Involvement of Peripheral Nervous System in Primary Sjögren Syndrome. A Gessar Analisys

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Background: Peripheral neuropathy (PN) is usually is a late onset event in primary Sjögren Syndrome (pSS) associated with purpura, cryoglobulinemia, hypocomplementemia and increased risk of lymphoma. Objectives: To describe the frequency of PN in patients with pSS and identify related factors.

Methods: Adult patients in the GESSAR database who met 2002 criteria for pSS. Demographic, clinical, laboratory and electromyogram (EMG) findings were recorded. PN was defined with clinic manifestations and EMG. Other causes of PN were excluded. To compare groups, all patients with PN were included (cases) and a random sample of patients without PN (controls) with a 1:4 ratio was used. Mann-Whitney was used for numeric variables and X2 or Fisher's for categorical. An α of 0.05 was considered significant.

Results: Of 368 patients, 95% were female. Mean age at analysis was 55 y/o (21-87) and 50 y/o (20-89) at diagnosis. The frequency of PN was 11.68% (43/368). Sensory PN was found in 63% (28/43), predominantly small fibers involvement in 41.8% (18/43), axonal PN in 20.9% (9/43) and ataxic in 2.3% (1 / 43). Somatosensory manifestations were found in 37% (16/43) with axonal involvement in 30.2% (13/43) and mononeuritis multiplex in 6.9% (3/43), none had autonomic PN. When comparing groups (43 vs 172 controls) patients with PN had a higher frequency of vasculitis (11.7% vs 1.7%;p=0.002), purpura (23.8% vs 4.7%,p=0.0001), renal tubular acidosis (7.6% vs 1.2%,p=0.020), leucopenia (30.7% vs. 12.1%,p=0.005), low C3 (48.5% vs. 10.3%, p=0.0001) and C4 (66.6% vs. 18.2%,p=0.0001), (+) Anti-Ro/SSA (85.3% vs. 66.6%,p=0.019), (+) RF (72.5% vs. 52.1%,p=0.022), cryoglobulinemia (42.1% vs. 10.9%,p=0.0001) and higher frequency of hypergammaglobulinemia (60.5% vs 44.6%,p=0.09), Raynaud's (27.5% vs 11.6%,p=0.051) and glomerulonephritis (4.6% vs 0.5%,p=0.018), although without statistical significance.

Conclusions: The frequency of PN was 12%, similar to other cohorts. Small fibers and axonal somatosensory PN were the most common. PN was significantly associated with vasculitis, purpura, renal tubular acidosis, cryoglobulinemia, leucopenia, hypocomplementemia and anti-Ro and RF positivity.