Spinal cord imaging in spastic paraparesis: are we cutting it thin enough?

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Introduction: Spinal dural arteriovenous fistula (SDAVF) is a rare condition, but accounts for great disability in those affected. Despite improvements in spinal imaging, SDAVF diagnosis is often difficult or masked by more common entities.

Case report: A 42-year-old woman, with a history of ileal resection 4 years prior, presented with a 6-month course of progressive walking difficulty, denying any sensory or bladder complaints. Physical examination revealed spastic paraparesis, brisk deep-tendon reflexes in the lower limbs, bilateral Babinski sign and bilateral foot drop. Blood chemistry was normal except for low vitamin B12 levels. Electromyogram was compatible with sensorimotor axonal polyneuropathy and cervical, thoracic and lumbar spinal MRI revealed no signal change. Parenteral cyanocobalamin supplementation was initiated and the patient was discharged with a diagnosis of polyneuropathy and probable subacute combined degeneration. In the following months, the patient’s gait improved, but the spastic paraparesis was unchanged. Repeat spinal cord MRI revealed no signal change. Given the lack of improvement, a SDAVF was suspected. Spinal MR-angiography and volumetric T1 acquisitions after contrast were obtained using thin-slice and multiplanar reconstruction, revealing dilated vessels in the spinal periphery from T11 to L2 levels. Digital subtraction angiography confirmed the diagnosis and the patient was referred for surgical occlusion of the fistula. After surgical intervention, there was a clinical improvement.

Conclusion: We describe a case of a SDAVF with superimposed vitamin B12 deficiency, whose diagnosis required extra-thin cuts of MR-angiography. SDAVFs are potentially reversible causes of myelopathy, thus emphasizing the importance of their early identification.

Keywords: Magnetic Resonance Angiography, Subacute Combined Degeneration, Dural Arteriovenous Fistula, Spastic Paraparesis

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Background

Spinal dural arteriovenous fistula (SDAVF) is a rare condition, but accounts for great disability in those affected. SDAVF are more common in men and typically present in the fifth and sixth decades of life [1]. The patients usually present with lower extremity spastic weakness, sensory symptoms, bowel/bladder disturbances and back pain [1, 2]. Despite improvements in spinal imaging, SDAVF diagnosis is often difficult or masked by more common entities. Magnetic resonance imaging (MRI) typically shows dilated perimedullary veins, spinal cord swelling, and central high signal intensities on T2-weighted images [3], but SDAVF presenting without spinal cord signal abnormalities have also been described [4].

Case report

We present a case of a 42-year-old woman, with a history of ileal resection for mesenteric cysts 4 years prior and epilepsy secondary to a cortical dysplasia. She drank around 25 cc. of wine with her meals and otherwise had unremarkable past history. Both her parents, non-consanguineous, had multiple vascular risk factors. No other relevant family history was reported.

She presented at our outpatient clinic complaining of progressive walking difficulties and symmetric weakness on both legs, which had developed in the previous 6 months. Other complaints, namely sensory or bladder/bowel disturbances, were denied. She reported significant weight loss, which she did not quantify. History of spinal trauma or back pain was absent. Physical examination revealed a symmetric, predominantly distal, spastic paraparesis (Medical Research Council scale [MRC], proximal 4+/5 and distal 3/5), brisk deep tendon reflexes in the lower limbs, bilateral Babinski sign and bilateral foot drop with steppage gait; there were no sensitive deficits (pinprick, light touch and vibration or positional) upon examination. The remainder of the physical examination was unremarkable.

The patient was admitted to the Neurology ward for further investigation. Blood chemistry (including a complete blood count, hepatic and renal function, electrolytes, folate, vitamin B12, and serologic tests for Borrelia spp, syphilis, HIV and hepatitis B and C viruses) was normal, except for borderline vitamin B12 levels (194 pg/mL, for a normal range of 187-883 pg/mL). The electromyogram was compatible with mild sensorimotor axonal polyneuropathy. Cervical, thoracic and lumbar spinal MRI revealed no signal change.

Despite the atypical findings for subacute combined degeneration and borderline vitamin B12 levels, the patient was discharged with the diagnosis of probable subacute combined degeneration and polyneuropathy secondary to vitamin B12 deficiency (in the context of the ileal resection). Intramuscular cyanocobalamin supplementation was prescribed.

Initially, there was a slight clinical improvement after cyanocobalamin supplementation, however, after 12 months, on re-evaluation, the patient’s walking difficulty had worsened (needing bilateral support for walking) and interfered markedly in daily-living activities. She denied any de novo sensitive or sphincter disturbances. On neurological examination, the patient’s steppage gait was improved, but the spastic paraparesis had aggravated. Marked muscular atrophy on both legs was noted. There were no sensitive deficits.

The patient was re-admitted, and additional work-up was ordered. Blood chemistry was normal; repeat cervical, thoracic and lumbar spinal MRI revealed no signal change.

Given the functional decline, despite adequate cyanocobalamin supplementation, further causes were pursued, and a SDAVF was suspected. Spinal MR-angiography and volumetric T1 acquisitions after contrast were obtained using thin slices and multiplanar reconstruction and revealed dilated vessels in the spinal cord periphery from T11 to L2 levels (Figure 1B). Digital subtraction angiography confirmed the diagnosis of SDAVF (Figure 1C&D). After consulting with Neuroradiology, the fistula was not considered amenable to endovascular treatment. Therefore, the patient was referred to the Neurosurgery department for surgical occlusion of the fistula, which was uneventful.

On a follow-up consultation, four months after surgical intervention, the symptoms had improved and the patient was able to walk unaided. Physical examination revealed markedly improved spasticity and weakness (MRC scale, proximal 4+/5 and distal 4-/5), maintaining brisk deep tendon reflexes in the lower limbs and bilateral Babinski sign.

Figure 1. Imaging study (23 months after symptom onset). Spinal MRI – (A-B): No signal change (A - sagittal T2). Vessel enlargement in thin section spinal MRI (B – sagittal T1-gadolinium, 0.9 mm slices). Spinal Digital Subtraction Angiography (C-D): Dural fistula, with its arterial afferent from the left L1 level segmentar artery (#), abnormal radicular vein drainage (not shown) and tortuous perimedullary venous vessel enlargement (*).
Discussion

We describe a case of a SDAVF with superimposed vitamin B12 deficiency, in which initial spinal MRI had unremarkable findings. This case highlights the importance of a high clinical suspicion since SDAVF are potentially reversible causes of myelopathy if identified early [1] and normal MRI does not exclude the diagnosis [4]. Thin-section volumetric post-contrast MRI and MR-angiography studies can reveal subtle details, not seen in routine protocols. In our case, due to atypical findings for subacute combined degeneration, including a normal MRI, further causes were pursued, emphasizing the importance of clinical judgment.

Abbreviations

SDAVF: Spinal dural arteriovenous fistula; MRI: Magnetic resonance imaging

Competing interests

The authors declare no conflict of interest.

References


