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Case Report

Spinal Dural Arteriovenous Fistula Presenting With a Radiological "H-Sign"

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Abstract

Background: Spinal Dural Arteriovenous Fistula (SDAVF) is a vascular abnormality of the spinal cord. Its clinical features include lower limb weakness, sphincter dysfunction and sensory deficits. Since these symptoms are seen in other more common causes of myelopathy, the diagnosis is often delayed. Several MRI findings suggestive of SDAVF have been described previously, but none of them was proven specific for the diagnosis. **Methods:** We describe a patient who presented with symptoms of progressive myelopathy. A written consent-to-disclose, was provided by the patient. **Results:** Workup included spinal MRI with findings suggestive of myelopathy and the presence of an"H-Sign", indicative of gray matter involvement. Treatment with intravenous steroids was in accordance with working diagnosis of myelitis. The patient's clinical deterioration following steroids, led to reevaluation of the diagnosis and the performance of spinal angiography, in which a SDAVF was detected. **Discussion:** The course of myelopathy in our patient was rather chronic and fluctuating, leading to long standing venous congestion which could possibly explain the involvement of the deep gray matter and the clinical deterioration following steroid-treatment. Our case shows that the finding of an"H sign" in spinal MRI should not dissuade clinicians from completing the relevant workup for SDAVF, especially in cases of steroid-non-responsive myelopathy and atypical clinical features.

Introduction

Spinal Dural Arteriovenous Fistula (SDAVF) is a common vascular abnormality of the spinal cord [1]. Clinical features include lower limb weakness, sphincter abnormalities and sensory deficits. The diagnosis is often delayed because clinical symptoms are not specific and usually similar to those of other, more common causes of myelopathy [1]. The strongest clinical clue is a dramatic worsening of symptoms with activities (or medications) that elevate the venous pressure, such as exertion, Valsalva maneuver, lumbar puncture and treatment with corticosteroids that cause venous congestion [2].

.Several common findings on MRI were described in the past, but none of them is specific [3]. The T2-hyperintense signal is often longitudinally extensive, predominantly involving the lower thoracic region and conus and occasionally extending along multiple segments and affecting white and gray matter within them [2]. Engorged perimedullary veins and subarachnoid posterior flow voids are also typical radiological findings. Variable gadolinium-enhancement patterns such as missing piece sign (a defect in contrast filling) or hazy enhancement with central canal prominence may be helpful indicators for SDAVF [2-4].

We describe here a patient with symptoms of progressive myelopathy caused by spinal dural arteriovenous fistula, associated with unusual findings on the spinal MRI.

Case Report

A 49 year old male patient, with no previous history of neurological diseases presented with progressive weakness of his lower limbs during the previous two months. He additionally complained of urinary retention and intermittent numbness in the distal parts of the lower limbs. He denied other neurological symptoms, fever, weight loss, night sweats or rheumatologic stigmata. Neurological examination demonstrated bilateral spastic paraparesis involving the proximal and distal muscles of the lower limbs (rated 4 out of 5), accompanied by exaggerated patellar and Achilles reflexes and a pathological plantar response, hypoesthesia of the lower limbs and a sensory level at T4. A long hyperintense T2 signal extending from T4 till cauda equina, without contrast enhancement, was detected on spinal MRI. Axial slices showed T2 hyperintensity with prominent involvement of the gray matter of the spinal cord, leading to a characteristic "H sign" [5]. The initial differential diagnosis based on the clinical presentation and the MRI findings included mainly inflammatory or paraneoplastic myelopathies. The putative immune-mediated pathogenesis was supported by the finding of high protein levels in the CSF (97 mg/dl) with normal glucose concentration (51 mg/dl), and 4 lymphocytes per visual field.

Serological tests for HIV, mycoplasma pneumonia, VZV, HSV and enterovirus, were all negative. Tests for paraneoplastic autoantibodies, anti AQP4 and anti MOG antibodies, were also negative. Whole-body CT was within normal limits.

Since the initial working diagnosis was inflammatory myelopathy, a treatment trial with IV Methylprednisolone (IVMP) was initiated. However, the following day, the patient experienced an acute deterioration in weakness of the lower limbs (graded 2 out of5 at this point), advocating against an inflammatory cause of the myelopathy. The acute clinical exacerbation following IVMP raised the suspicion of SDAVF and therefore, a diagnostic angiography was performed which demonstrated an arteriovenous fistula in T4-5 with venous congestion. On the next day, embolization of the fistula was performed and a substantial and fast clinical improvement was observed; the paraparesis improved from 2/5 to 4/5 on his examination. Sensory modalities to touch and temperature also greatly improved. After discharge he continued treatment in our rehabilitation center with improvement in the strength of the antigravity muscles. Three months later, a repeated MRI of thoracic and lumbosacral parts of the spinal cord showed significant improvement in the T2-hyperintense sign (Figure 1).

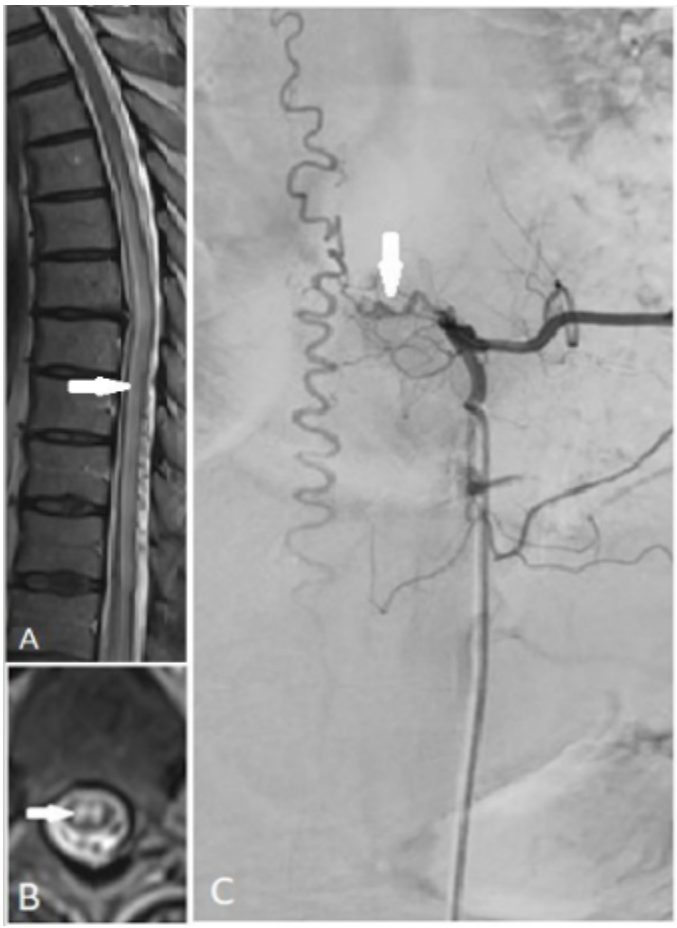

Figure 1: A) MRI T2 of thoracic spine showing hyperintense signal of longitudinal extensive myelopathy. B) An axial MRI demonstrating T2-hyperintensinity without contrast enhancement. C) Diagnostic angiography confirming location of dural arteriovenous fistula at T4 level.

Discussion

The presented patient was admitted to the Neurology department of our hospital with a picture of progressive and fluctuating myelopathy, and a suspicion of immune mediated pathogenesis (indicated by the presence of a long T2 lesion in the spinal MRI and high CSF protein), but non-responsive to steroid treatment. The final diagnosis was myelopathy due to SDAVF with an atypical imaging finding on MRI of an "H-sign" consistent with edema in the distribution of the gray matter of the spinal cord. Such radiological finding was previously described in autoimmune myelopathies, including anti-MOG-ab myelitis [5] and paraneoplastic myelopathy [5], in acute flaccid myelitis caused by enteroviruses [6] and Western Nile virus [7] and in spinal cord

infarction [8]. To our knowledge, radiological spinal "H-sign" has not been reported in the literature in association with myelopathy caused by SDAVF.

The spinal cord receives its blood supply from one Anterior Spinal Artery (ASA) (75%) and two Posterior Arteries (PSAs) (25%). ASA supplies most of the spinal cord gray matter and the anterior two-thirds of the spinal cord white matter, and PSAs supply the posterior one-third of the white matter of the cord. Unlike the arterial blood supply, there is no dominance of the dorsal or ventral venous return. The venous system of the spinal cord is arranged radially over an almost equally distributed axial territory. The venous system frequently formulates many anastomoses in and on the surface of the spinal cord. Therefore, the damage of the spinal cord due to venous congestion is usually more prominent in the deeper areas of the cord (gray matter), especially in cases of severe and longstanding venous congestion [9].

The course of myelopathy in our patient was rather chronic and fluctuating (regarding mainly the sensory symptoms), leading to longstanding venous congestion which could possibly explain the involvement of the deep gray matter that was expressed by the appearance of a radiological "H-sign" in the spinal MRI.

Worth noting, is that raised CSF protein and pleocytosis were previously reported in patients with SDAVF [10]. These findings could be explained by the development of venous hypertension from an arterialized venous drainage system, which may result in raised intravenular pressure and transudative flow of protein and cells into the extravenular space and CSF [10]. Therefore, such CSF findings (as in our patient) should not exclude the diagnosis of SDAVF.

In conclusion, our case shows that a radiological finding of a spinal "H sign" should not dissuade clinicians from completing the relevant workup for SDAVF, especially in cases with non-steroid-responsive myelopathy and other atypical clinical or radiological signs.

These authors contributed equally to the manuscript.

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