CASE REPORTS

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Recurrent longitudinally extensive transversal myelitis in a patient with Sjögren's syndrome

Rekurentni longitudinalno ekstenzivni transverzalni mijelitis kod bolesnika sa Sjegrenovim sindromom

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Abstract

Introduction. Longitudinally extensive transverse myelitis (LETM) is a transversal myelitis that extends through three or more vertebral segments in length. **Case report.** A 52-year-old woman was hospitalized due to pain in the lumbar region, difficulty in walking, hypoesthesia of the anogenital area and urinary retention. In the past medical history, two years earlier, the patient had been diagnosed with transversal myelitis confirmed by MRI of the cervical spine and six months earlier, the patient was diagnosed with primary Sjögren's syndrome (SS). During the current hospitalization MRI of the spinal cord revealed extensive inflammatory lesions of almost the whole spinal cord. Lumbar puncture (LP) revealed mild pleocytosis and slightly increased protein level. Isoelectric focusing of cerebrospinal fluid (CSF) and serum proteins was normal. Visual evoked potentials were normal. Serological testing excluded acute viral infections. Corticosteroid therapy was applied with good therapeutic response. Control MRI revealed regression of pathological changes in the spinal cord. **Conclusion.** A wide range of disorders can cause LETM, but usually the first line diagnosis is neuromyelitis optica (NMO). Based on the detection of NMO immunoglobulin G in the serum of affected patients, a variety of allied disorders were grouped under the name of NMO spectrum disorders, including recurrent myelitis associated with LETM and myelitis associated with autoimmune disorders such as SS. There have been only a few cases reported in the literature with recurrent LETM associated with non-organ specific autoimmune disorder.

Key words:
myelitis, transverse; sjorgen's syndrome; neuromyelitis optica; diagnosis; drug therapy; treatment outcome.

Apstrakt

Uvod. Longitudinalno ekstenzivni transverzalni mijelitis (LETM) je transverzalni mijelitis koji zahvata tri ili više susednih segmenata kičmene moždine. **Prikaz bolesnice.** Bolesnica, stara 52 godine, hospitalizovana je zbog bolova u lumbalnom delu kičme, otežanog hoda, hipestezije anogenitalne regije i retencije urina. Pre dve godine, magnetnom rezonancom (MR) cervikalne kičme dijagnostikovan joj je transverzalni mijelitis, a pre šest meseci primarni Sjegrenov sindrom (SS). Tokom sadašnje hospitalizacije MR kičmenog stuba verifikovana je ekstenzivna inflamatorna lezija koja je zahvatala kičmenu moždinu skoro celom dužinom. Analizom likvora utvrđena je blaga pleocitoza i blaga hiperproteinorahija. Nalaz izoelektričnog fokusiranja proteina likvora i seruma bio je uredan, kao i nalaz vizuelnih evociranih potencijala. Serološkim testovima isključena je akutna virusna infekcija. Ordinirana je kortikosteroidna terapija uz dobar terapijski odgovor. Kontrolnim nalazom MR potvrđena je regresija inflamatorne promene kičmene moždine. **Zaključak.** Brojne bolesti i poremećaji mogu uzrokovati LETM, ali na prvom mestu neuromijelitis optika (NMO). Na osnovu detekcije NMO imunoglobulina G u serumu bolesnika, brojne srodne bolesti grupisane su pod imenom NMO spektar bolesti, koji uključuje rekurentni mijelitis povezan sa LETM, kao i transverzalni mijelitis u sklopu autoimunih oboljenja kao što je SS. U literaturi je opisano samo nekoliko slučajeva rekurentnog LETM povezanog sa organ nespecifičnim autoimunim bolestima.

Ključne reči:
mijelitis, transverzalni; sjegrenov sindrom; neuromijelitis optika; dijagnoza; lečenje lekovima; lečenje, ishod.

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Introduction

Longitudinally extensive transverse myelitis (LETM) is a relatively recent term designating a transversal myelitis (TM) that extends through three or more vertebral segments in length [1]. It is much rarer and has more severe prognosis than other types of TM. LETM is usually associated with neuromyelitis optica (NMO), but as a possible diagnosis systemic autoimmune diseases [Sjögren's syndrome (SS), systemic lupus erythematosus (SLE), antiphospholipid syndrome], neuroinflammatory conditions (Behcet's disease, sarcoidosis), multiple sclerosis (MS), and infectious diseases should always be considered [2].

Case report

A 52-year-old woman suddenly felt severe pain in the right lumbar region without irradiation. Two days later, the patient noticed difficulty in walking and hypoesthesia of the anogenital area accompanied with urinary retention. Since the patient had had similar problems before, she immediately contacted the physician who referred her to our hospital.

Regarding the past medical history from other hospital center, the patient had been diagnosed with TM confirmed by magnetic resonance imaging (MRI) of the cervical spine that showed intramedullary lesion extending from medulla oblongata downwards to Th3 level of the spinal cord (Figure 1A), two years earlier. At that time, computed tomography (CT) scan of the brain revealed no abnormalities. Lumbar puncture (LP) showed increased cell count with lymphocyte predomination. Serological laboratory tests of serum and cerebrospinal fluid (CSF) for common viruses and *Borrelia burgdorferi* were negative. The deficit that remained after the rehabilitation treatment was discrete muscular weakness of the left arm and the right leg. Control MRI of the cervical spine, performed tree times in a 2-year period, revealed regression of pathological changes in the spinal cord. Meanwhile, after some period of pain, swelling and stiffness in both hands and feet, in the Institute for Rheumatology the patient was diagnosed as primary SS based on: clinical findings (xerophtalmia, xerostomia) laboratory findings [positive antinuclear antibodies (ANA), anti Ro/SS-A antibodies, rheumatoid factor (RF)], ultrasound of salivary glands (focal areas of inflammation)]. It is important to emphasize the fact that patient did not use the prescribed therapy – hydroxychloroquine.

On current admission to our neurological department, the patient was alert, orientated, with all vital signs within normal limits. Neurological findings were as follows: deviation of tongue to the right, mild to moderate quadriparesis with increased tendon reflexes, hypoesthesia of all extremities with a level of decreased sensibility on Th4 dermatome and below, urinary retention and obstipation.

Standard laboratory tests showed elevated sedimentation rate, and mild anemia. C-reactive protein (CRP) level was normal and lactate dehydrogenase (LDH) elevated. MRI of the spinal cord revealed extensive inflammatory lesions of almost the whole spinal cord (Figure 1 B). MRI of the brain showed a few confluent T2W and fluid attenuated inversion recovery (FLAIR) hyperintense lesions of brainstem and cerebellum. Lumbar puncture revealed pleocytosis (101/mm^3) with lymphocyte predomination (82%) and increased protein level (0.82 g/L). Isoelectric focusing of CSF and serum proteins was normal. Visual evoked potentials (VEP) were normal. Abdominal ultrasound showed only liver hemangioma. Chest x-ray was without pathological changes. Serological testing for viruses, in consultation with an immunologist and infectologist, excluded acute viral infections.

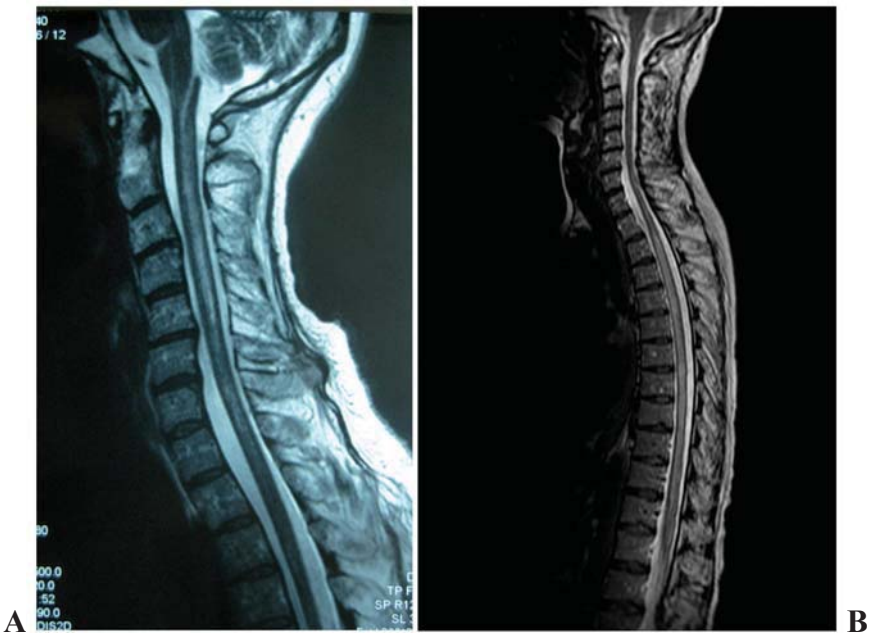

Fig. 1 – Magnetic resonance (MR) T2-weighted sagittal image from 2009.
A – The area of increased signal intensity accompanied with swelling (smooth expansion) of the medulla oblongata, the entire cervical part and the first three levels of the thoracic part of the spinal cord, that could be attributed to acute longitudinal myelitis. The same sequence on the magnetic resonance imaging (MRI) of the spine from 2011. B – Newly formed intramedullary lesions of the same MR characteristics, from Th3 downwards making a conus that correlates with actual clinical findings. Note the chronic atrophic changes of the previously affected medulla oblongata and cervical spine.

Based on immunological findings – increased RF, positive ANA, decreased levels of C3 and C4, normal lupus anticoagulant (LA) and anti double stranded (anti-ds) DNA antibodies, and positive Schirmer's test, regarding all previous and new findings, an immunologist confirmed the diagnosis of SS.

We applied corticosteroid therapy with intravenous methylprednosolone 1 g daily during six days, switched to prednisolone 1 mg/kg daily, with good therapeutic response and gradual recovery of symptoms.

Control MRI of the spine, performed two weeks later, revealed initial regression of pathological changes in spinal cord. Control LP findings were within normal limits.

The patient was discharged after 18 days on prednisolon therapy, with her previous deficit (mild hemiparesis and discrete hemihypesthaesia of the left limbs), but incontinent.

Discussion

A wide range of disorders can cause LETM, but usually the first line diagnosis is NMO [1]. Since the discovery of a specific serum biomarker, neuromyelitis optica immunoglobulin G (NMO-IgG), the concept of understanding and definition NMO has changed [3]. This biomarker distinguishes NMO from other demyelinating disorders. Based on the detection of this biomarker in the serum of affected patients, a variety of allied disorders are grouped under the name of NMO spectrum disorders (NMOSD), also including recurrent myelitis associated with LETM and optic neuritis or myelitis associated with autoimmune disorders such as SLE and SS [4]. Recurrent TM associated with longitudinal spinal cord lesions appear to be rather NMO-IgG seropositive. NMO-IgG seropositivity after the first attack predicts a relapsing course in most of the patients within three years of the first attack [5].

Sjögren's syndrome can be complicated with neurological problems, and frequently neurological signs are the first manifestations of SS. According to the study of Delalande et al. [6], neurological complications are the first symptom of SS in 81% patients, about 35% have spinal cord involvement and one third of these have acute myelopathy. In some patients, NMO and non-organ specific autoimmune disorders, particularly SS or SLE, coexist. It is the result of the presence of NMO-IgG in some patients with SS or SLE who have neurological involvement [7].

In the presented case, the first attack of LETM was initially considered idiopathic, but after confirming the diagnosis of SS it was assumed as first manifestation of SS. Current clinical findings and diagnostic procedures excluded sarcoidosis, Behcet's disease, SLE, metastatic tumors, viral infection as possible diagnosis. The patient did not fulfill diagnostic criteria for MS. Devic's disease was considered unlikely due to the absence of absolute diagnostic criteria – optic nerve lesions on MRI, also negative VEP, but subclinical NMO or NMOSD could not be excluded because we were not able to test NMO-IgG (not available in our laboratories). In the context of current findings, recurrent LETM in the presented case, even without testing NMO-IgG, made us to conclude that the presented patient probably had NMOSD.

There have been a few cases in the literature on LETM and recurrent LETM associated with non-organ specific autoimmune disorder [8–10], but, to our knowledge, there have been no reports on recurrent LETM associated with SS. Moreover, we found the presented case interesting because inspite of the fact that LETM extended to practically entire spinal cord, the patient had good outcome.

Conclusion

A wide range of disorders can cause LETM, but usually the first line diagnosis is NMO. Based on the detection of NMO immunoglobulin G in the serum of affected patients, a variety of allied disorders were grouped under the name of NMO spectrum disorders, including recurrent myelitis associated with LETM and myelitis associated with autoimmune disorders such as SS. There have been only a few cases reported in the literature with recurrent LETM associated with non-organ specific autoimmune disorder.

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