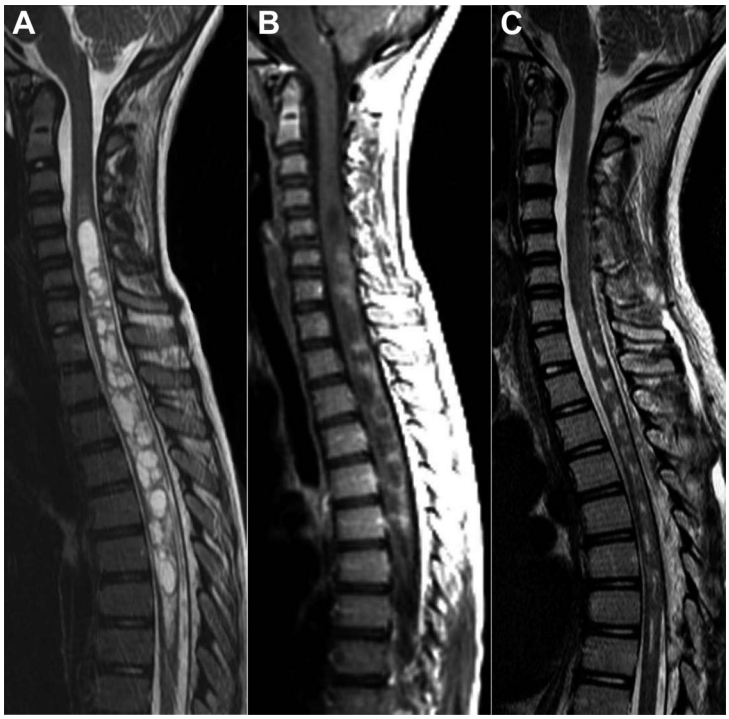
# Multicystic demyelinating myelopathy

## Widening spectrum of pediatric aquaporin-4 autoimmunity

**Figure 1** Baseline and repeat spinal cord MRI after immunotherapy




Sagittal T2-weighted MRI demonstrates a large multiloculated cystic lesion within the spinal cord extending from C4 to T7 (A), with patchy peripheral nodular enhancement on postcontrast scan (B). (C) Decreased axial extension of the lesion and improvement in spinal cord edema and expansion 4 weeks after steroid therapy.

A 10-year-old girl presented with subacute lower limb weakness and gait ataxia. MRI revealed a large multicystic spinal cord lesion with patchy enhancement (figure 1, A and B) and 3 small (<6 mm) periventricular and deep white matter brain lesions. The presence of serum anti-aquaporin-4 (AQP4) immunoglobulin G (ELISA assay) and compatible neuropathologic features from neurosurgical specimens[1] (figure 2) suggested the diagnosis of a neuromyelitis optica spectrum disorder.[2] Targeted immunotherapy was started, with partial lesion resolution (figure 1C).

This case provides neuroradiologic evidence for macroscopic multicystic cord demyelination in AQP4-related disorders and highlights the role of inflammatory etiologies in childhood spinal cord disease.

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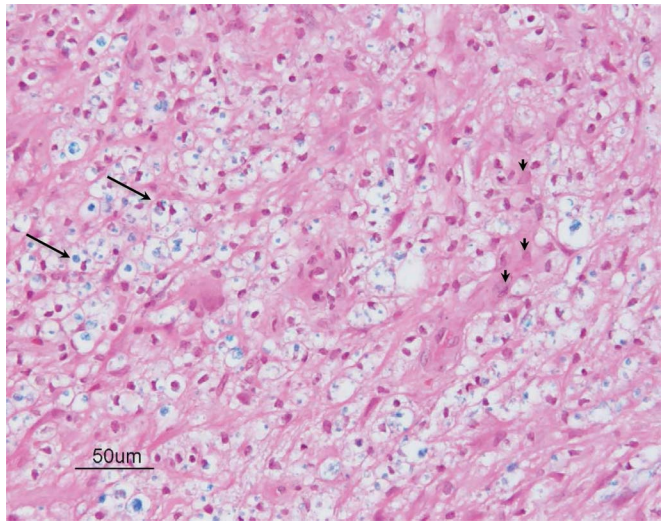
**Figure 2** Spinal cord biopsy




Hematoxylin & eosin/Luxol fast blue (LFB) stained section from the spinal cord biopsy demonstrates sheets of macrophages (arrows) containing LFB-positive debris and scattered reactive astrocytes (arrowheads) suggestive of an active demyelinating process (200×).

1. Lucchinetti CF, Mandler RN, McGavern D, et al. A role for humoral mechanisms in the pathogenesis of Devic's neuromyelitis optica. Brain 2002;125:1450–1461.
2. McKeon A, Lennon VA, Lotze T, et al. CNS aquaporin-4 autoimmunity in children. Neurology 2008;71:93–100.

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