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SENSORY NEURONOPATHY AS SENTINEL SYMPTOM OF PRIMARY SJÖGREN'S SYNDROME: A CASE REPORT

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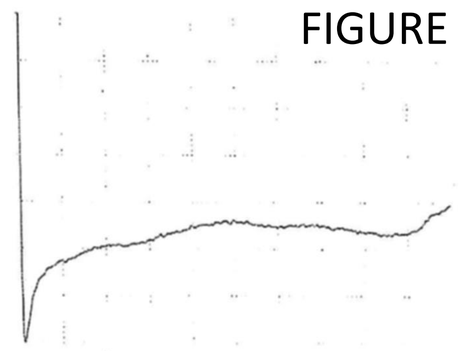
INTRODUCTION: Primary Sjögren's Syndrome (pSS) is a chronic multisystem autoimmune disorder characterized by lymphocytic infiltration of exocrine glands leading to a sicca syndrome. Sensory neuronopathy (SN) could be a complication of pSS mainly with severe impairment of deep sensation and less frequent mild involvement of the motor system. On the other hand, neurological manifestations may precede pSS diagnosis.

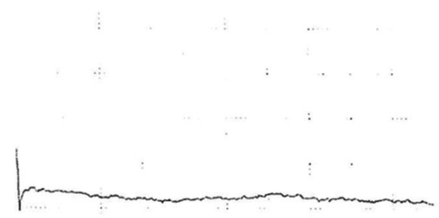
CASE REPORT: A 61 year-old woman came to our attention referring a 10 years history of dysesthesias of the right hand, difficulties of the right upper limb movements and progressive gait abnormalities. She complained a more rapid worsening in the last year limiting daily life activities. One year before hospitalization she received a pSS diagnosis and she began treatment with methotrexate. At the admission to our Neurological Department, she was able to walk only with bilateral support because of severe gait unsteadiness. Neurological examination revealed sensory ataxia with increased difficulty in finger-to-nose test with closed eyes, apallesthesia and pseudoathetosic movements of both arms. Dystonic attitude of the fingers and areflexia were observed, whereas no motor deficits and/or superficial sensitivity abnormalities were found. Electrophysiological study revealed a sensory-motor axonal neuropathy with main involvement of the sensitive component (left median nerve, right ulnar nerve and right superficial peroneal nerve stimulation did not produce any sensory action potentials; figure 1 a-c). Brain and spinal cord MRI were normal. Cerebrospinal fluid (CSF) analysis showed normal cells and protein content and intrathecal IgG synthesis (11 CSF restricted oligoclonal bands). Laboratory tests revealed ANA positivity with nuclear speckled pattern (titer 1:160), anti-SSA 185.8 UA/mL (<10). Cryoglobulins, onconeural and anti-ganglioside antibodies were absent. According to Camdessanché criteria (figure 2), a diagnosis of probable sensory neuronopathy pSS-related was performed and she was treated with periodic intravenous immunoglobulins (ivIg 0.4 gr/Kg/die for 5 days). Both gait disturbance and involuntary pseudoathetosic movements clearly improved during ivIg treatment.

FIGURE 1-a



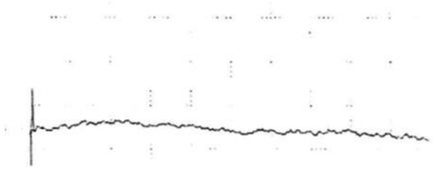
LEFT MEDIAN NERVE

FIGURE 1-b



RIGHT ULNAR NERVE

FIGURE 1-c



RIGHT SUPERFICIAL PERONEAL NERVE

CONCLUSIONS: This case-report confirms that severe SN could be a sentinel symptom of pSS. In our experience, ivIg has been effective in pSS-related SN, in disagreement with other authors that reported a better efficacy of the treatment in patients with pSS-related sensory-motor neuropathy than in patients with pSS-related SN.

A. In a patient with clinically pure sensory neuropathy a diagnosis of sensory neuronopathy is considered as possible if score is >6.5		
	Yes	Points
a. Ataxia in the lower or upper limbs at onset or full development		+3.1
b. Asymmetrical distribution of sensory loss at onset or full development		+1.7
c. Sensory loss not restricted to the lower limbs at full development		+2.0
d. At least 1 SNAP absent or 3 SNAP < 30% of the lower limit of normal in the upper limbs, not explained by an entrapment neuropathy		+2.8
e. Less than 2 nerves with abnormal motor nerve conduction studies in the lower limbs		+3.1
If > 6.5, a diagnosis of sensory neuronopathy is possible	Total score:	
B. A diagnosis of sensory neuronopathy is probable if the patient's score is > 6.5 and if:		
1. The initial work-up does not show biological perturbations or electroneuromyography findings excluding sensory neuronopathy *and*:		
2. The patient has one of the following disorders: onconeural antibodies or a cancer within 5 years, cisplatin treatment, Sjögren's syndrome		
3. Or MRI shows high signal in the posterior column of the spinal cord		
C. A diagnosis of sensory neuronopathy is definite if dorsal root ganglia degeneration is pathologically demonstrated although dorsal root ganglia biopsy is not recommended		

FIGURE 2: Camdessanché criteria for the diagnosis of SN

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