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Case Report

Axonal Myelin Polyneuropathy of the Facial Nerve Revealing a Gougerot Sjogren Syndrome about of a Presentation

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Abstract

Primary Sjögren's syndrome is a chronic autoimmune disease associated with a variety of systemic presentations. Sjögren's syndrome (SS) is one of the most common autoimmune diseases. It can exist as a primary syndrome or as a secondary syndrome when associated with other autoimmune diseases, such as rheumatoid arthritis. Here we report a patient who presented with Sjögren's syndrome, the diagnosis of which was oriented by an axonal impairment of nerve VII on electro neuromyography.

Keywords: axonal myelin; polyneuropathy; gougerot sjogren syndrome

Introduction

Primary Sjögren's syndrome is a chronic autoimmune disease associated with a variety of systemic presentations. [1] Sjögren's Syndrome (SS) is one of the most common autoimmune diseases. It can exist as a primary syndrome or as a secondary syndrome when associated with other autoimmune diseases, such as rheumatoid arthritis, systemic lupus erythematosus, systemic scleroderma, and primary biliary cirrhosis. [2] Nerve involvement is possible but rarely seen in this syndrome (20%). The attack of the cranial pairs is even more rare and predominates especially on the trigeminal [3]

The neurological manifestations of Gougerot-Sjögren syndrome (GSS) are variously appreciated. This is essentially the attack of the peripheral nervous system. [4] Various damage to the cranial nerves has been reported in the context of Gougerot-Sjögren syndrome. Involvement of the V, III and VII nerves have been reported, the most characteristic being nerve V, especially its lower branch. Rare and well-documented cases of facial paralysis have also been described [5]. Here we describe a patient who presented with peripheral facial nerve damage.

Presentation of the patient

Medical observation A 32-year-old patient had consulted the neurology department for a deviation of the mouth and innoclusion of the left eye for which peripheral nerve damage had been diagnosed, . Background: business manager by profession followed since 2017 in internal medicine for inflammatory joint rheumatism. Treated for 6 months for 6 months for lymph node tuberculosis declared cured without histological proof with articular symptoms classified as Poncet's rheumatism. Polyarthritis involving globally symmetrical non-deformed addictive fixed large and small synovial joints The afebrile patient Patient well oriented in time and space left palpebral occlusion impossible and ocular deviation up and out. Erasure of forehead wrinkles and inability to raise the eyebrow. Pockmarks The anamnesis found a dry mouth evolving for about two years. Complete blood count, creatinine, ionogram and blood sugar were normal. Entry elevated C-reactive protein at 98 mg/L quickly normalized. The autoimmune assessment showed positive antinuclear Abs (> 1/1000) with a speckled appearance of anti-SSA and SSB specificity, a strongly positive rheumatoid factor greater than 300 UR/mL (N < 20 UR/mL), whereas the native anti-DNA and polymorphonuclear anti-cytoplasmic antibodies were negative. TSH was normal. HIV, hepatitis A, B, C, syphilis, lyme and Campylobacter jejuni serologies were negative and the profile of EBV, mumps and CMV serologies was not suggestive of a recent infection. The cerebrospinal fluid was clear with the presence of two lymphocytes per cubic millimeter, without hyperproteinorachia. At the EMG, Nerve conduction: - near inexcitability of the left facial nerve - blink reflex: impairment of the trigemino facial axis upstream of the brainstem on the left, Detection: - rest absence of spontaneous activity - Effort: electrical silence on the left facial muscles, - Conclusion: electroneuromyography in favor of left facial involvement with involvement of the trigeminofacial axis. Parotid ultrasound showed aspects of sialectasis and glandular biopsy found lymphoid clusters of variable size compatible with non-specific sialadenitis corresponding to stage 2 of Gougerot's and Sjögren's syndrome. Whole body CT, spinal cord and brain MRI were normal. Treatment corticosteroid therapy by bolus of methylprednisolone 1 g/day for five days, followed by prednisone 1 mg/kg, combined with physiotherapy. The evolution was marked, during the first two months, by a recovery of the facial paralysis).

Discussion

Epidemiology of Sjögren's syndrome in Africa twenty-one hospital studies were included. These studies reported 744 cases of SS. The mean age at diagnosis varied between 28 and 73.6 years, and the proportion of women varied from 83.3% to 100%. There was no population-based incidence or prevalence. Among people with autoimmune and other rheumatic conditions, the frequency of primary SS ranged from 1.9% to 47.6%, while that of secondary SS associated with rheumatoid arthritis ranged from 4 .3% and 100%. Symptoms of dryness were the most common features, with the most commonly affected organs being the joints, lungs, and neurological structures. [6] The main autoantibodies were anti - Ro /SS A antigen, anti-La/SS B antigen and antinuclear antibodies [6].

Several forms of peripheral neuropathy occur in Sjögren's syndrome (sec). Symmetrical sensorimotor polyneuropathy most commonly occurs followed by sensory neuropathy. Pure sensory neuropathy, trigeminal sensory neuropathy, and autonomic neuropathy are also common [7]

Axonal polyneuropathy They generally represent the most common damage and include sensory-motor polyneuropathy and sensory axonal polyneuropathy [8] damage to the peripheral nervous system can be explained by vasculopathy of the vasa nervum. The frequency of lesions of the vasa nervosum suggests that the involvement axonal or secondary to an ischemic phenomenon. Elsewhere, a humoral or cellular immune reaction directed directly against neurons has been evoked. [9]

Vaso nervorum vascular lesions documented by pathological studies are associated with a higher incidence of serum anti-SS-A (Ro) antibodies [10]

Although spinal ganglia involvement may have explained some of the clinical and neurophysiological findings, shows that necrotizing vasculitis was involved in fiber degeneration. All nerve biopsies revealed perivascular inflammatory infiltrates and other vascular abnormalities, which were diagnostic and strongly suggestive of necrotizing vasculitis [11] The most commonly affected cranial nerve is the trigeminal (V) nerve [12] . This damage affects the sensory contingent of the nerve, predominates on the lower branches and is often unilateral. It accounts for about 15% of peripheral damage. Tajima et al. [13] found a higher frequency since they noted 50% trigeminal neuropathy among 21 patients with neurological manifestations. It can be isolated but is often associated with neuropathy, more often with sensory neuropathy [14]. For some authors, it could be due to an infiltration at the Gasser's ganglion [12]. Some attacks are asymptomatic, revealed by a neurophysiological study [15], thus increasing their prevalence (up to 45% of peripheral attacks). Damage to other cranial nerves has been described but appears to be rarer: The facial nerve (VII) is sometimes affected [16] . The first observation of neurological damage during SS is also that of facial diplegia [13]. Damage to the cochleo vestibular nerve (VIII) has also been described [17] and may be responsible for deafness [18]. In the experience [13] involvement of VIII would be more frequent (35% of cranial nerve involvement) than that of V (29%). The oculomotor nerves can also be affected: common ocular motor nerve [19] or even more rarely external ocular nerve [20] . Finally, the mixed nerves can be affected [21]. The possibility of recurrent and multiple paralysis of the cranial nerves during SS must also be emphasized [16]. Treatment of neurological Manifestations There is no consensus on the treatment of neurological complications of SS. Treatment with corticosteroid therapy is generally started [22], whether it is an attack of the PNS or the CNS, except in the case of pauci symptomatic attack. For neuropathies, corticosteroid therapy is more effective in the case of vasculitis [22]; during multineuritis or when there is cryoglobulinemia Immunosuppressants are sometimes used, but are generally reserved for severe pictures. Treatments by plasma exchange or intravenous immunoglobulins have also been proposed. The latter seem effective especially in cases of peripheral damage [23,24] particularly in ataxia-like neuropathies such as ganglionopathies [15].

Conclusion

Systemic pathology characterized by damage to the peripheral nervous system, damage to cranial nerves dominated by damage to the trigeminal. peripheral VII auusi is part of the peripheral attien as a demo, drawn this paper, in front of an attack of the peripheral VII researched the clinical and biological argument in report of a syndrome of gougerot sjogren.

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