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Case Report
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A Case Report of Acute Transverse Myelitis with Anti-Amphiphysin Positivity

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Abstract

Transverse Myelitis (TM) is a rare, acquired spinal cord disease that may present with rapid-onset weakness, sensory changes, and bowel or bladder dysfunction. It may occur different reasons. Paraneoplastic myelopathies are one of the rare etiologies. Anti-amphiphysin positivity associated with breast cancer and Stiff-Person Syndrome in women may rarely lead to isolated TM. We report a 35-year-old case that presents with paraplegia which occurred gradually in 20 days accompanied by as sensory defect at T8 level. MRI showed multiple peripheral hyperintensities localized at cervical and lower thoracic levels, with millimetric contrast enhancement along with anti-amphiphysin positive results from paraneoplastic antibodies after IVMP and plasmapheresis treatment with maintenance oral prednisolone.

Keywords: Transvers myelitis; Paraneoplastic myelitis; Anti-amphiphysin

Introduction

Transverse Myelitis (TM) is a rare, acquired spinal cord disease that may present with rapid-onset weakness, sensory changes, and bowel or bladder dysfunction. Although it may be post-infectious, it may be the clinical manifestation of a neuro-inflammatory disease such as acute disseminated encephalomyelitis, multiple sclerosis, Myelin Oligodendrocyte Glycoprotein Antibody Disease (MOGAD), Neuromyelitis Optica Spectrum Disorder (NMOSD), and acute flaccid myelitis. Paraneoplastic syndromes are among the rare causes of TM [1]. In this report, we present a 35-year-old female patient who applied to our hospital with a TM clinic and was found to be positive for anti-amphiphysin antibodies in the examinations.

Case Presentation

A 35-year-old female patient was admitted to our hospital with complaints of weakness and numbness in the legs that started 20 days ago. Her complaints started as numbness in the right leg and weakness occurred within the same day, and spread to the left leg the next day. Within a week, she was unable to walk due to increasing complaints. There was no known disease in her history. In her family history, her father was diagnosed with Brucella infection 9 years ago with the complaint of progressive gait disorder and completely recovered with treatment in her Neurological Examination (NM), Medical Research Council (MRC) grades were 5/5 on her upper limbs and 1/5 on her bilateral lower limbs. Her systemic physical examination was unremarkable. She accompanied sensory defect on T8 level. Her cerebellar tests were normal. Her vibration was decreased in bilateral lower extremities. Deep Tendon Reflexes (DTR) were symmetrically brisk at the upper extremities and hypoactive at the lower limbs. Plantar Skin Reflexes (PSR) had bilateral extensor response. There was no sphincter defect.

MRI showed no pathological findings in brain and had multiple peripheral T2 hyperintensities localized at cervical and lower thoracic levels, with millimetric contrast enhancement in spinal cord (Figure 1, 2).

In the lumbar puncture, Cerebrospinal Fluid (CSF) protein was 30 mg/dL (N: 15-45 mg/dL) and 2 leukocytes were observed. There was no growth in CSF culture. Brucella, Borrelia tests results were negative in CSF and serum. The oligoclonal band was found type 1 pattern. NMO-MOG antibodies were negative. In Visual Evoked Potentials (VEP) examination, bilateral P100 wave lateness slightly long. Upon detection of CSF ACE 4 IU/L (0-2.6 IU/L), Serum ACE 165 IU/L (52 IU/L), contrast-enhanced thorax CT and 24-h urine calcium were requested for sarcoidosis, it was found to be

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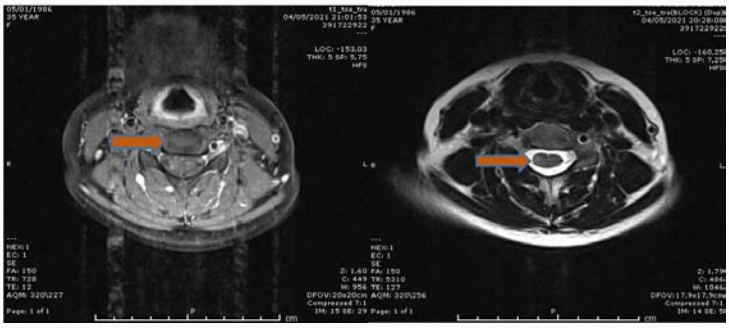

Figure 1: C4-5 level T1 sequence with contrast and T2 axial sequence.

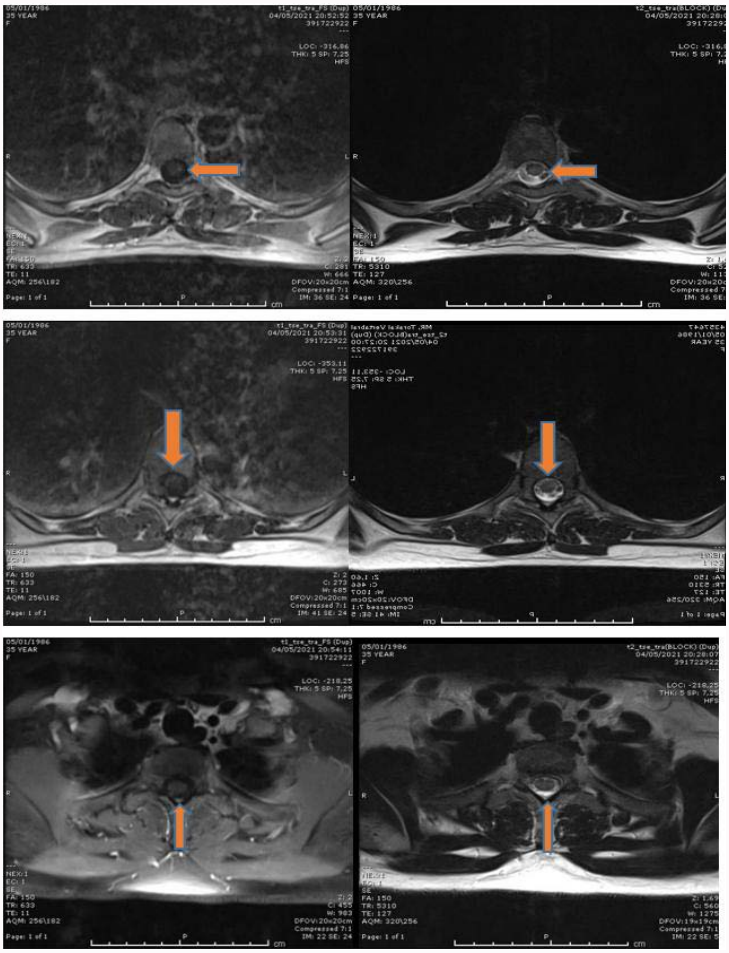

Figure 2: T5-6, T9-10, T11-12 level T1 with contrast sequence and T2 axial sequence.

normal. The COVID PCR test was negative, COVID IgM antibody negative, anti-SARS-CoV IgG antigen 162.8 (normal value: Negative) positive, which was performed 1 month before her clinic due to a history of mild flu infection.

In the researches for malignancy screening, CA-125: 114 U/mL (0-35 U/mL). In her abdominal CT; in the middle quadrants of the left breast, which entered the image area, the area evaluated in favor of the linearly extending asymmetric dilated duct, which is observed asymmetrically compared to the right breast, was observed. But her breast ultrasonography revealed no pathological findings.

With the diagnosis of autoimmune TM, 1 gr/day Intravenous Methylprednisolone (IVMP) was given for 7 days. After treatment, MRC grades were 2/5 on her bilateral lower extremities. A decrease in sensory defect was detected. Since the motor weakness continued significantly, 7 sessions of plasmapheresis and 10 gr IVMP were applied. After the treatment, MRC grades were 3/5 on proximal parts and 4/5 on distal parts of her bilateral lower limbs.

The patient was discharged while walking with support, with weekly IVMP and Azathioprine (AZA) 50 mg/day planned to be started and gradually increased. AZA was discontinued at 1 month of treatment due to skin rashes. Oral steroid treatment was given at a dose of 32 mg/day, gradually tapering off.

After the treatment was planned, the paraneoplastic panel was found to be anti-amphiphysin + positive. Since MS, NMO Spectrum diseases, MOGAD, sarcoidosis and other autoimmune and infectious TM causes were excluded in the investigations of her, it was thought that the current clinical and imaging findings might be related to anti-amphiphysin. The whole-body PET-scan was made. Focally increased FDG uptake was observed in the posterior part of the uterine corpus. It was evaluated as myoma in the foreground. No findings in favor of malignancy were observed in the gynecological examination and biopsy. The paraneoplastic antibody panel was repeated in the first year and it was found to be anti-amphiphysin negative. On her last NM; MRC grades were 5/5 on her lower and upper extremities, increased DTR responses in her lower limbs, PSRs had bilateral extensor response. Her clinical follow-up and malignancy scans are continuing.

Discussion

Paraneoplastic syndromes are one of the rare causes in the etiology of transverse myelitis. The most frequently observed autoantibodies; anti-Hu, anti-Collapsin-Response Mediator Protein 5 (CRMP5), and lesser anti-amphiphysin is observed [1]. Amphiphysin is an intracellular neuronal protein that plays a role in vesicle membrane uptake at the axon terminal [2]. Somer et al. conducted studies describing the direct pathogenic role of amphiphysin in paraneoplastic Stiff-Person syndrome [2]. Although its association with SPSS is common in the literature, isolated cases of TM accompanying malignancies have also been reported. Galassi et al. reported that a 40-year-old female patient with subacute onset paresthesia, predominant paraparesis in the distal lower extremities, ataxia and found to be positive for long segment myelitis anti-amphiphysin was diagnosed with breast cancer during the follow-up [3]. Carrette et al. reported a 70-year-old male patient who presented with bilateral optic neuritis and had rapidly developing progressive paraparesis and urinary incontinence on the 10th day of hospitalization, with short segment multiple demyelinating lesions on cervical and dorsal MRI in spinal examinations and diagnosed with prostate cancer in his scans [4].

Paraneoplastic causes should also be considered in cases of transverse myelitis with atypical imaging and CSF findings, and appropriate treatment should be initiated for patients with antibody positivity. Although malignancy is not detected in patients with paraneoplastic antibody positivity, as in our case, malignancy screening should be performed regularly.

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