"Severe Autonomic Dysfunction and Chronic Myopathy as Initial Manifestations of Sjogren's Disease with Suspected Sporadic Inclusion Body Myositis"

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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)

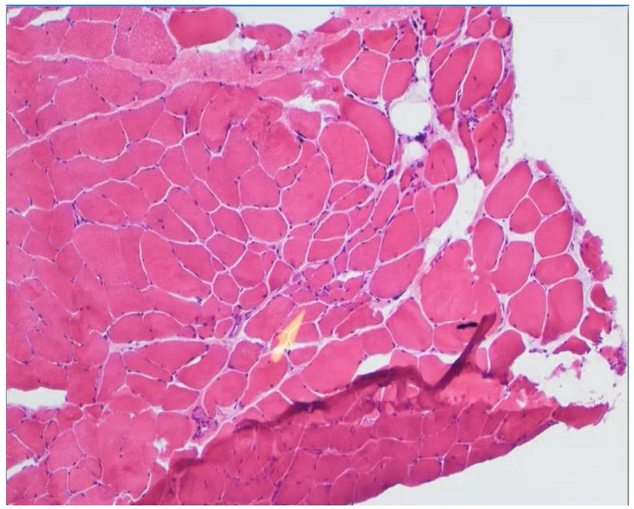
The case

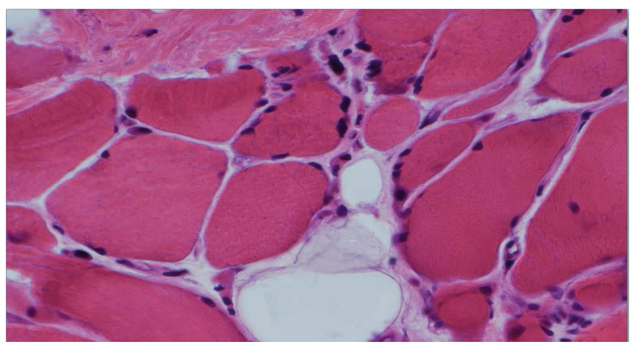
A 36-year-old African American woman presented with a five-year history of persistent dizziness, frequent syncope triggered by positional changes, muscle weakness in her upper and lower extremities, and shock-like sensations in her arms. She was initially diagnosed with dysautonomia and postural orthostatic 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.

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.

Figures 1 and 2 show a muscle biopsy of the anterior tibialis muscle showing a single focus of group atrophy with intrafascicular fibrosis and atrophy of Type 2 muscle fibers.





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 . 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.

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.

Conclusion

This patient's slow, progressive muscle weakness and autonomic dysfunction led to a diagnosis of Sjögren's syndrome, characterized by severe keratoconjunctivitis sicca and xerostomia. The potential overlap with inclusion body myositis (IBM) is significant, as both conditions share genetic backgrounds and similar clinical features. Diagnosing such cases is challenging, requiring careful evaluation of both autonomic and myopathic symptoms in Sjögren's syndrome patients.

References

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