



## **PERIPHERAL NEUROPATHY AS FIRST MANIFESTATION OF SJOGREN'S SYNDROME.**

Fernanda Maria Borghi (Universidade Estadual de Maringá, Maringá, PR, Brasil), Lorena Bossoni Miosso Marchiotti (Universidade Estadual de Maringá, Maringá, PR, Brasil), Sueny de Paula Monarin (Universidade Estadual de Maringá, Maringá, PR, Brasil), Pedro Henrique Pelissari (Universidade Estadual de Maringá, Maringá, PR, Brasil), Paulo Roberto Donadio (Universidade Estadual de Maringá, Maringá, PR, Brasil)

### **BACKGROUND**

Primary Sjogren's syndrome (SS) is an autoimmune inflammatory disease, characterized by lymphocytic infiltrates in exocrine glands, resulting in the classic ceratoconjunctivitis sicca (xerophthalmia and xerostomia). However, many patients may present with extraglandular manifestations, such as articular, muscular, pulmonary, renal, cutaneous and neurological. The occurrence of neurological involvement is very variable in the literature, with an average of around 20%, which may precede the syndrome itself.

### **CASE REPORT**

Female patient, black, 54 years old, with previous diagnosis of sickle cell anemia, sought care with complaint of paresthesia and paresis in the upper left and lower right limbs. Initial investigation revealed rheumatoid factor 741 IU / ml (normal <15 IU / ml), ESR 80 mm3, and ANA: 1: 640 fine speckled pattern. With the diagnostic suspicion of Sjögren's Syndrome, new exams were requested. Months later she had complaints of dryness in his eyes and mouth. The exams revealed: ANA 1: 2560 fine speckled pattern, Anti-RO: 254 U/ml; Anti-LA: 52.7 U/ml; Anti-DNA, Anti-Sm, Lupus Anticoagulant, Anti-cardiolipin IgG and IgM, and ANCA-C and P, negatives. Initiated hydroxychloroquine with partial improvement of symptoms. Electroneuromyography of the four limbs showed: sensitive and motor axonal neuropathy with sensory predominance, with important asymmetry in the lower limbs suggesting two types of neuropathy: 1. polyneuropathy; 2. sensory neuropathy (ganglionopathy). Initiated pregabalin, with partial improvement of complaints of paresthesia.

### **CONCLUSION**

improvement of complaints of paresthesia.

Discussion: The prevalence of neurological involvement in SS is very variable, averaging 20%. It can be isolated or preceding the syndrome in months to years. Sensory neuropathy of small fibers is the most prevalent. The electroneuromyography study allows the definition of the subtype sensory ataxia/ganglionopathy, characterized by the involvement of the dorsal root ganglia. Regarding pathophysiology, the main hypothesis is vasculitis, through inflammatory infiltrate in the vascular wall, with destruction of endothelial cells, fibrinoid necrosis, luminal obstruction and ischemia. Treatment is symptomatic, with tricyclic antidepressants, gabapentin, pregabalin, duloxetine and opioids. Glucocorticoids and immunosuppressants may be used in severe cases. Data are conflicting in relation to the increased association with the risk of lymphoma, but one should always be aware of this possibility. There seems to be a worse prognosis with the presence of palpable purpura, vasculitis, c4 consumed and cryoglobulinemia. The authors report this case with the aim of alerting the need for a clinical screening of SS for acute or chronic myelopathy, axonal sensory motor neuropathy or involvement of cranial nerves.