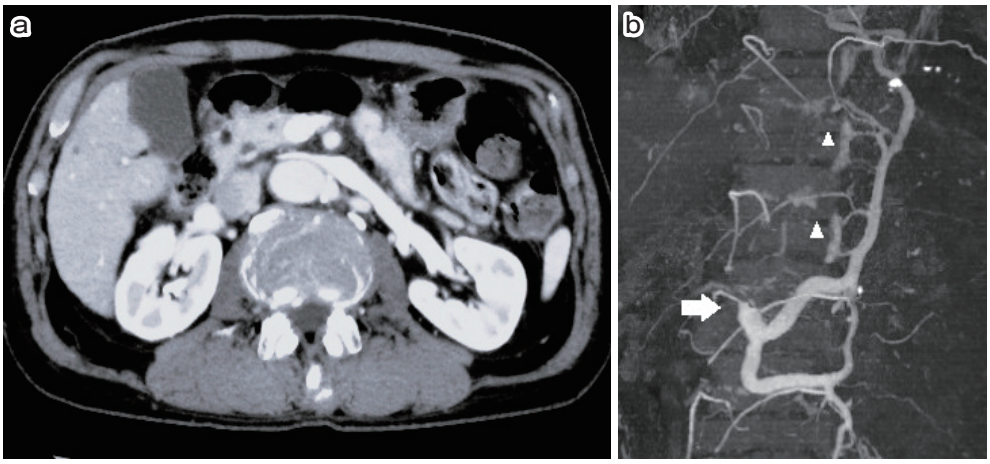

Fig. 2 a, b: Computed tomography angiography showing venous outflow obstruction of the left renal vein with a narrowed space between the aorta and superior mesencephalic artery (Fig. 2a), resulting in collateralization of venous circulation involving the hemiazygos vein, superior lumbar vein, lumbar veins, radicular veins, and epidural venous plexus (white arrowheads). The left renal vein is cut at the narrowest part of the stenosis (white arrow).

that SEH is related to NCS, but Ishida A. et al. have reported SEH caused by left internal jugular vein thrombotic occlusion and dilation of the surrounding venous plexus.[7] The pathogenesis of their case was thought to be the venous hyper pressure due to venous flow congestion, which we believe is similar to our case. When the cause of SEH is investigated, the venous flow should be also assessed.

As NCS is rare, there have been no established treatment guidelines as of Feb 2022, but the principle is conservative treatment. The main anatomical feature of NCS is the compression and obstruction of the left renal vein between the SMA and aorta. Narrowing of the angle between those arteries is related to body fat, so very thin patients may benefit from weight gain. Most patients can be treated conservatively; however, more invasive treatment options, such as an open surgical procedure or endovascular stenting, may be necessary for patients who have severe symptoms, such as significant hematuria, persistent orthostatic proteinuria, and severe flank or pelvic pain.[8,9] In this case, NCS was diagnosed after the occurrence of SEH. We decided to treat the patient conservatively for two reasons. First, the only symptom caused by NCS was mild microhematuria. Second, the level of the spine where the epidural veins were enlarged was laminectomized, so the influence was thought to be limited if SEH recurred. Consequently, SEH did not recur until the patient died 6 years after the first bleeding episode. We believe that invasive treatment for NCS is not necessarily needed for preventing SEH recurrence when the spinal level at which the epidural veins are congested is laminectomized.

Conclusion

SEH could occur secondary to venous congestion including NCS. To evaluate the etiology of SEH, venous return should be investigated, which can be clearly visualized using CTA.

Abbreviations

Spinal epidural hematoma: SEH
Digital subtraction angiography: DSA
Computed tomography angiography: CTA
Nutcracker syndrome: NCS
Superior mesenteric artery: SMA

Informed Consent

Informed consent was obtained from the family of the patient described in this report.

Conflicts of Interest Disclosure

All the authors have no conflicts of interest.

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