Central and Peripheral Neurologic During Sjogren's Syndrome

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Abstract

The Sjögren's syndrome is a frequent exocrinopathy autoimmune (0.5% to 3% prevalence). Furthermore exophthalmia and xerostomia, it can be complicated by extra-glandular systemic manifestations. That's including neurological complications, which are rare and little known; they may precede or occur simultaneously with the sicca syndrome. The diagnosis of these neurological manifestations is often difficult with a long diagnostic delay. Neurological manifestations encountered can affect the peripheral or central nervous system, the coexistence of the two is rare. We report a patient case, 51 years old, followed since 2003 for goiter treatment. The diagnosis of syndrome Sjogren primitive landed in 2007 according to the criteria of Vitali. Admitted to the rheumatology department for assessment of impaired general condition, dyspnea stages III according to NYHA, a depressive syndrome, extremity paresthesia, and inflammatory polyarthralgia. Electromyogram four members showed a myogenic syndrome lower limb motor and sensory neuropathy and axonal origin. Muscle biopsy was normal at the boundary of the part taken. Brain magnetic resonance imaging performed before the headache and depressive syndrome has objectified hyper intensities signals of the periventricular white matter and subcortical left. The patient received a course of Rituximab, symptomatic treatment of xerophthalmia and xerostomia.

Keywords: Gougerot sjogren; Neurological complications; Central neuropathy

Introduction

Sjögren's Syndrome (SGS) is a common autoimmune disease. Apart from xerophthalmia and xerostomia, it can be complicated by extra glandular systemic manifestations, including the nervous system. The frequency of this localization is variable according to the studies (10% to 70%), with an average between 15% and 25% [1]. Neurological disorders can be indicative of the disease in 39% of cases. They can affect the Peripheral Nervous System (SNP) or Central Nervous System (CNS), the coexistence of both is rare. We report a new case.

Case Report

Mrs. H.F, 51 years old, It has been followed since 2003 for a goitre under treatment. Sjögren's syndrome was diagnosed in 2007 according to Vitali’s classification and treated by synthetic antimalarials. She was admitted for general deterioration assessment, with NYHA stage III dyspnea, depressive syndrome, paresthesia of extremitis, as well as inflammatory polyarthralgia since one year. In addition, the patient reported chronic otitis media and epigastric episodes complicating the dry syndrome. The physical examination did not objectify a clinical tumor syndrome or parotidomegaly. On the other hand, there were diffuse osteomyalgia, redness of the eye, the tongue removed, and thyroid hypertrophy. In the inflammatory assessment, the blood count was normal, the sedimentation rate at 25 mm/1 st hour, the CRP at 19 mg/L, and a hypergammaglobulinemia at 15 g/l wide and polyclonal at electrophoresis of plasma proteins. The Rheumatoid Factor, C4 and Cryoglobulinemia were without abnormality. The CPK level was normal, while the LDH and Beta 2 micro globulin levels were at 340 IU/l and 2.62 mg/l, respectively. The anti CCP Ab was greater than 320 IU. The rest of the biological assessment, including renal blood and urine, the metabolic balance was normal. Electromyography examination revealed a myogenic syndrome in the lower limb and an axonal sensitivomotrice neuropathy. The neuromuscular biopsy in the territory of the anterior peroneal muscle did not find any anomaly. The exploration of the central nervous system using cerebral magnetic resonance imaging revealed hyper intensities of the left and sub-cortical periventricular white matter (Figure 1). The study of cerebrospinal fluid was normal. Cervical ultrasound revealed an atrophic parotid gland with a 4.8 mm thyroid nodule. Radiography and chest CT showed bronchial dilatation sites and an 8 mm pulmonary nodule. The respiratory functional exploration
found a mixed syndrome with normal DLCO. Fibroscopy revealed diffuse esophageal myositis. The remainder of the visceral assessment, including abdominopelvic ultrasound, was normal. The treatment was a course of Rituximab 1g at 15-days intervals, preceded by the treatment of infectious sites, the stop of corticosteroid therapy, and the symptomatic treatment of xerophthalmia and xerostomia. Apart from the persistent dry syndrome, the patient reported a good clinical response, both neurologically and generally. The biological control was normal. Decline: 2 years.

Discussion

The manifestations of the peripheral nervous system during the SGS are numerous and quite concordant (20% of the patients) [2,3]. Central nervous system damage is less common, more discordant and more recent. One of 63 cases in the Andonopoulos et al. [4] series was recorded. In the study including 400 patient's four cases (i.e. 1% of patients) had this complication [3]. Initially described by Alexander et al. [5] clinical manifestations may be diffuse (cognitive, meningoencephalitis) or focal (medullary, encephalic or optic nerve involvement) [5]. Some may mimic multiple sclerosis. They can precede or occur at the same time as Sjögren’s syndrome [6-8]. The diagnosis of these neurological manifestations is often difficult with a long diagnostic delay.

NPS involvement is more often associated with other systemic manifestations [9-12]. Among these disorders, axonal polyneuropathies, mostly vascular, predominantly distal and symmetrical, and motor signs are generally unobtrusive. EMG is typical with a slow evolution [1,13,14]. Sensory neuropathies, due to lymphocyte infiltration, in the dorsal roots and spinal ganglia, called ganglionopathies, can also be encountered [15]. The involvement of the cranial nerves mainly affects the lower sensory branch of the V nerve, which is often unilateral, isolated or associated with another sensory involvement [1]. Multiple mononeuropathies are rarer. Indeed, the presence of muscle pain without rhabdomyolysis or electromyography or pathological abnormalities is common in the SGS. Also cases of sub acute or chronic polyradiculoneuropathies associated with SGS have been described [16]. Many authors include carpal tunnel syndrome in peripheral involvement despite its frequency in women during the fifth decade [17].

The pathophysiology can be of vascular origin, vasa nervosum or directly via autoimmune cells. Some manifestations are asymptomatic and are only discovered when paracranial explorations are carried out: electroencephalogram, cerebral or spinal MRI, or cerebrospinal fluid study, which are the main explorations associated with the assessment. Immunologic for the diagnosis of neurological complications of the SGC.

There is no therapeutic consensus regarding neurological complications. It can be based on steroids, immunosuppressant’s, reserved for severe forms, as the case of our patient. Plasma exchanges and intravenous immunoglobulins have also been proposed [18].

Conclusion

The neurological manifestations during the Sjögren syndrome are multiple. They rarely affect the central nervous system unlike peripheral damage. Some neurological signs must systematically search for their link with Sjögren’s syndrome. The particularity of our observation is the combined involvement of both the central and peripheral nervous system. Nevertheless, the diagnosis can sometimes be difficult when the attack is asymptomatic.

References