

The diagnostic challenge between Longitudinal extensive transverse myelitis (LETM) and spinal Dural Arteriovenous Fistula: Importance of Early Diagnosis.

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Backgrounds and aim: Spinal dural arteriovenous fistulas (SDAVF) are rare vascular lesions in the spine, with an estimated incidence of 5–10/million/year. The exact pathophysiology is not known, but it probably consists in an abnormal connection between a spinal radicular artery and vein disrupting the tissue perfusion with venous infarction as consequence. SDAVF are often seen in middle-aged or elderly patients presenting with non-specific symptoms of progressive myelopathy, which may lead to delayed diagnosis. Aim: We report a case of a patient with myelopathy firstly misdiagnosed as inflammatory myelitis with persistently inflammatory cerebrospinal fluid (CSF) findings.

Case report: A Man in his 60s presented with an 8- months history of paresthesias in the right lower limb, involving within few days the contralateral limb, and determining progressive legs weakness and urinary incontinence. An inflammatory pathology was initially suspected due to the cerebrospinal fluid (CSF) findings, which revealed lymphocyte pleocytosis, elevated protein levels (approximately 1 g/dL), and the presence of anti-sulfatide antibodies. The MRI showed extensive central-medullary hyperintense signal (STIR),

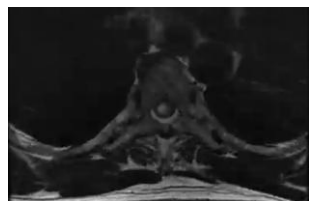


Fig. 1

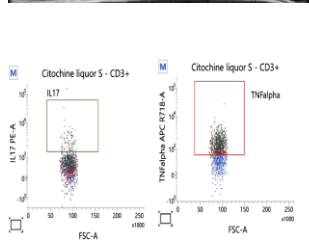


Fig. 2

involving initially the medullary conus. The patient was, indeed, treated with high dose intravenous immunoglobulins with no benefit. The patient was then admitted to our Neurology Department experiencing further worsening consisting in paraplegia and slight sensory involvement of both hands. CSF analyses confirmed CD3⁺ activation with a marked increase in inflammatory cells releasing TNF α and IFN γ and, to a lesser extent, IL-17. The patient underwent a third MRI at our hospital, which was crucial in raising the suspicion of a spinal arteriovenous fistula (after contrast, an inhomogeneous patchy enhancement was observed at the D4–D5 level, a more subtle enhancement at the conus medullaris, and a faint, inhomogeneous enhancement at the D8–D9 level). The final diagnosis was established through angiography, confirming the presence of a dural arteriovenous fistula that was successfully treated by endovascular approach.



Fig. 3

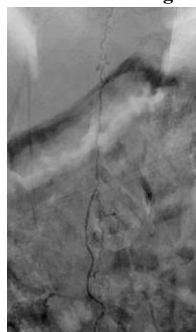


Fig. 4

Fig. 1 Axial MRI scan performed at another center on May 18, 2024: Extensive central spinal cord signal alteration, characterized by hyperintensity on long TR sequences and a mildly swollen appearance of the spinal cord

Fig. 2 CFS Analysis enlightens markers as TNF α and IFN γ .

Fig. 3 MRI (June 2024): After contrast, an inhomogeneous patchy enhancement is observed at the D4–D5 and D8–D9 level.

Fig. 4 Angiography (June 2024) confirming diagnosis of SDAVF, after which an embolization procedure was performed.

Conclusions: SDAVF may be misdiagnosed with inflammatory myelitis especially in the case of findings of neuroinflammation in the CSF. Our case indicates that the finding of an inflammatory CSF profile should not discourage clinicians from pursuing investigations for this potentially treatable condition favoring earlier diagnosis and appropriate treatment.

References

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