Case Report

Primary Sjögren's syndrome presenting as a generalized Chorea

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Abstract

We report on a patient who presented with generalized chorea as the first manifestation of Sjögren Syndrome, and review the possible pathogenesis.

Keywords: Chorea; Sjögren syndrome; Dyskinesia

1. Introduction

Sjögren's syndrome (SS) is an autoimmune disorder, characterized by a lymphocytic infiltration of the exocrine glands. It can present alone (primary SS) or in association with another autoimmune disease, for example Systemic Lupus Erythematous (SLE, secondary SS).

The case we present is a primary SS (PSS), which first manifested as generalized chorea. PSS rarely affects the central nervous system (CNS) and to our knowledge this is the first report in the literature of chorea as the initial manifestation.

2. Case report

Our patient is a 43 year-old woman with no family history of movement disorders who worked in a public management company. In November 2002, she began to develop progressive generalized choreic movements affecting the axial musculature, including her face and pharynx, which was slightly worse on her left side. Two months later she also developed depressive symptoms and a mild cognitive impairment; this was according to a psychiatrist who prescribed venlafaxine, mirtazapine and clonazepam, there was no improvement in her mood.

As the result of the progression of the choreic movement, the patient had to give up her work and social activities. In February 2003, we assessed her at the movement disorders unit of The Chile University Clinical Hospital. At that initial evaluation she was rated at 10 out of 23 on the Marsden and Schachter Chorea Scale [1]. She was treated with haloperidol in doses from 1 to 10 mg for symptomatic treatment, with little benefit. Haloperidol doses were stable until February 2004.

The neuropsychological assessment included: a learning words test, categorical evoking test (both from the Barcelona battery), Stroop colors and the Wisconsin cards tests. The results demonstrated that the patient was of normal intelligence, with a dysexecutive pattern in concordance with a frontal lobe dysfunction. The cerebral magnetic resonance imaging and transesophageal echocardiogram were normal. Auto-antibody tests, studied with anti-nuclear, anti-neutrophil cytoplasm, anti-DNA and anti-cardiolipin were negative. Plasma protein electrophoresis was normal, rheumatoid factor was negative and viral hepatitis markers were negative. Levels of anti-streptolisin O antibody were within normal range. Genetic testing for Huntington disease was negative.

The immune test performed showed that the Anti Ro and Anti La antibodies were positive at medium levels. A Schirmer test demonstrated xeropthalmia and a salivary gland scintigram showed xeroftomia. PSD diagnosis was

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made according to the American–European Consensus Group criteria [2]. In September 2003, the patient was prescribed prednisone 40 mg per day and azatriopine 50 mg per day. With this treatment regimen her neurological (chorea) and psychiatric (depression) symptoms improved quickly (within a month). At present, the patient has a mild axial chorea (two points in the Marsden and Schachter chorea scale) and is working at her previous job.

3. Discussion

All autoimmune diseases can affect the central or peripheral nervous system [3–6]. The mechanisms involved are not clear at all, but at least two hypotheses have been proposed:

1. An indirect mechanism through cerebral arteries vaculitis, in the CNS, or vasa-nervorum vaculitis in the peripheral nervous system. In the CNS vaculitis, the MRI is abnormal showing ischaemic features, like infarcts involving predominantly the subcortical white matter.
2. A direct mechanism has been described through the formation of anti-neuronal antibodies, without MRI abnormalities [7,8].

Both mechanisms can coexist in the same patient, for example in systemic lupus.

PSS frequently affects the peripheral nervous system [9–11], as in motor and sensitive polyneuropathy. Unusual involvement of the CNS, however, has been reported. In these cases, patients present a transverse myelitis-like syndrome or rarely hemispheric white matter disease, a multiple sclerosis-like syndrome.

Chorea has not been reported as the initial manifestation of PSS until now. In our case there is no family history of movement disorders and no other etiology for the chorea could not be determined. The treatment with haloperidol did not significant affect the severity of the chorea (improve or worsen), which only improved with the initiation of immunosuppressive drugs. Moreover, magnetic resonance imaging was normal. For these reasons, we concluded that PPS was the cause of the chorea, occurring via the formation of anti-neuronal antibody.

Our report emphasizes that autoimmune diseases, and particularly SS, can be a possible etiology of adult onset chorea, even if no other symptoms or features of the disease are present. An autoimmune battery tests including Anti Ro and Anti La antibody, should be added in the evaluation of a patient with an adult onset chorea.

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