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Chorea as the presenting manifestation of primary Sjögren's syndrome in a child

Running title : Chorea revealing Sjögren's syndrome

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Visual diagnosis

A 12-year-old boy presented with subacute chorea (**Video**). He had no personal or familial medical history. Chorea occurred without any history of fever or infection, and rapidly worsened over 3 weeks. Examination was otherwise normal. Brain Magnetic resonance imaging (MRI) and cerebrospinal fluid examination were normal. Diagnosis of chorea associated with a primary Sjögren's syndrome was made according to American-European Consensus Group (AECG) criteria (1), because of the association of xerophthalmia (Schirmer test < 5mm in 5 minutes), positive antinuclear antibodies (1:320) with anti-SSA specificity, and focal lymphocytic infiltration on salivary gland histology (2 infiltrates containing at least 50 lymphocytes in a 4mm² glandular section, focus score =2). Screening for other causes of chorea was negative, in particular anticardiolipin and antistreptococcal biology. There was no other clinical manifestation of Sjögren's syndrome. The patient was treated symptomatically with Tetrabenazine and Valproic acid. Chorea gradually improved within 6 months, until the patient became asymptomatic without any treatment.

The association of chorea and primary Sjögren's syndrome has never been reported in children and is rare in adults. Various central nervous system (CNS) disorders have been linked to Sjögren's syndrome, including movement disorders such as parkinsonism and dystonia (2). Diagnosis can be very challenging, as these symptoms are non-specific and may precede the onset of systemic features of Sjögren's syndrome. The causal association between Sjögren's syndrome and CNS involvement is a subject of debate. In our patient, the association of a remitting subacute chorea with unequivocal Sjögren's syndrome features, and the absence of another cause of chorea support a direct causality. Two mechanisms may account for CNS injury in Sjögren's syndrome: an indirect mechanism involving a vasculitis, and a direct pathogenicity of anti-SSA antibodies. Two adult patients with chorea and Sjögren's syndrome have been previously reported and both improved under steroid regimen (3,4). In our child patient, we decided to postpone steroid therapy, taking into account i) the normality of brain MRI, which was not in favor of vasculitis ii) the potential deleterious effect of this treatment on growth, iii) the possibility of a spontaneous remission in autoimmune choreas. Childhood-onset primary Sjögren's syndrome itself is very rare and usually revealed by an isolated parotid swelling. Recognition and follow-up of this condition is crucial as it may precede the onset of severe autoimmune diseases such as systemic lupus erythematosus or juvenile arthritis.

Subacute chorea can be the initial manifestation of autoimmune diseases. Sjögren's syndrome should be considered in this setting, even in children, and immunosuppressive drugs should be used only in case of symptomatic treatment failure.

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Video legend

The patient has generalized chorea involving the trunk, the four limbs, and the oro-facial area. Abnormal movements occur on a background of hypotonia. Tandem gait is altered due to choreic movements of the legs. As illustrated at the end of the video, chorea impacts activities of daily living, such as feeding or writing.