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Spinal arteriovenous malformation associated with congenital dermal sinus: a case report

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Abstract

Background A spinal arteriovenous malformation is a rare condition caused by abnormal anastomoses in the spinal cord, in which blood flows from arteries to veins without capillary involvement. A congenital dermal sinus is a form of spinal dysraphism that results from aberrant development of ectodermal, mesodermal, and neuroectodermal tissues. We herein present the first reported case of an arteriovenous malformation with arteriovenous anastomosis adjacent to a congenital dermal sinus.

Case presentation A 75-year-old Japanese man without underlying disease or trauma presented to a local hospital with a 4-month history of progressive bilateral lower limb paresthesia. Magnetic resonance imaging revealed the tip of the conus medullaris to be low at the L3/4 level, with hypersignal intensity of T2-weighted images in the spinal cord extending from the conus medullaris to T11. We diagnosed the lesion as a spinal arteriovenous malformation with congenital dermal sinus and surgically resected both structures. Postoperative radiological findings showed the disappearance of abnormal vascular shadows. The patient's balance was stable, and he was able to walk independently.

Conclusion We encountered a spinal arteriovenous malformation with abnormal anastomosis of the arteriovenous vein at a different site than previously reported. In the management of spinal arteriovenous malformations, confirmation of the patient's cutaneous findings, appropriate history taking, and careful clinical and imaging examination may enable successful treatment.

Keywords Arteriovenous malformation, Spine, Congenital dermal sinus, Surgical procedure, Case report

Background

Spinal arteriovenous malformations (AVMs) are rare lesions caused by abnormal anastomoses in the spinal cord, whereby blood flows directly from arteries to veins without capillary involvement [1]. The majority of spinal AVMs occur at birth and are thought to progress with age, although the precise mechanism of lesion development is not understood [2]. Meanwhile, a congenital dermal sinus (CDS) is a form of spinal dysraphism that is caused by aberrant development of ectodermal, mesodermal, and neuroectodermal tissues [3].

To the best of our knowledge, we herein report the first case of an AVM with arteriovenous anastomosis adjacent to a CDS.

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Case presentation

A 75-year-old Japanese man without underlying disease or trauma presented to a local hospital with the chief complaint of a 4-month history of progressive bilateral lower limb paresthesia. The patient had no medical, family, or psychosocial history, nor had he undergone any relevant past interventions. He had been diagnosed as having a spinal cord tumor at the same hospital and was scheduled for further investigation and treatment at our department. However, he required emergency hospitalization owing to the inability to maintain a standing position for more than 3 minutes during the week before his appointment. He also reported no longer being able to ascend or descend stairs without the use of a handrail.

Physical examination on admission revealed a skin ostium in the medial lumbar region (Fig. 1a, b). Radiographs of the lumbar spine showed spondylosis without other abnormalities (Fig. 1c, d). A continuous ductal structure was observed from the skin ostium in the medial lumbar region. It appeared to run deeply toward the L3/4 interspinous process, terminating at the dural surface and entering the thecal sac (Fig. 1e, f).

The patient rated his low back pain as 30 mm on a visual analogue scale. Neurological evaluation disclosed spastic gait, bilateral lower limb paresthesia, hyperreflexia, slight difficulty urinating, and the presence of Romberg's sign. Manual motor testing results of the lower extremities, including the iliopsoas, quadriceps, hamstring, tibialis anterior, triceps surae, extensor digitorum longus, and flexor digitorum longus muscles, were all scored as 5. However, his movements were slow and spastic, and he had difficulty maintaining elevation of his lower extremities for more than 1 minute. Magnetic resonance imaging (MRI) of the lumbosacral spine showed a low conus medullaris tip at the L3/4 level, with hypersignal intensity of T2-weighted images in the spinal cord extending from the conus medullaris to T11.

Sagittal T2-weighted imaging of the lumbosacral spine revealed tortuous intradural flow voids along surfaces of the spinal cord from the lower thoracic to lumbar spine (Fig. 1g, h). Contrast-enhanced computed tomography (CT) showed enlarged tortuous veins from L4 to T12, with filling in the early arterial phase (Fig. 1i, j). A mixture of thin and thick contrasted veins at L3/4

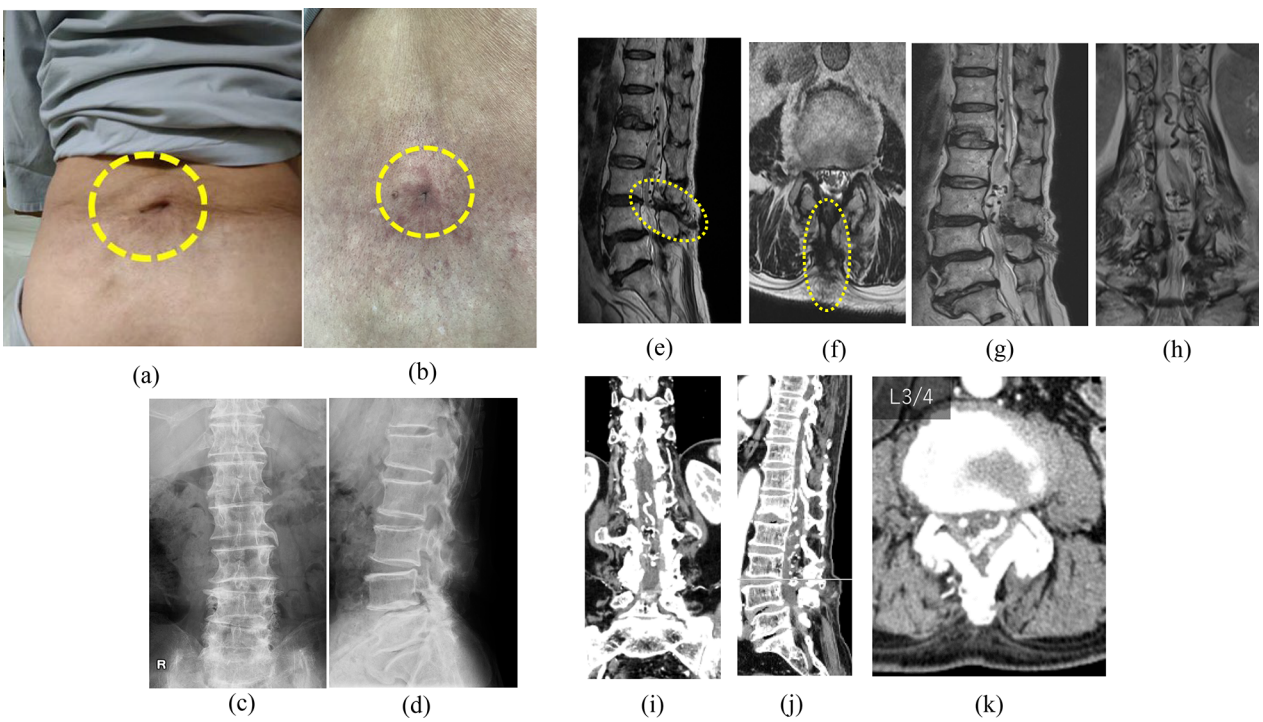

Fig. 1 Preoperative photograph of the patient's back and radiological findings of the lumbosacral spine. (**a**, **b**) Patient exhibited a skin ostium (dotted circle) in the medial lumbar region. (**c**, **d**) Radiographs of the lumbar spine showing spondylosis without other abnormalities. (**e**, **f**) A continuous ductal structure (dotted ovals) observed from a skin ostium appearing to run deeply toward the L3/4 interspinous process, terminating at the dural surface and entering the thecal sac. (**g**, **h**) Tortuous intradural flow voids were apparent along surfaces of the spinal cord from the lower thoracic to lumbar spine. (**i**, **j**) Enhanced computed tomography showing enlarged tortuous veins from L4 to T12, with filling in the early arterial phase. (**k**) We observed a mixture of thin and thick contrasted veins at the L3/4 level

suggested an arteriovenous anastomosis near the same level (Fig. 1k).

Following the diagnosis of a spinal AVM with cutaneous sinus, we performed AVM and CDS resection. We consulted the neurosurgeon about embolization of the feeder vessel prior to surgery, but since it was a semi-emergency procedure under time constraints, the neurosurgeon recommended prioritizing surgery over embolization. After the initiation of general anesthesia, the patient was placed in a supine position. We delineated a margin around the dermal sinus, and skin incision lines were marked in the cephalic and caudal directions of the dermal sinus. Taking care not to cut into the dermal sinus out of concern for possible spinal cord infection, L3/4 laminectomy was performed as well. When the dura mater was exposed, the dermal sinus was seen to be continuous with the dura mater. Accordingly, we incised the dura mater above and below the sinus. A fatty mass was found cephalad to the dermal sinus that was continuous with the sinus base by a cord-like substance (Fig. 2a, b). A dilated abnormal vessel caudal to the dermal sinus was judged as the inflow vessel for the spinal AVM. This vessel was temporarily blocked, and after the confirmation of no abnormalities by intraoperative neuromonitoring, it was severed (Fig. 2c). The dermal sinus was excised at the base of the cord-like structure and removed together with the abnormal blood vessels (Fig. 2d). The procedure was time consuming owing to the difficulty in finding the AVM and detaching the strong adhesions around it. Fluorescence contrast-enhanced examination with indocyanine green confirmed the disappearance of abnormal vessels (Fig. 2e). As the dura mater was largely deficient after removal of the dermal sinus, the dural defect was patched using the fascia of the back muscles. Ultimately, his operation time was 468 minutes, and blood loss was 280 mL. We encountered no perioperative complications, including surgical site infection or cerebrospinal fluid leakage. Histopathological examination of the excised lesion suggested a CDS with an adjacent arteriovenous connection (Fig. 3). No neural tissue was identified in the lesion. Postoperative contrast-enhanced CT and MRI images at 2.5 months demonstrated that the abnormal vascular shadows had disappeared without recurrence along with a reduction of the intramedullary T2-weighted high-signal area (Fig. 4a, b). Furthermore, the patient's Japanese Orthopaedic Association (JOA) score improved from 8 points preoperatively to 10.5 (maximum: 11) points postoperatively, his balance was stable, and he could walk independently at 7 months after surgery.

Discussion and conclusion

Spinal vascular malformations are a rare but important group of lesions that, if left undiagnosed and untreated, can lead to serious complications. The reported phenomena of mass effect, stolen blood effect, and venous stasis caused by enlarged abnormal blood vessels often produce a variety of nonspecific neurological symptoms; without

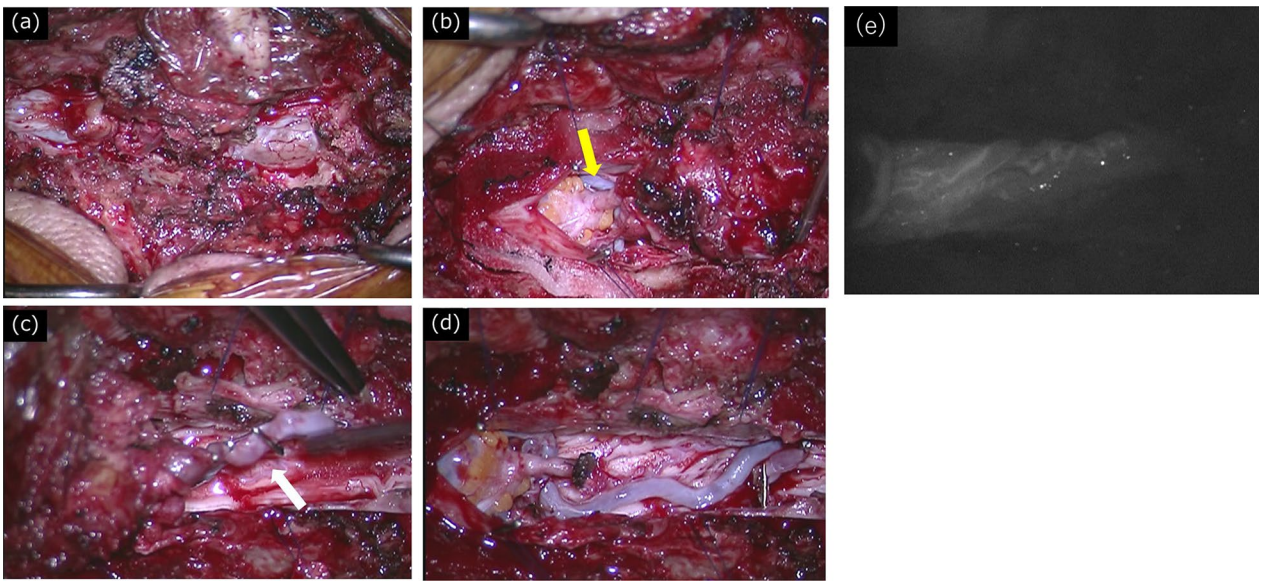

Fig. 2 Surgical site photographs. (**a**) After L3/4 laminectomy, the cutaneous sinus was continuous with the dura mater. (**b**) A fatty mass was present on the cephalic side of the dermal sinus. It was contiguous with the base of the sinus by a cord-like substance (yellow arrow). (**c**) Abnormal dilated vessels caudal to the dermal sinus were observed. (**d**) Intradural photograph after colectomy of the dermal sinus and abnormal vessels. (**e**) Fluorescence contrast-enhanced examination with indocyanine green in the dura after resection of the abnormal vessels

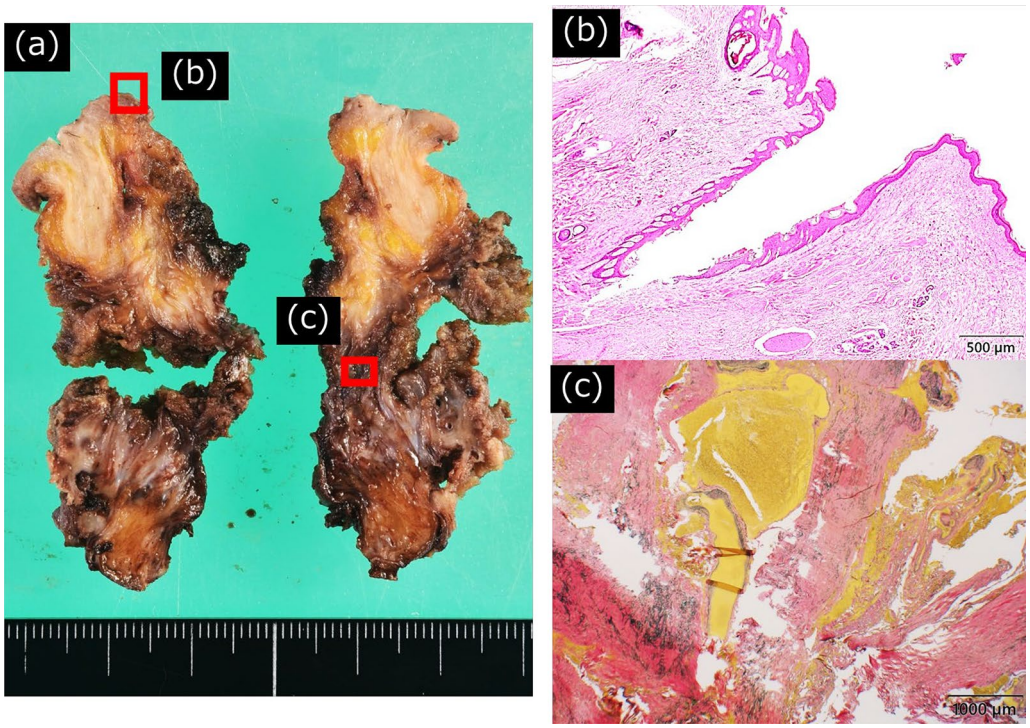

Fig. 3 Histopathological findings. (**a**) Extracted lesions cut in the sagittal plane. Panels show the areas observed under a microscope. (**b**) The fistula area showed a luminal structure covered by multilayered squamous epithelium from the epidermis to deeper layers. Abnormal anastomotic findings of blood vessels in the tissue adjacent to the dermal sinus were evident

timely and appropriate treatment, progressive spinal cord symptoms and myelopathy may occur [4].

A CDS is a form of spinal dysplasia associated with disjunction defects when the closed neural tube separates from the overlying cutaneous ectoderm during primary neural tube formation [5]. It is characterized by a tract lined with stratified squamous epithelium extending from the subcutaneous tissue to the underlying thecal sac or neural tube [6]. The incidence of this malformation is 1 case per 2500 live births [7]. The CDS passes through the dorsal dysplastic elements of the spinal column and dura mater as a thin fiber band connecting the skin (dermal ectoderm) to the central nervous system (neuroectoderm). It usually runs through the dystrophic dorsal elements of the spinal column and through the dura and attaches to the spinal cord or another intradural neural element [8]. This fibrous band should be completely removed as it can compress surrounding nerve structures and cause infection associated with abnormal dermal and epidermal tissue [8, 9].

Spinal dysplasia associated with spinal AVMs has been reported, including lipomyeloceles, lip myelomeningoceles, spinal lipomas, and filum terminal lipomas [10]. Spinal AVMs associated with spinal lipomas may have an arteriovenous anastomosis within the lipoma, and so radical resection of the lipoma is recommended along with the treatment of shunt blood flow [11]. However, there are several subtypes of spinal lipomas, some also being malformations of the spinal cone, that can be difficult to recognize intraoperatively owing to interspersion with lipomas [12].

It is generally accepted that spinal adipose species and spinal AVMs are congenital, with both originating from mesenchymal tissue and showing no regression during the fetal period. Several reports have suggested a causal relationship between the lesion types, as pressure from intramedullary lipoma tissue may produce abnormal local angiogenesis and spinal AVMs [11]. The present case exhibited a spinal AVM with a spinal lipoma. Unlike previous reports, the arteriovenous anastomosis was adjacent to the CDS and separated from the lipoma, making it difficult to determine the extent of intraoperative resection. In this patient, we considered the possibility that AVM development may have been accelerated by irritation from extracorporeal traffic through the CDS rather than by pressure from the lipoma.

Good short-term results were achieved by resecting only the CDS and abnormal vessels while preserving the lipoma with reference to intraoperative neuromonitoring and indocyanine green fluorescence contrast results.

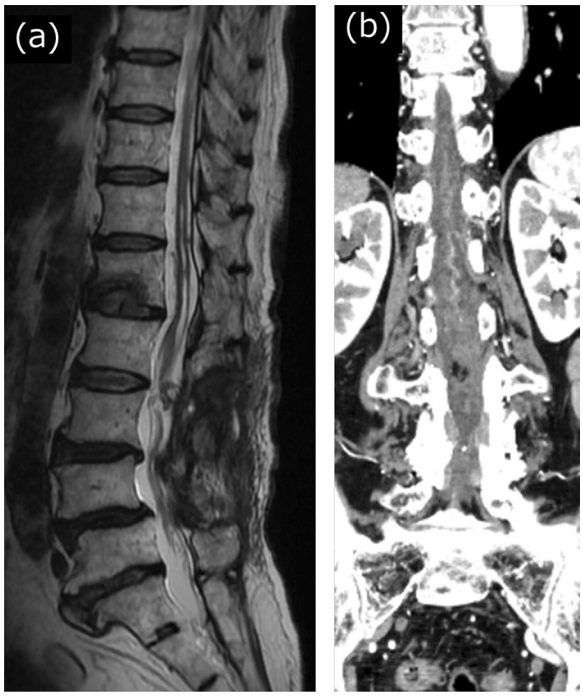

Fig. 4 Postoperative radiological findings. Postoperative (**a**) magnetic resonance imaging and (**b**) contrast-enhanced computed tomography at 2.5 months showed that the abnormal vascular shadows had disappeared, and the intramedullary T2-weighted high-signal area was reduced

However, as the presence of lipomas may lead to recurrence in the long term, careful observation is necessary.

We successfully treated a case of spinal AVM with abnormal anastomosis of the arteriovenous vein at a different site than reported to date. In the management of spinal AVMs, confirmation of the patient's cutaneous findings, appropriate history taking, and careful clinical and imaging examination may enable effective treatment.

Abbreviations

AVM	Arteriovenous malformation
CDS	Congenital dermal sinus
MRI	Magnetic resonance imaging
CT	Computed tomography

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Not applicable.

Author contributions
YK wrote the initial draft of this manuscript. MU, SI, YM, HO, TH, DK, SS, KS, TM, MD, and JT assisted with drafting the manuscript and data collection. MU was responsible for the oversight of the report and editing the manuscript. All authors read and approved the final manuscript.

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Availability of data and materials
The data that support the findings of this study are available from the corresponding author upon reasonable request.

Declarations

Ethics approval and consent to participate
This is a case report. The institutional ethics review board of the authors' facility has confirmed that no ethical approval is required. Consent was obtained from the patient for participation in this study.

Consent for publication
Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing interests
The authors declare no competing interests.

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