Central Nervous System Involvement Mimicking Multiple Sclerosis in a Patient With Sjogren's Syndrome

İsmail BOYRAZ, Bünyamin KOÇ, Selma YAZICI

Department of Physical Medicine and Rehabilitation, Medical Faculty of Abant Izzet Baysal University, Bolu, Turkey

Sjogren’s syndrome (SS) is a systemic autoimmune disease that can involve both the peripheral nervous systems and central nervous systems (CNS). It is characterized by a lymphocytic infiltration of all exocrine glands. Neurologic symptoms including transverse myelitis, optic neuritis, stroke-like acute symptoms or several neurological disorders like progressive relapsing remitting attacks mimicking multiple sclerosis (MS) may develop in patients with SS. Herein, we reported a patient who had been treated/followed-up previously with the diagnosis of MS and was diagnosed subsequently with SS.

A 55-year-old male patient was admitted to the neurology outpatient clinic with complaints of weakness and numbness in left leg. Steroid treatment was administered with a diagnosis of MS in 1998, based on the findings of a cranial magnetic resonance imaging. Cerebrospinal fluid examination was normal, except for the slightly elevated protein level and presence of a single oligoclonal band. His complaints-recurred one week later following the treatment, and speech disorder developed one year later. The patient suffered two episodes of the disease within one year, and prophylactic interferon beta-1a was initiated. The magnetic resonance imaging in 2005 revealed resolution in the brain lesions and chronic infarctions. In the first physical examination at the initial period of the complaints, he had common paresis and cerebellar functions were impaired. Muscle weakness deteriorated gradually. The diagnosis of primary SS was considered in 2005 upon onset of mouth and eye dryness. Anti-Sjogren syndrome antigen B and other autoimmune antibodies were negative and inflammatory markers were within the normal range. Anti Ro-52 was weakly positive. A salivary test was positive, while the Schirmer and pathergy tests were negative. No sign of vasculitis was detected in the selective cerebral angiography. Sialography was normal and a salivary gland biopsy was compatible with SS score of 3. A skin biopsy revealed ruptured/destructed folliculitis, and therefore we excluded vasculitis. As the patient had dry eyes for at least three months and symptoms of dry mouth for at least three months, unstimulated whole salivary flow, abnormal salivary gland biopsy, and Anti-Ro (+), we have diagnosed the patient as SS according to European diagnostic criteria and determined CNS involvement of SS mimicking MS.

The incidence of neurological involvement in primary Sjogren's syndrome (pSS) is about 10-60%. The exact pathogenic mechanisms of the neurological involvement are still unknown. Some authors suggested that CNS involvement results from small vessel vasculitis. The rate of nerve involvement is 20-25% in SS, 87% of which can be attributed to peripheral nervous systems involvement. Cerebral involvement is heterogeneous, while CNS involvement may...
sometimes mimic MS. Peripheral neuropathy, myositis, seroimmunological disorders and cutaneous vasculitis, which are rarely seen in cases of MS, may indicate “MS-like pSS”. Cerebrospinal fluid electrophoresis in pSS demonstrates one or two oligoclonal band formation, while multiple bands are seen in MS. Oligoclonal bands may be detected in 90% of MS patients and in 20-25% of pSS patients, although these bands may resolve with steroid treatment.

In conclusion, CNS involvement in patients with pSS may mimic some other diseases like MS, as seen in our patient. Clinical, laboratory, and radiological findings should be evaluated and examined carefully for differential diagnosis.

Declaration of conflicting interests
The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

Funding
The authors received no financial support for the research and/or authorship of this article.

REFERENCES