






NMO Spectrum Disorder or Mass?

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ABSTRACT

Longitudinally extensive transverse myelitis (LETM) generally present a destructive clinical syndrome which has come into focus for its association with neuromyelitis optica spectrum disease (NMOSD). In clinical practice, both LETM and NMO have a close relation so they thought to be synonymous with each other. Other causes of LETM are infective, neoplastic and connective tissue disorders. Similar symptoms and even similar CSF abnormalities can cause difficulties in differential diagnosis. In suspicious cases, beside more detailed history, tight observation of clinical progress and follow-up MRI, pathological evaluation can be helpful to certain diagnosis.

INTRODUCTION

Longitudinally extensive transverse myelitis (LETM) is a typical feature of neuromyelitis optica, but such spinal lesions can also seen in multiple other autoimmune and inflammatory diseases that involve the CNS (such as acute disseminated encephalomyelitis, multiple sclerosis, sarcoidosis or Sjögren syndrome) or in infectious diseases with CNS involvement. Patients with a neoplastic disorder or traumatic spinal cord injury can also present with longitudinal spinal lesions like inflammatory diseases.^[1]

Intramedullary tumours of the spinal cord are quite infrequent. The most common intramedullary tumors are astrocytomas and ependymomas which together account for 90% of them. These lesions can cause important difficulties in the differential diagnosis between inflammatory diseases (such as acute disseminated encephalomyelitis, multiple sclerosis), vascular abnormalities and neoplasms.^[2]

Intramedullary tumours, particularly ependymomas and astrocytomas often appear hyperintense on T2-weighted

imaging and usually extend across multiple vertebral segments like neuromyelitis optica spectrum diseases.^[3] Here we report a case which has been treated as NMOSD first but after diagnosed as intramedullary tumor.

CASE REPORT

18-year-old male patient admitted to our hospital with complaints of weakness and numbness in both legs. It was learned from the patient's history that he was referred to a tertiary care center with bilateral weakness in lower extremities and urinary incontinence complaints which started 40 days ago. In the radiological images performed there, a spinal lesion was detected and pulse steroid was started with a pre-diagnosis of demyelinating disease. After a while, he stated that the weakness partially resolved but started again. He did not state any complaints about vision. Past history included only a vehicle accident one year ago which did not cause any neurological symptom. No alcohol or smoking was reported. There was no feature in his family history.



Figure 1. Hyperintense lesion in the t2-weighted imaging along T1-T3



Figure 2. Hyperintense lesion in the T2-weighted imaging along C6-T2.

In the neurological examination; muscle strength was detected at upper limb bilateral 5/5, at right lower limb proximal and distal 1/5, at left lower limb proximal 2/5 distal 3/5. Bilateral hypoesthesia was obtained below T12. Bilateral position sense loss, bilateral decreased sense of vibration was detected in lower extremities. Deep tendon reflexes have increased globally. Babinski was extensor on the right and there was no response on the left.

Complete blood count, liver, kidney and thyroid function tests, blood glucose, electrolytes; sodium, potassium, calcium, ionized calcium, phosphorus, magnesium, erythrocyte sedimentation rate and C-reactive protein were normal.

His spinal MR imaging showed a hyperintense lesion was seen extending longitudinally along T1-T3 levels in T2-weighted sections (Fig. 1).

Vitamin B12 level was normal and syphilis serology was negative. The patient underwent lumbar puncture. No cells were seen in the cerebrospinal fluid (CSF). Oligoclonal band in CSF and Serum anti-aquaporin 4 antibody (NMO IgG) were negative. Vasculitis markers, angiotensin converting enzyme level and paraneoplastic markers were negative. There was no finding that could be evaluated in favor of malignancy in thorax CT.

OCT was performed on the patient who did not have symptomatic optic neuritis. On the left superior and inferior; on the right superior and nasal retinal nerve fiber layers (RNFL) thinning was observed.

A diagnosis of seronegative NMO spectrum disorder was considered accompanied by longitudinal transverse myelitis and optic nerve involvement as asymptotically. Along with longitudinal transverse myelitis and optic nerve asymptomatic effect, a diagnosis of NMO spectrum disorder was considered. 0.4 mg / kg / day IVIG treatment was given for 5 days. The patient gained increase his muscle strength with pulse steroid and IVIG treatment was called for a control examination 15 days later.

Progression was observed in the control visit. Follow up spinal MRI was taken and expansive view at the thoracic level was shown growth towards the cervix. (Fig. 2) Plasmapheresis was added to treatment scheme but patient had no clinical benefit. Biopsy decision was given and Diffuse midline glial tumor grade 4 was shown (Fig. 3). Tumor was resected by surgeons and radiotherapy was applied to patient.

DISCUSSION

Current diagnostic criteria of NMO spectrum diseases were published by the International Panel for NMO diagnosis in 2015. The criteria classify the diagnosis by those with AQP4-IgG and those without AQP4-IgG (including those whom AQP4-IgG negative or not tested). The main clinic features are determined as the three cardinal manifestations of optic neuritis, myelitis, and an area postrema syndrome, in addition to less common manifestations of other brainstem attacks, diencephalic episodes, and cerebral episodes. AQP4-IgG positivity and one main feature are sufficient for diagnosis, whereas the criteria for patients who are AQP4-IgG seronegative are more stringent, requiring additional characteristic radiologic features be present to help avoid misdiagnosis.^[4]

Almost 20% to 25% of patients with NMO spectrum disorder are AQP4-IgG seronegative. The treatment approach is similar in AQP4-IgG-seronegative NMO spectrum disorder and AQP4-IgG-seropositive NMO spectrum disorder; Assay techniques have improved over time, cell-based assays are now recommended and they yield a sensitivity of 75% to 80% and specificity of greater than 99%.^[5]

Clinical symptoms of intramedullary tumors are nonspecific, including local or less frequently irradiating pain. Motor weakness, gait problems and bowel and bladder dysfunction are another common symptoms of intramedullary tumors.^[6] Demyelinating disease, MS or ADEM also present with the same symptoms.

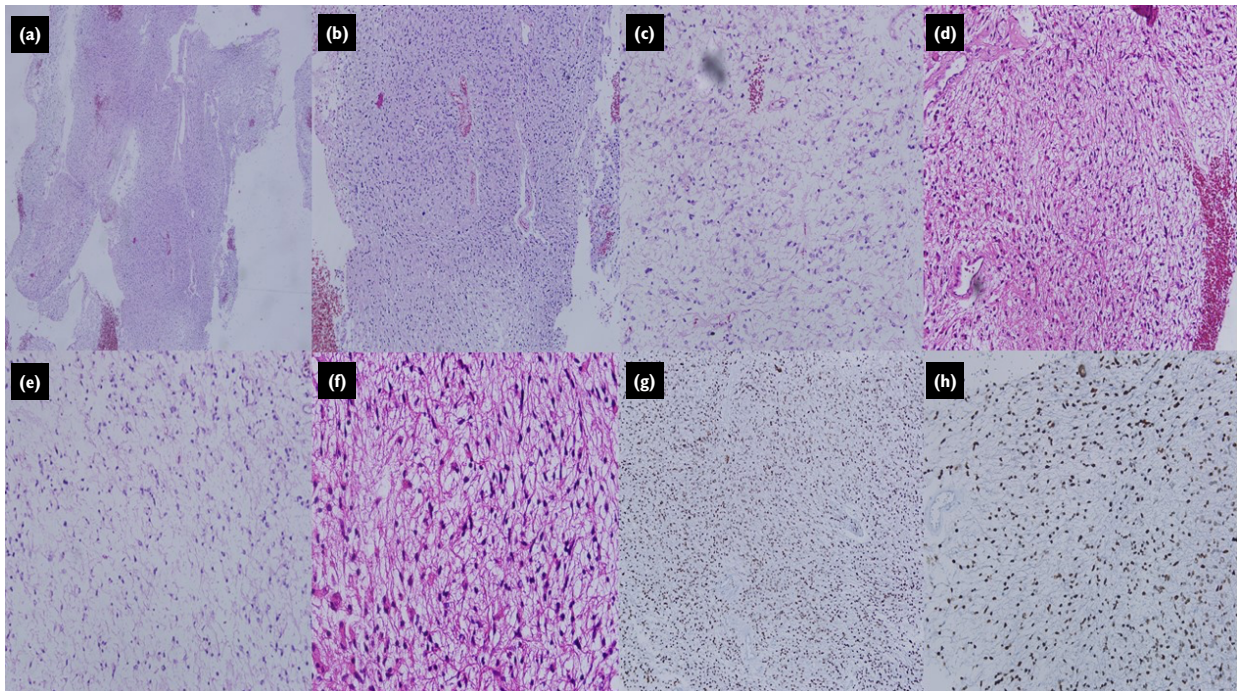


Figure 3. (a,b,c,d,e,f) Hematoxylin and eosin stain showing neoplastic cells, and (g,h) H3 K27M immunostain showing positive neoplastic nuclei.

In a study conducted by Brinar and friends^[2] with five cases with spinal lesions presenting with weakness and paresthesia, open biopsy was considered in all patients. It was performed in two patients. Inflammation related tissue has been reported in both.

In the case reported by Habek and friends,^[7] the patient was admitted with the acute myelopathy clinic, and pulse steroid therapy was started considering the demyelinating disease. Although the treatment, progression was continued as clinical and radiologically. Biopsy was performed and interpreted as an intramedullary tumor. Intense inflammation was seen in pathology. Additionally performed CSF analysis, which revealed positive oligoclonal IgG bands. Serum NMO-IgG antibody was positive. The patient was diagnosed with spatially limited NMO spectrum disorder, treated with plasma exchange, high-dose corticosteroids, and cyclophosphamide, and recovered well.

In the case reported by Jakob and friends^[8] Inflammatory LETM has been considered due to its clinical features, oligoclonal band positivity and positive response to corticosteroid treatment. Biopsy performed and spinal tumor was diagnosed.

In our case, seronegative NMO Spectrum disease was considered in the patient who had transverse myelitis attacks and had asymptomatic optic neuritis findings. Despite applying pulse steroid, ivig and plasmapheresis treatments, clinical and radiological progression continued. Biopsy was performed considering intramedullary mass in differential diagnosis, midline diffuse glial tumor stage 4 was detected.

CONCLUSION

Although longitudinal extensively spinal cord lesions are most frequently caused by inflammatory lesions (especially NMO), non-inflammatory causes should also be considered. Clinic of myelopathy related with spinal masses deteriorates in an acute-subacute course but it should be remembered that acute-subacute onset can also be observed in advanced masses with rapid progression.

Clinical features, neurological examination, laboratory tests and MRI findings should be evaluated together. Suspicious cases should be followed up closely. Clinical and radiologic progression despite the treatment should alert clinician.

Informed Consent

Retrospective study.

Peer-review

Externally peer-reviewed.

Authorship Contributions

Concept: N.Y.A, A.K.K.; Design: N.Y.A, A.K.K., D.Y., T.H., B.Ö.B.; Supervision: N.Y.A, A.K.K., D.Y., T.H., B.Ö.B.; Fundings: N.Y.A, A.K.K., D.Y., T.H., B.Ö.B.; Materials: D.I.; Data: N.Y.A, A.K.K., D.Y., T.H., B.Ö.B.; Analysis: N.Y.A, A.K.K., D.Y., T.H., B.Ö.B.; Literature search: N.Y.A, A.K.K., D.Y., T.H., B.Ö.B.; Writing: N.Y.A, A.K.K., D.Y., T.H., B.Ö.B.; Critical revision: N.Y.A, A.K.K., D.Y., T.H., B.Ö.B.

Conflict of Interest

None declared.

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NMO mu? Kitle mi?

Longitudinal yayılan transverse miyelit, genellikle nöromyelitis optika spektrum hastalıkları ile ilişkili ağır bir kinik sendrom olarak ortaya çıkar. Ancak etiolojide otoimmün nedenler yanında enfektif, neoplastik ve bağ doku hastalıkları ile ilişkili süreçler bulunabilir. Benzer semptomlar ve BOS bulguları ayırıcı tanıda zorluğa sebep olabilir. Şüphe durumunda; ayrıntılı öykü, yakın klinik ve görüntüleme yanında patolojik değerlendirme doğru tanıya ulaşmada yönlendirici olabilir.

Anahtar Sözcükler: Demiyelinizan; glial tumor; nmo, spinal kitle.