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Seminars in
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Transverse Myelitis and Neuromyelitis Optica Spectrum Disorders

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Transverse myelitis is defined as inflammation of the spinal cord, named because of its typical clinical presentation with bandlike symptoms of altered sensation or pain in a horizontal fashion—at a specific dermatome level. Radiographic patterns might vary but the idiopathic form is more frequent to present as involvement of 3-4 vertebral segments and both sides of the cord. It is now recognized that there are numerous other causes as well as the idiopathic type, with often atypical features and geographic variation. There is also increasing recognition of other forms of myelitis, particularly the longitudinally extensive manifestation with involvement of 3 or more vertebral segments. Neuromyelitis optica, one of these subtypes can be diagnosed by means of an antibody assessment. The picture is more complicated with the expansion of the description to involve neuromyelitis optica spectrum disorders, new antibodies such as myelin oligodendrocyte glycoprotein and the inclusion of an antibody-negative variant. This article describes the different entities of transverse myelitis, with a particular focus on neuromyelitis optica spectrum disorders.
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Introduction

The term "Transverse myelitis" refers to a *clinical* syndrome affecting the spinal cord. There are many causes and increased use of magnetic resonance imaging (MRI) is central to establishing the correct diagnosis. Transverse myelitis is a diagnosis of exclusion and imaging may be used initially to diagnose other causes of the same clinical symptoms (eg, extrinsic compression). Once other causes are excluded, cases can be subdivided into either idiopathic "acute" transverse myelitis or disease-associated "acute" transverse myelitis secondary to conditions such as multiple sclerosis. The onset is typically acute or subacute and it is recommended that the term "transverse myelitis" is used in all cases.[1]

The inflammation of the spinal cord generally affects a clinical level but deficits are not always bilateral and do not always affect all long tracts in the cord, leading to the use of terms such as partial transverse myelitis. The use of the word "transverse" is not a description of the changes demonstrated radiologically or pathologically, but rather is a clinical term referring to the usual bandlike symptoms that patients experience—classically an area of altered sensation or pain in a horizontal (ie, transverse) band usually at the dermatomal level corresponding to the lesion within the cord. Weakness, sensory disturbance, and autonomic dysfunction are characteristic.[2]

Transverse myelitis may extend across several segments of the cord leading to the use of the term longitudinally extensive transverse myelitis (LETM). This finding is traditionally associated with neuromyelitis optica (NMO) but can also be associated with other diseases.[3]

It is important that reporting radiologists recognize the terminology and understand the implications as determining the cause of the myelitis can be challenging and increasingly early treatment is guided by detailed imaging with effective interpretation. There has been significant development in the treatment options for the various subgroups of transverse myelitis and these often need to be started quickly to obtain the best possible outcome for the patient. Several of the treatment options are toxic with significant side effects and their usage may be guided by accurate imaging to prevent exposing patients to unnecessary risk.

In this article, we aim to describe the imaging findings in idiopathic and disease-associated transverse myelitis and

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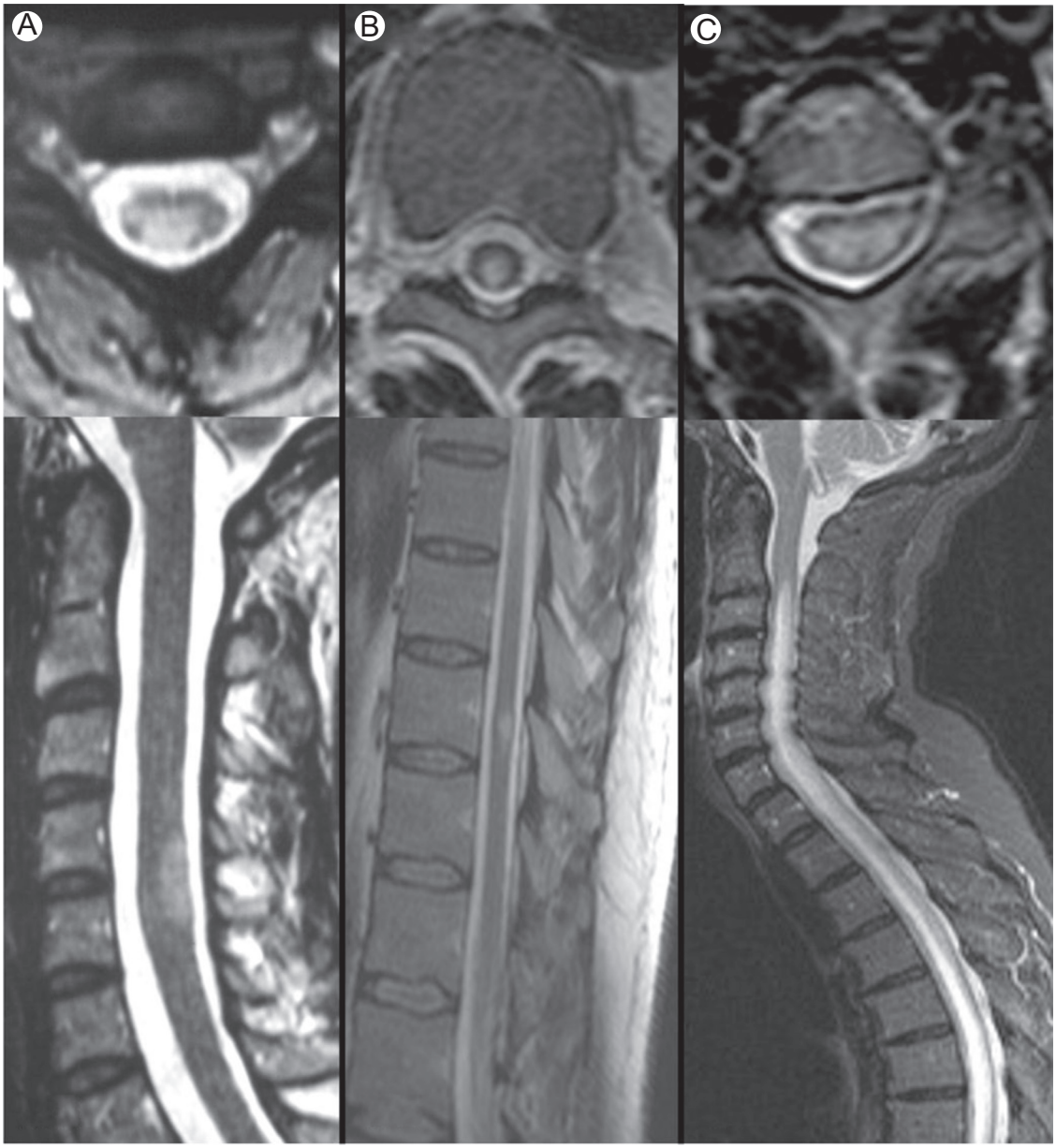

Figure 1 Appearances of different inflammatory lesions within the spinal cord. (A) A typical slightly eccentric T2-hyperintense multiple sclerosis lesion causing focal cord expansion, involving less than 2 vertebral segments (A-2), the dorsal aspect of the cord and not respecting the gray-white matter boundary (A-1). (B) A case of idiopathic transverse myelitis—although the lesions are classically described as involving >2 segments of the cord (and typically 3-4 segments), in clinical practice they are often smaller; the lesion is T2 hyperintense centrally within the cord and is causing mild cord expansion. (C) A longitudinally extensive transverse myelitis in a patient with antibody-positive neuromyelitis optica; there is a central T2 hyperintensity in the expanded cord (C-1), extending from the C2-3 level through the visualized thoracic cord (C-2).

demonstrate how MRI can be used to assist in the diagnosis of the various subgroups of "transverse" myelitis.

Background

Transverse myelitis is a generic clinical (but translated into a radiological) description for a group of conditions causing cord dysfunction due to a *noncompressive* myelopathy. Clinical symptoms generally develop over hours to days. The term "transverse myelitis" was previously reserved for the idiopathic cases but the current practice is to use it in all cases to describe the general clinical syndrome.

The etiology can be inflammatory, vascular, paraneoplastic, treatment-related (radiation), or idiopathic. Transverse myelitis has an estimated incidence of between 1.34 and 4.6 per million[4,5]; or an estimated incidence of up to 3 per 100,000 patient-years (0.003%). There is no known familial, ethnic, or geographical variation in the incidence.[6] The seeming female preponderance is probably explained by the fact that this