

Development of Neuromyelitis Optica Spectrum Disorder and Spinal Arachnoid Cysts in a Patient With Intractable Epilepsy

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Neuromyelitis optica is an inflammatory, demyelinating disease of the central nervous system that is characterized by severe relapsing attacks of optic neuritis and transverse myelitis. The current case describes a 29-year-old man with intractable epilepsy and diplegic spastic cerebral palsy who was given the diagnosis of neuromyelitis optica spectrum disorder after presenting with weakness, incontinence, and decreased visual acuity. His symptoms recurred 21 months after initial presentation. Magnetic resonance imaging of his spine revealed arachnoid cysts with regional mass effects. Differentiation of arachnoid cysts from a demyelinating process may be difficult in the early stages of the disease. Close monitoring of patients with neuromyelitis optica spectrum disorder is important, especially in patients with recurrent or refractory symptoms.

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Neuromyelitis optica (NMO; also known as *Devic disease*) is an inflammatory disorder of the central nervous system characterized by severe, immune-mediated demyelination and axonal damage, predominantly targeting the optic nerves and spinal cord.¹ The first clinical descriptions of NMO emerged more than a century ago when Eugène Devic and Fernand Gault documented a series of patients with a monophasic course of bilateral or rapidly sequential optic neuritis and myelitis.² Disability after these attacks was often severe. This disease is distinct from classic relapsing-remitting multiple sclerosis with respect to pathogenesis, imaging results, biomarkers, neuropathologic symptoms, and response to treatment.³

Early-stage diagnostic specificity is critical in cases of NMO because observational data suggest that some treatment options used in patients with multiple sclerosis, which can be mistaken for NMO, may worsen NMO symptoms.⁴ The discovery of a disease-specific serum NMO immunoglobulin G (IgG) antibody that selectively binds to aquaporin-4 (AQP4) has led to increased understanding of a diverse spectrum of disorders.⁵ The 2015 diagnostic criteria for NMO spectrum disorder (NMOSD)⁶ facilitates earlier and more accurate diagnosis, especially for cases in which the AQP-4 antibodies are not detectable or not available. Alternatively, the treatment path for NMOSD may be useful for future prevention of the disorder.

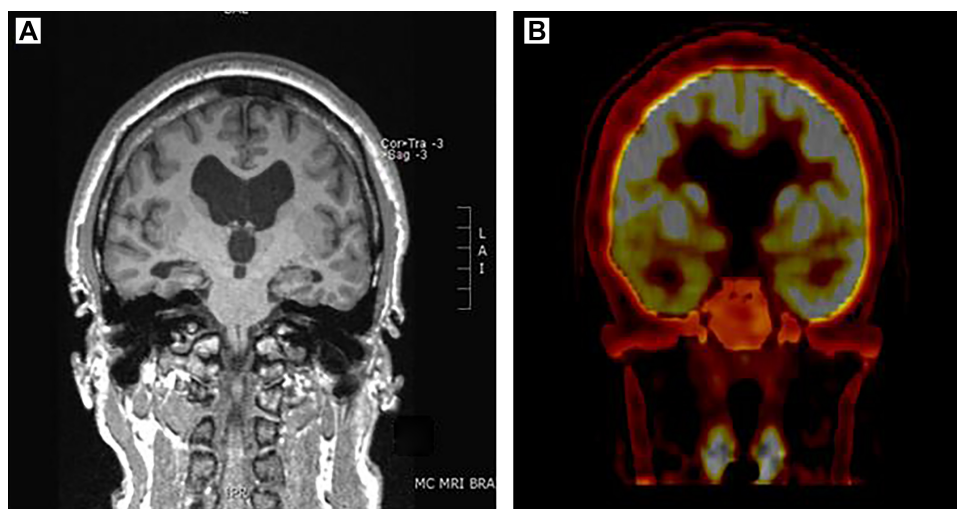


Figure 1.

Magnetic resonance images of the brain of a man presenting with increased bilateral distal leg weakness and spasticity showed (A) stable, moderate enlargement of the lateral, third, and fourth ventricles and (B) asymmetric decreased right temporal lobe volume.

Report of Case

A 29-year-old man with childhood-onset medically intractable epilepsy and spastic diplegic cerebral palsy presented to the clinic with a 6-month history of increased bilateral distal leg weakness, spasticity, rapidly worsening urinary and bowel incontinence for the past 3 weeks, and decreased visual acuity and pain with eye movements. Magnetic resonance (MR) images of the brain showed stable, moderate enlargement of the lateral, third, and fourth ventricles (**Figure 1A**), which is consistent with a compensated communicating hydrocephalus, and asymmetric decreased right temporal lobe volume (**Figure 1B**).

Physical examination revealed 5 out of 5 muscle strength in the bilateral upper extremities. Hip flexion was 4/5 strength; knee extension, 4/5; and knee flexion, 4/5 bilaterally. Additional manual muscle testing scores were as follows: right dorsiflexion, 3/5; plantar flexion, 4/5; left dorsiflexion, 0/5; and plantar flexion, 1/5. A sensory level was found at T4. Deep tendon reflex strength scores were 2/5 on the bilateral upper extremities, 4/5 on the right patella, and 3/5 on the left patella. Sustained clonus was present in both

ankles. The patient had an up-pointing toe on the right foot and severe spasticity in the bilateral lower extremities.

An MR image of the thoracic spine showed deformation of the spinal cord from T5 to T7 and multiple small arachnoid cysts. Abnormal T2 hyperintense signal within the spinal cord and multiple outward septations suggested circumferential loculations around the spinal cord (**Figures 2A** and **2B**). Myelography was performed to further investigate the circumferential loculations, and the results showed a spinal cord deformity from T5 to T7 with a hyperdense right-sided extramedullary component, which could have represented a communication between the lesion and the cerebrospinal fluid. Spinal arteriography showed no evidence of early venous drainage or arteriovenous fistula. Results of a visual evoked potential test demonstrated a P100 latency of 102 ms on the left and 125 ms on the right. Amplitude was 2.9 μ V on the left and 0.2 μ V on the right. Differential white blood cell count, blood glucose level, and total protein level were normal. Cerebrospinal fluid IgG index and oligoclonal band levels were within the normal range. Serum

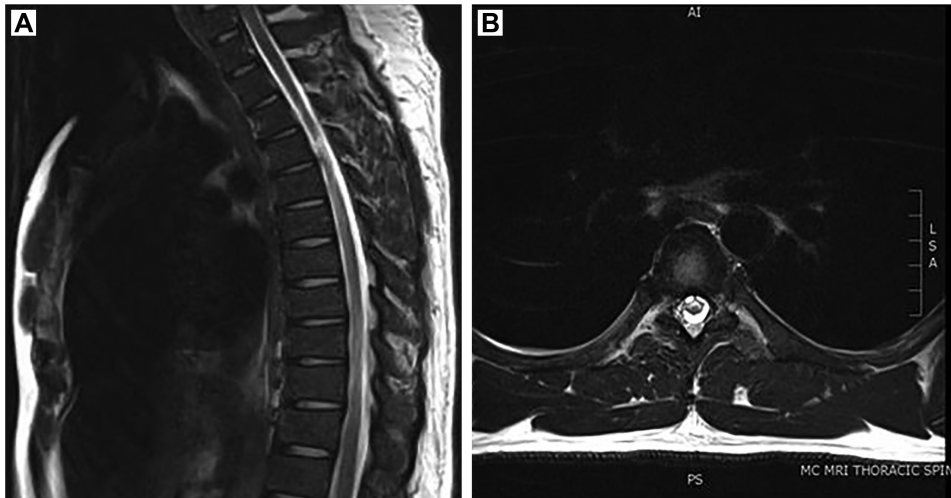


Figure 2.

Magnetic resonance images of the thoracic spine of a man presenting with increased bilateral distal leg weakness and spasticity showed demyelinating changes from T5 to T7 and (B) a deformation of the spinal cord.

NMO-IgG assay test results were negative. Test results for infectious diseases, autoimmune disorders, and nutritional abnormalities in the blood were negative.

Because of the patient's eye pain, vision loss, visual evoked potential test results, and acute myelitis extending over more than 3 continuous segments in the thoracic spine, the patient was given the diagnosis of NMOSD.¹ He was given 250 mg of methylprednisolone intravenously every 6 hours for 7 days. Rituximab infusion was subsequently initiated at 375 mg/m² once per week for 2 weeks.

At an 18-month follow-up appointment, results of a repeated serum NMO-IgG assay test were negative. The patient underwent a right temporal lobectomy 1 month after the follow-up appointment and has been seizure-free while taking his current medications (gabapentin, lamotrigine, and zonisamide). Two months after the lobectomy (21 months after initial presentation), the patient had a relapse of NMOSD and presented to the clinic with decreased visual acuity and increased weakness. Rituximab was initiated at 375 mg/m² once per week for 2 weeks, and his vision gradually improved over the following 2 months. However, no change in his bilateral

lower extremity weakness and spasticity was noted. An MR image of the thoracic spine showed an intradural and extramedullary nonenhancing cyst containing cerebrospinal fluid signal along the ventral thoracic spinal cord from T3 to T5. The cyst was eccentric to the left and measured 4×7 mm. Associated mild mass effect on the ventral T3-5 thoracic spinal cord was noted (Figure 3A). A similar 5×4-mm nonenhancing cyst containing cerebrospinal fluid signal was also present along the ventral T5 to T6 thoracic spinal cord on the right with regional mass effect (Figure 3B). No abnormal enhancement was noted (Figure 3C). Progression of myelomalacia involving the anterior thoracic spinal cord was present at T3 and T4 and abnormal T2 hyperintensity and myelomalacia involving the thoracic spinal cord from T5 to T7 (Figure 3D). The patient subsequently underwent drainage of the arachnoid cysts. Physical therapy was initiated while the patient was in the hospital and continued as outpatient sessions. He had a poor clinical response because of baseline weakness and spasticity and, as of December 2017, continues to be dependent on using a front-wheeled walker at home and a wheelchair for longer distances.

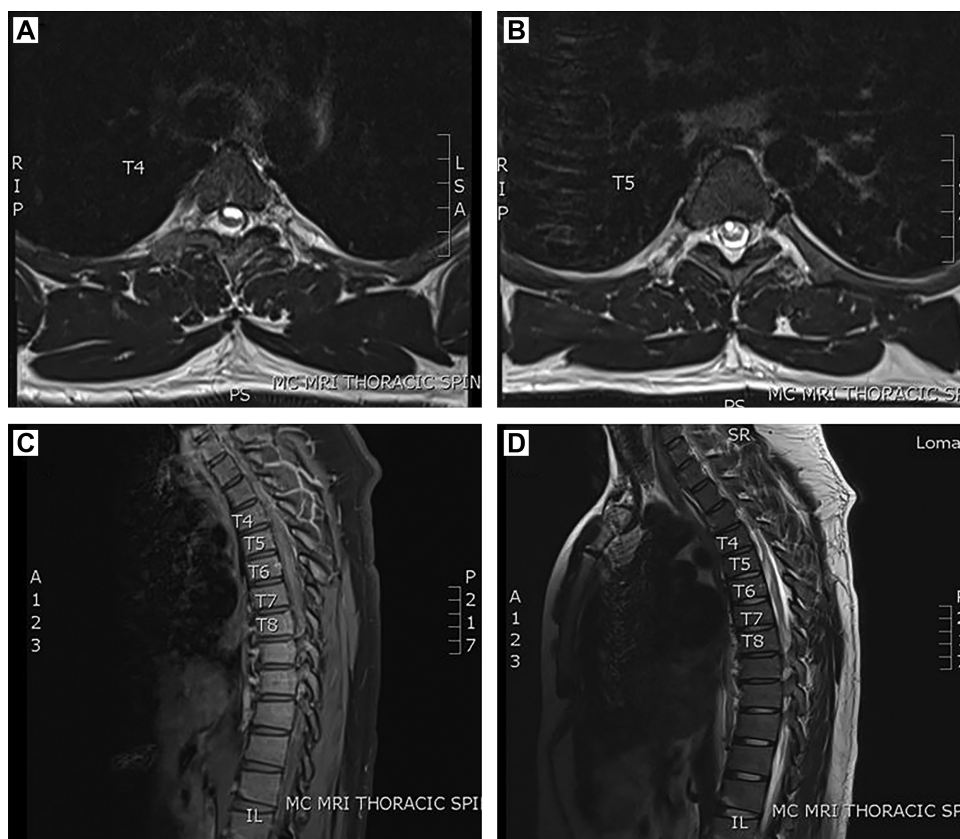


Figure 3.

Magnetic resonance image taken 18 months after the patient's initial presentation showed (A and B) probable arachnoid cysts along the ventral surface of the T3 to T5 and T5 to T6 thoracic spinal cord with regional mass effect, (C) no abnormal enhancement, and (D) progression of myelomalacia involving the anterior thoracic spinal cord at T3 and T4.

Discussion

Treatment should be initiated early in patients with NMOSD. Azathioprine and rituximab are suggested as first-line treatments, the latter being increasingly regarded as an established treatment option with long-term efficacy and an acceptable safety profile.⁷ Methotrexate, mycophenolate mofetil, and mitoxantrone are recommended as second-line treatments.^{7,8}

In the current case, the diagnosis of NMOSD was made according to the 2015 diagnostic criteria.⁶ The overall clinical presentation of the patient was complicated by a rare occurrence of progressively enlarging spinal arachnoid cysts. To the authors' knowledge, no reported correlations between NMOSD and idiopathic spinal arachnoid

cysts have been reported. Reported contributing factors to spinal arachnoid cysts include traumatic spinal cord hemorrhage,⁹ iatrogenic causes,¹⁰ and genetics.^{11,12} Because of the rarity of spinal arachnoid cysts, the optimal surgical treatment remains controversial.¹³ Compared with fenestration, total resection carries a greater risk of complications, including kyphosis.¹⁴ Minimally invasive surgical procedures may be used in the management of intradural spinal arachnoid cysts to reduce complications.¹⁵

Conclusion

The 2015 NMOSD diagnostic criteria allows for early diagnosis and management of the disorder.

Differentiation of arachnoid cysts from a demyelinating process on MR imaging in the early stages can be difficult. Close monitoring of patients with NMOSD is important because of high relapse rates, especially in patients with recurrent or refractory symptoms.

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