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Tumefactive Demyelination of the Spinal Cord

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Summary

Demyelinating lesions of the brain have been frequently reported to mimic tumors, whereas very few cases of tumefactive demyelination of the spine have been reported. We report a patient with tumefactive demyelination of the spinal cord who was clinically, by MRI, and by frozen section diagnosed with tumor and had complete surgical resection. Hence patients with spinal cord lesions suspicious of tumor should be biopsied before complete surgical resection to rule out demyelination.

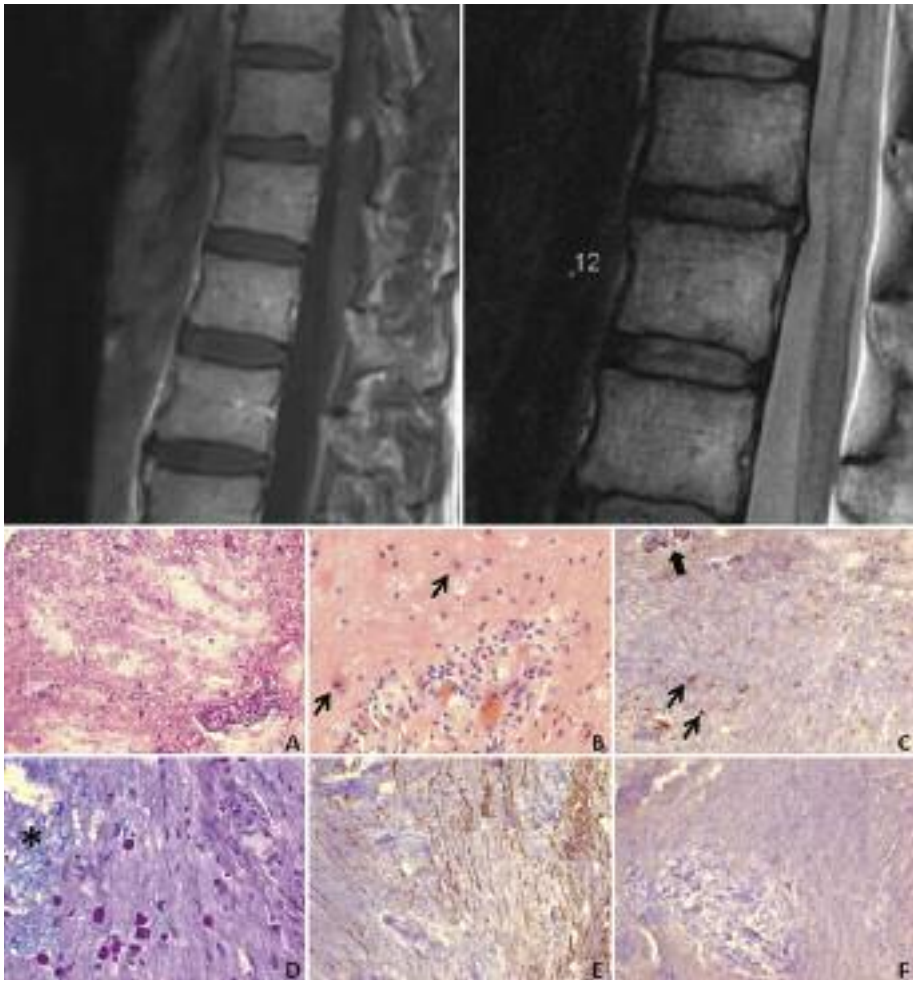
Key words: Tumefactive demyelination; Spinal Cord; Tumor; Multiple Sclerosis.

Background

Tumefactive demyelinating lesions (TDL) of the brain have been described, whereas very few cases of the spinal cord were reported. These lesions can often clinically and radiologically mimic central nervous system tumors or infections. The largest study to date on brain TDL showed a broad spectrum of radiological characteristics (location, size, mass effect, edema, and enhancement pattern) of TDL making it difficult to distinguish from neoplasm with regular MR and CT imaging modalities (Lucchinetti et al., 2008). Although perfusion weighted MRI and spectroscopy have been utilized to differentiate between TDL and tumors of the brain, they are of limited use in spinal cord disorders due to decreased tissue volume as compared to the brain. Despite the fact that there is lower frequency of positive oligoclonal bands in patients with acute monosymptomatic demyelinating syndrome (Rolak *et al*., 1996), CSF testing for oligoclonal bands is very useful in the diagnostic workup of such patients as their presence would point to an underlying demyelinating disease.

Case

We report on a 49 year-old man who presented with four months history of worsening bilateral lower extremity weakness with bowel and bladder urgency. He had no history of back pain. On neurological examination he had bilateral lower extremity spasticity, clonus, hypereflexia, and moderate weakness of proximal and distal muscle groups. Sensory examination showed a T12 sensory level and moderate decrease in sensing vibration at the ankles. Review of system was negative and routine labs were within normal limits. MRI of the spine with gadolinium revealed a T2 hyperintense signal with cord swelling involving the thoracic cord at T11 and T12 levels with enhancement along the left half of the thecal sac at T11 level, which was suggestive of a neoplastic lesion like glioma (Fig. 1). Head MRI with contrast was normal. CSF studies were not done initially due to high suspicion of malignancy based on MRI results and were declined by the patient later on. After the tissue frozen section revealed gliosis and leukomalacia and was negative for inflammation, the mass was completely resected. Later immunostaining revealed a considerable inflammatory component with gliosis and axonal loss suggestive of a subacute/chronic demyelination. On 3 month follow up after surgical resection and intravenous steroid treatment (methylprednisolone, 1gram intravenously daily for 5 days), the patient had no improvement in his neurological symptoms. After 1 year, the patient had no new neurological symptoms, neurological exam was unchanged, and repeat brain MRI with contrast remained normal. Although it might have been useful to do an MRI of the spine at 1 year follow up, it was felt unnecessary at that time as the cord lesion the patient previously had was completely resected and the patient had no



Upper part: T2 MRI of the spine revelaed a T2 hyperintense lesion with cord swelling involving the thoracic spine at T11 and T12 levels (right part). T1 MRI with guadolinium showed a focus of enhancement along the left half of the thecal sac at T11 level (left part)

Lower part: Fig. — A. Frozen section shows mildly increased cellularity due to reactive astrocytes and macrophages; B. Perivascular lymphocytes (center bottom) are also seen on permanent sections, in addition to reactive astrocytes (arrows); C. CD68 immunohistochemistry highlights macrophages within the neuropil (arrows) and around the vessels (block arrow); D. Only small areas of residual myelin is seen (asterix) by luxol fast blue/PAS/cresyl violet (LFB/PAS/CV) stain with extensive myelin loss in the rest of the tissue; E. Neurofilament immunohistochemistry reveals the relative preservation of axons (brown lines); F. No proliferative activity is identified by Ki-67 immunohistochemistry.

new neurological symptoms to suspect a new spinal cord lesion.

Discussion

Demyelinating diseases of the central nervous system is a spectrum of conditions that include multiple sclerosis, acute disseminated encephalomyelitis, neuromyelitis optica, and acute hemorrhagic leukoencephalopathy. These conditions rarely present as TDL involving the spinal cord, hence they can be mistaken on MRI by other more common entities such as tumors or infections. Since non invasive tests could not provide definitive diagnosis and frozen section results may miss the diagnosis, and resection based solely on these results may worsen the outcome, a biopsy of the lesion for tissue histology and immunostaining to provide definitive diagnosis should be performed prior to any further treatment.

Our patient is one of the very few reported cases of TDL of the spinal cord having normal brain MRI. It is also the only case in the literature of tumefactive demyelination of the lower spinal cord since these

lesions usually involve the cervical cord (Lei *et al*., 2009). In addition to a normal brain MRI, our patient's spinal lesion had significant edema on MRI which makes the diagnosis of TDL less likely on MRI basis. Moreover, the frozen section of the lesion was negative for inflammation. Although the frozen section was negative for malignancy, the high radiological suspicion of the lesion being a neoplasm and since no other diagnosis was established on frozen section, complete surgical resection was performed. However, the immunohistochemical staining done later on for histiocytes (CD68), the LFB/PAS/CV (Luxol fast blue/ periodic acid-Schiff /cresyl violet) stain showing extensive myelin loss, and the neurofilament stain showing some degree of myelin loss with relative preservation of axons as compared to myelin provided the definitive diagnosis of TDL rather than tumor (Fig. 1).

Although an uncommon condition, TDL of the spinal cord should always be on the differential diagnosis of spinal cord tumors and thus a biopsy with immunostaining should be performed for all suspicious cord lesions to confirm the diagnosis before any treatment. Since the prognosis of TDL is the same as that for population based MS, medical treatment of TDL usually improves the neurological symptoms of these patients (Scott *et al*., 2005); hence it is very important to distinguish between TDL and glioma early before surgery since this has a major impact on the patient's outcome.

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