Case Report and Literature Review: A 36-Year-Old Woman with Recurrent Syncope and Neuromuscular Symptoms

Introduction

Sjogren's syndrome (SS) is the second most common autoimmune disease after rheumatoid arthritis. Neurological impairment in SS varies from 10% to 60% across studies, mainly due to differences in case detection methods. Autonomic dysfunction in SS can range from mild symptoms to more severe conditions like orthostatic hypotension and excessive postural tachycardia. Although inflammatory myopathy is rare in SS, sporadic inclusion body myositis (sIBM) has a particular association with primary SS (pSS) (1-2).

Case Report

A 36-year-old African American woman presented with a five-year history of persistent dizziness, frequent syncope triggered by positional changes, muscle weakness in her upper and lower extremities, and shock-like sensations in her arms. She was initially diagnosed with dysautonomia and postural orthostatic tachycardia syndrome (POTS) by cardiology and was treated with beta-blockers, though her symptoms did not improve. A physical exam revealed muscle weakness in the distal extremities and decreased grip strength. Cardiac evaluations revealed no cardiac cause for her syncope, leading to a differential diagnosis of dysautonomia due to neuromuscular issues, adrenal insufficiency, or autoimmune disease.

Further Diagnostic and Therapeutic Course

Rheumatological workup showed a positive antinuclear antibody (ANA) test and strongly positive anti-SSA antibodies. A salivary gland biopsy confirmed Sjogren's syndrome, showing focal lymphocytic sialadenitis with atrophy. The patient was started on Plaquenil (5 mg/kg), IVIG (400 mg/kg), and azathioprine. Although the muscle weakness was not initially attributed to myopathy, suspicion arose for inclusion body myositis (IBM). Testing for the anti-cN1A (NT5c1A) IBM (RDL) antibody yielded a moderately positive result. An MRI of her right thigh muscles was normal, but a muscle biopsy showed group atrophy with intrafascicular fibrosis and type 2 muscle fiber atrophy.

Discussion

Although uncommon in Sjogren's syndrome, autonomic dysfunction can involve any organ regulated by the autonomic nervous system. Identifying myopathy, particularly sporadic inclusion body myositis (sIBM), is particularly challenging when it coexists with autoimmune conditions such as SS. Although the muscle biopsy results were atypical, the patient's moderately positive anti-cN1A antibodies raised further suspicion of sIBM. Muscle biopsies can appear normal in 10% of inflammatory myopathy cases (3). Sporadic inclusion body myositis has a recognized association with Sjogren's syndrome, sharing a common MHC class II genetic predisposition. In some instances, sIBM may manifest before or simultaneously with the diagnosis of SS (4-5).

Conclusion

The patient’s progressive muscle weakness and autonomic dysfunction led to a diagnosis of Sjogren’s syndrome, with suspected sIBM due to elevated cN1A antibodies. Accurate diagnosis and treatment require a thorough evaluation of muscular and autonomic symptoms.

References

1. Vivino FB, et al. "Sjogren's syndrome: An update on disease pathogenesis, clinical manifestations, and treatment." Clin Immunol. 2019;203:81-121.
2. Goodman BP, et al. "Spectrum of Autonomic Nervous System Impairment in Sjögren Syndrome." Neurologist. 2017;22(4):127-130.
3. Gran JT, Myklebust G. The concomitant occurrence of Sjögren's syndrome and polymyositis. Scand J Rheumatol. 1992;21(3):150-4.
4. Chung SH, et al. "Sporadic inclusion body myositis and primary Sjogren's syndrome: An overlooked diagnosis." Clin Rheumatol. 2021;40(10):4089-4094.
5. Kanellopoulos P, Baltoyiannis C, Tzioufas AG. Primary Sjögren's syndrome is associated with inclusion body myositis. Rheumatology (Oxford). 2002;41(4):440-4.