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Tremor and Other Hyperkinetic Movements

Case Reports

Athetoid Movements as Initial Manifestation of Primary Sjögren Syndrome

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Abstract

Background: Primary Sjögren syndrome (pSS) is an autoimmune disorder characterized by exocrine gland and extraglandular symptoms. We present a case report of pSS with an initial presentation of athetoid movements.

Case Report: A 74-year-old female presented with a 2-month history of slow undulating movements in her trunk and thighs that eventually spread to her neck and lower extremities. She also reported dry eyes, dry mouth, as well as pain in her shoulders and thighs. Her proinflammatory markers and rheumatologic profile were positive. Her salivary gland biopsy revealed a Focus score > 2. Brain magnetic resonance imaging was normal. A diagnosis of pSS was made. The patient's symptoms improved with hydroxychloroquine, pilocarpine, gabapentin, and clonazepam.

Discussion: Clinicians should consider and screen for primary autoimmune disorders as a cause of subacute athetoid movements in elderly patients. Although aggressive treatment has been recommended, treatment should be tailored to each patient's specific needs.

Keywords: Autoimmune disorders, Sjögren syndrome, neuroimmunology, chorea, athetosis

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Introduction

Primary Sjögren syndrome (pSS) is a systemic autoimmune disorder that impairs exocrine gland function and is associated with extra-glandular manifestations. The central and peripheral nervous system is involved in 15% of cases. The most commonly reported neurological extraglandular manifestations are sensory polyneuropathies. Only 2% of pSS patients have been reported to develop movement disorders.[1] We present a case of pSS that initially presented with athetoid movements.

Case report

A 74-year-old female with a history of hypothyroidism, treated with levothyroxine, presented to the clinic with a 2-month history of involuntary movements. The movements developed slowly over a couple of days. They involved her trunk and thighs, and then spread to her neck and lower extremities over the following weeks. The movements were associated with neck, shoulder, and thigh pain. The movements were functionally and socially impairing with no aggravating or relieving factors reported. Movement did not relieve the associated pain and there was no sense of urgency reported with the movements. She noted the ability to suppress the movements at times. She also reported the presence of dry eyes and mouth over the previous 6–8 weeks. Her physical examination exhibited globally decreased reflexes. Her movements were described as slow, undulating movements of the trunk and legs, which moved and rotated from one side to the other in a non-specific pattern. The movements were only present during rest, either when sitting or lying down and were more intense when lying down. They did not interfere with walking and were absent during sleep (Video 1). The remainder of the neurological examination was unremarkable.

Video 1. **Clinical Characteristics of Involuntary Movements**. In the pre-treatment segment, we can observe the patient at rest and in recumbent supine position, generalized slow undulating writhing movements of the head, trunk, pelvis, arms, and legs. Movements have no rhythm, do not follow a specific pattern, and seem to be flowing from one side to the other. In the post-treatment 6-month follow-up segment, the patient is in recumbent position and sitting position, and no movements are observed.

Toxic/metabolic and infectious causes were ruled out. A magnetic resonance imaging (MRI) brain scan, electroencephalogram, nerve conduction studies (NCS), and electromyography of all four extremities revealed no abnormalities.

A contrasted thoracic–abdominal–pelvic computed tomography scan was normal. Her proinflammatory markers showed highly abnormal acute-phase markers. In addition, her autoimmune profile was positive for antinuclear antibodies (1:640 mottled/spiculated), anti-Ro/SS-A (22.9 U/mL, negative <12 U/mL), rheumatoid factor (immunoglobulin M >200.0 U/mL, negative <6 U/mL), anti-topoisomerase I (29.67 U/mL, negative <32 U/mL), and anticentromere (141.48 U/mL, negative <50 U/mL). A paraneoplastic panel was negative. A Schirmer's test revealed <3 mm and <4 mm of moisture in the left and right eye, respectively. Her salivary gland biopsy showed sialadenitis with a Focus score >2. Lumbar puncture revealed no cells, normal glucose and protein, with positive oligoclonal bands. A diagnosis of pSS was made according to the 2016 American College of Rheumatology/European League Against Rheumatism (ACR/EULAR) criteria. Treatment was started with hydroxychloroquine, 200 mg per day; pilocarpine, 5 mg per day; gabapentin, 300 mg per day; and clonazepam, 0.5 mg per day. At the 6-month follow-up, the patient reported 90% improvement of symptoms. The frequency of athetoid movements decreased substantially to a frequency of one or two events per month.

Discussion

Athetoid movements are rare extraglandular symptoms of pSS. Current data suggest that choreoathetoid movements associated with pSS may present in isolation or in combination with neuropsychiatric symptoms and radiological findings. Evidence also shows that neurologic disease, when present in pSS, can be a strong indicator of disease activity and morbidity. In these cases, early initiation of treatment has contributed to good recovery.[2]

Several theories are available to explain the pathogenesis of choreoathetoid movements associated with autoimmune disorders. It is hypothesized that autoantibodies can 1) induce vasculitic changes in the basal ganglia, 2) inhibit M3 muscarinic receptors in the basal ganglia, or 3) induce a neuroendocrine–immune imbalance causing neuronal damage.[3] All of these chemical changes may cause dysregulation in the function of the basal ganglia.

A small number of cases reporting autoimmune choreoathetoid movements are reported in the literature, which we describe below. The first case reported a 69-year-old male with right limb choreoathetosis. Treatment with steroids was not effective; however, the symptoms resolved after treatment with thioridazine hydrochloride.[4] The second case described a 43-year-old female who subacutely developed chorea and neuropsychiatric symptoms. Her symptoms improved after a 1-month course of prednisone, 40 mg per day, and azathioprine, 50 mg per day.[5] The third report, described an elderly male patient with chorea, personality changes, and bilateral basal ganglia hyperintensities on MRI. His symptoms and radiological findings completely resolved after 2 months of treatment with oral prednisolone, 60 mg/day.[6] The Mayo Clinic's 16-year study of adult-onset autoimmune choreas reported two patients with pSS and two with combined autoimmune disorders. Choreas improved in only one patient who was treated with intravenous methylprednisolone for 5 days.[7]

Our case highlights the importance of considering autoimmune disorders as a cause of subacute athetoid movements in elderly patients. Although aggressive treatment is recommended, treatment should be individualized to each patient's needs. The present case should help physicians raise their clinical suspicion of this disorder in order to identify, and thus implement, early and appropriate support and care.

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