Sjogren’s myelitis presenting as hemicord syndrome

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We present a case where a 63-year-old right-handed man who presented with a 6-month history of progressive asymmetrical sensorimotor symptoms in lower limbs. This was associated with concomitant rash on the lower limbs, and mild sicca symptoms. MRI spine showed focal T2 hyperintensity in the left hemicord at C3-4 level. Skin biopsy of the rash revealed urticarial vasculitis, and lip biopsy revealed lymphocytic sialadenitis. Initial anti-Ro antibody was negative, but subsequent Ro52 antibody testing returned positive. There was also matched serum and cerebrospinal fluid oligoclonal bands. He was subsequently diagnosed as Sjogren’s myelitis and treated with intravenous methylprednisolone, then transitioned to a steroid sparing agent. This case highlights the difficulties in reaching a rheumatological diagnosis in the early stages with typical negative antibodies, and shows a rare neurological manifestation of a systemic rheumatological condition.

KEYWORDS: Sjogren’s syndrome, myelitis, hemicord syndrome

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Case report

A 63-year-old right-handed man presented with a 6-month history of acute onset, asymmetrical, progressive sensorimotor disturbance in the lower limbs. He experienced a burning sensation associated with tingling and reported heaviness in the legs. His exercise tolerance had dropped in the preceding 3 months, and his gait became unsteady. A rash in his legs also appeared in parallel with the neurological symptoms. He also reported mild sicca symptoms (dryness in eyes and mouth) over the preceding few years. He was previously fit and healthy and his only notable medical history was a ruptured spleen from a car accident.

On examination, he had predominantly right-sided weakness (Medical Research Council (MRC) score 4 on the upper and lower limbs) and left-sided paraesthesia. A sensory level was apparent (Medical Research Council (MRC) score 4 on the upper and lower limbs). Multiple rashes were present on both legs, suggestive of vasculitis. MRI (Fig 1) showed focal T2 hyperintensity in the right lateral aspect of the spinal cord at C3-4 level without any enhancement. There was moderate to severe spondylosis, with exit foraminal stenoses at the cervical level, which were considered unrelated to his symptoms.

Sjogren’s myelitis was considered because of the vasculitic rash and sicca symptoms. A skin biopsy of the rash was performed, which confirmed urticarial vasculitis. He also had positive anti Ro52 antibody, and matched serum/cerebrospinal fluid (CSF) oligoclonal bands. A lip biopsy revealed lymphocytic sialadenitis with focus score of 2.

A diagnosis of Sjogren’s myelitis was made and he was treated with 1 g intravenous methylprednisolone for 3 days, followed by oral prednisolone. Due to the ongoing severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2) pandemic at the time, we opted to maintain immunosuppression with azathioprine instead of more potent immunosuppression with rituximab. His progress was complicated by autonomic dysreflexia and associated fatigue. After 3 months of neurorehabilitation, he was able to stand using his walking frame, and mobilise around in a manual self-propelled wheelchair. He remains under the care of both the neurology and rheumatology teams.

Discussion

Sjogren’s syndrome is a relatively rare autoimmune inflammatory condition affecting mainly exocrine functions, with an estimated prevalence of 60 per 100,000. Dermatological manifestations are the most common presenting feature. However, neurological symptoms are not uncommon, and were found to affect 18.9% in a French cohort of primary Sjogren’s syndrome.

Diagnosis was difficult initially, given the patient presented with a hemicord syndrome and a vasculitic rash in the absence of classical rheumatological phenotype (no dry eyes or mouth) and having sparse serological findings (negative ANA, ANCA/hepatitis screen, normal immunoglobulin panel) to support an underlying rheumatological condition. Ro52 antibodies, in the absence of ANA antibodies, are of uncertain significance. However, according to the joint diagnostic criteria from American College of Rheumatology and the European League Against Rheumatism (ACR-EULAR), we were able to make a diagnosis of Sjogren’s syndrome based on positive salivary gland biopsy.

In the context of neurological symptoms, peripheral neuropathy occurs more commonly than central symptoms; in the instances of central cases, a longitudinally extensive transverse myelitis is often found. However, what we found in our patient was an area of inflammation affecting his hemicord at the C3/4 level. Anti-aquaporin antibodies were negative, making neuromyelitis
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in Sjögren’s-associated myelitis. Chlorambucil, azathioprine, methotrexate and rituximab have also been used. In our case, azathioprine was chosen in agreement with the patient in consideration of his concerns about stronger immunosuppression at the time of the Sars-Cov2 pandemic.

Conclusion

We present a rare neurological manifestation of a systemic rheumatological condition. In the workup of myelitis, we often check extractable nuclear antigen (ENA) titres. In the presence of sicca symptoms, a positive ENA is more supportive of a systemic inflammatory condition, highlighting the importance of clinical correlation and role of lip biopsy (tissue diagnosis) where suspicion is high. Finally, although cyclophosphamide is often the preferred therapy in this cohort of patients, the selection of the immunosuppressant remains a multi-disciplinary decision. Deliberations need to be made with regards to the potential risks against benefits, both in the short and long term. The patient needs to understand and be comfortable with the treatment plan.

References

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