Multicystic demyelinating myelopathy
Widening spectrum of pediatric aquaporin-4 autoimmunity

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 1A 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 (figure 2) suggested the diagnosis of a neuromyelitis optica spectrum disorder. 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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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×).

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