

# Idiopathic Recurrent Transverse Myelitis With Syringomyelia: A Case Report

## Siringomyeli ile Birlikte Tekrarlayıcı İdiopatik Transvers Myelit: Olgu Sunumu

### ABSTRACT

Idiopathic transverse myelitis is a monophasic demyelinating disease of the central nervous system (CNS) and its recurrence is a rare entity. Existence of syringomyelia in the spinal cord in inflammatory CNS disease is not usually encountered. This case study describes a 45-year-old man who was admitted with features of a cervical demyelinating disease. The history of the patient indicated that this could be a recurring attack. Cervical MRI and laboratory examination of the patient showed evidence of recurrent transverse myelitis. The patient was managed with medical therapy. The follow-up cervical MRI of the patient 2 years later indicated syringomyelia in the cervical cord.

**KEY WORDS:** Multiple sclerosis, Spinal cord, Transverse myelitis

### ÖZ

İdiopatik transvers myelit santral sinir sisteminin monofazik demyelinizan bir hastalığıdır ve tekrarlaması nadir olarak görülür. İnflamatuvar santral sinir sistemi hastalıkları ile beraber spinal kordda siringomyeli görülmesi sık karşılaşılan bir durum değildir. Sunduğumuz 45 yaşında erkek olan bu olgu servikal demyelinizan hastalık özellikleri ile başvurdu. Olgunun alınan öyküsünde bunun bir tekrarlayan atak olduğu öğrenildi. Olgunun servikal MR ve laboratuvar tetkikleri tekrarlayan transvers myelit tanısını doğruladı. Olgu medikal yöntemlerle tedavi edildi. Olgunun 2 yıl sonraki takip MR larında servikal kordda siringomyeli gözlemlendi.

**ANAHTAR SÖZCÜKLER:** Multiple sklerosis, Spinal kord, Transvers myelit

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## INTRODUCTION

Idiopathic transverse myelitis (TM) is a monophasic demyelinating illness of the central nervous system (CNS) and its recurrence is a rare entity. It is characterized by bilateral motor, sensory, and autonomic dysfunction of the spinal cord in the absence of any pre-existing neurological disease. Recurrence usually heralds, or is accompanied by, the evidence of dysfunction elsewhere such as the presence of multiple sclerosis (MS) (2,3,5,8,10).

Syringomyelia may exist secondary to congenital anomalies, neoplasms of the spinal cord, arachnoid adhesion, spinal cord injury and myelomalacia (6,9,10). The existence of syringomyelia in the spinal cord in inflammatory CNS disease is not usually encountered (6). In this case, we have described idiopathic recurrent transverse myelitis (RTM) with syringomyelia and spinal cord atrophy.

## CASE REPORT

A 45-year-old male patient was admitted to our emergency room with complaint of a sudden loss of power in four extremities while defecating. Neurological examination revealed quadriplegia and incontinence of miction and defecation.

The patients' history indicated that he had previously been admitted to the hospital with emergent double vision, paresthesia and loss of power in left side four years previously. He became a fully remitted outpatient within 15 days after high doses of intravenous methylprednisolone (1g/day for 7 days) for a diagnosis of myelitis.

Routine laboratory examination indicated that human immunodeficiency virus (HIV), human T-lymphotropic virus (HTLV) and collagen vascular screening tests were negative. Electroencephalography (EEG) and electromyography (EMG) were normal. Routine studies including visual evoked potentials (VEP) and brainstem auditory evoked potential (BAEP) were normal but lengthened latency was observed in somatosensorial evoked potentials (SEP). Antibodies to Epstein-Barr virus, cytomegalovirus, herpes virus and a panel of respiratory viral pathogens were negative. There was slight cerebrospinal fluid (CSF) pleocytosis (13 wbc) with normal protein, and no oligoclonal bands (OCBs).

Cranial MRI findings were normal, but cervical MRI showed an intramedullary lesion at the C3-Th1 segments of the spinal cord with a low-intermediate

signal on T1WI and a high signal on T2WI without Gd-contrast enhancement. The anteroposterior diameter of the cord was found to be increased minimally at the lesion level (Figure 1, 2).

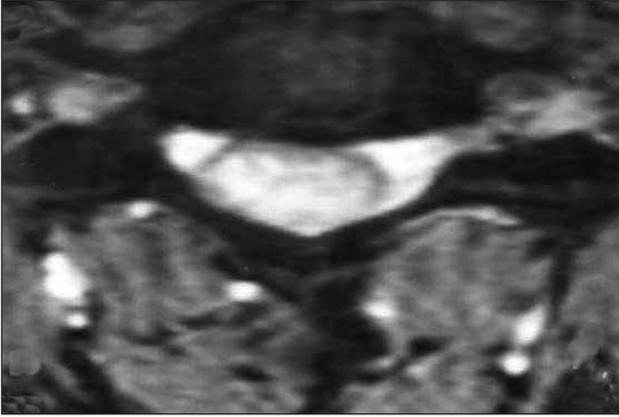
Idiopathic RTM was considered in the differential diagnosis bearing in mind the history of the patient and the findings described above. The patient was treated with high doses of intravenous methylprednisolone (1g/day for 10 days) and a tapering course of oral prednisone. He was discharged without symptoms after 20 days. No symptoms were reported by the patient during the two years of follow-up except fatigue and paresthesia in lower extremities. At the end of this period, there was no indication of any previous lesion, the diameter of the spinal cord was decreased and syringomyelia was seen in follow-up MRI (Figure 3).

## DISCUSSION

Idiopathic RTM with syringomyelia is a rare entity. It is an interesting topic in terms of the causative factors and therapeutic considerations. It is



**Figure 1:** T2W1 sagittal MRI scans showing hyperintense image in the spinal cord at the C3-Th 1 levels.



**Figure 2:** T2W1 axial MRI scan showing hyperintense image in the spinal cord.



**Figure 3:** T2W1 sagittal MRI scans showing syringomyelia at the C1-7 levels together with spinal cord atrophy.

difficult to differentiate from MS but can be differentiated from MS-RTM on the basis of the clinical manifestations of myelopathy, or MRI or CSF examination findings. Additionally, systemic lupus erythematosus, antiphospholipid antibody syndrome, isolated angiitis of the CNS, HIV, herpes simplex infections, and spinal vascular malformations have been reported to produce recurrent isolated cord syndrome (10).

MRI scans indicated no difference in the location of spinal cord lesions between the idiopathic RTM and MS-RTM groups on T1WI- and T2-weighted images. Lesions in the middle or upper cervical cord regions were more frequently involved in MS studies (2,3,5,10). Eccentric or peripheral enhancement was detected mostly in myelinated spinal cord white matter. As described for patients in other studies, characteristic patterns of myelopathic MS lesions involving discrete multiple segments or oval enhancement were not detected in idiopathic RTM. A few focal spinal lesions were detected in idiopathic RTM as in other cases of MS-RTM (5,10).

In the case under discussion, the patient had normal cranial MRI with idiopathic RTM and did not demonstrate paraclinical evidence of the disease process of spatial dissemination beyond the spinal cord (5,10). The cervical MRI showed an intramedullary lesion at the C3-Th1 levels of the spinal cord with low-intermediate signal on T1WI and high signal on T2WI without Gd-contrast enhancement.

Idiopathic RTM occurs primarily in male patients and presents more often with acute TM than with MS-RTM (1-3). Jeffrey et al. (4) reported recurrence in one of the six cases of idiopathic TM. Deeb et al. (1) reported that the lesion of TM occurs more frequently at the thoracic spinal cord rather than the cervical cord. However, the affected region was cervical (C3-Th1) in our case. Although cord atrophy is emphasized in TM, there are no reports about the formation of syringomyelia in RTM cases (2,3,5,8,10). Patel et al. (9) have previously reported a case with posttraumatic syringomyelia and transverse myelitis.

This case suggests that idiopathic RTM might be a distinct morbid entity from MS-RTM, with male preponderance, absence of OCBs, frequent multiple relapses, and frequent presentation with acute TM (5,7,10).

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