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Sjögren's syndrome

ABS0762 FREQUENCY, CHARACTERISTICS AND PREDICTORS OF CENTRAL AND PERIPHERAL NEUROLOGICAL INVOLVEMENT IN SJÖGREN'S DISEASE: DATA FROM PORTRESS, THE PORTUGUESE REGISTRY OF SJÖGREN'S DISEASE

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Abstract

Background:

Sjogren's disease (SjD) systemic affection is being increasingly recognized. Prevalence of peripheral (PNS) and central (CNS) nervous system involvement in SjD and risk factors for these manifestations have not been clearly established.

Objectives:

To define the clinical characteristics of SjD patients with CNS and/or PNS involvement and to find predictors of this involvement.

Methods:

We included patients from the Portuguese registry of SjD (PORTRESS) who had information regarding presence or absence of PNS and/or CNS involvement. Demographic, clinical and treatment data were compared according to parametric or non-parametric tests, as applicable. Predictors of neurological (PNS and/or CNS), PNS (vs. no PNS) and CNS (vs. no CNS) involvement were identified through binomial logistic regression modelling.

Results:

We included 1233 patients, 94.2% female, with a mean age of 61.8±14.3 years and a median disease duration of 11.0 [IQR 11.2] years (Table 1). 65 patients (5.3%) had a history of neurological involvement, 48 (3.9%) with PNS and 20 (1.6%) with CNS affection (Figure 1). Sensorimotor polyneuropathy (20.8%) and sensory polyneuropathy (20.8%) were the most common subtypes of PNS affection. Aseptic meningitis (25.0%) was the most common CNS manifestation. Patients with neurological involvement were more commonly male and had more active disease, translated by higher ESSDAI scores, markers of B cell activity (cryoglobulinemia, low C4, monoclonal gammopathy), and skin, glandular and respiratory involvements (according to ESSDAI definition). On multivariate analysis, male sex (OR 2.77 [1.22-6.31], p=0.015), purpura (OR 2.72 [1.20-6.19], p=0.017), monoclonal gammopathy (OR 2.49 [1.00-6.17], p=0.049) and a higher non-neurological baseline ESSDAI (OR 1.08 [1.02-1.15], p=0.011) were associated with neurological involvement. PNS involvement was associated with older age at symptom onset (OR 1.03 [1.01-1.06], p=0.008), low C4 (OR 3.08 [1.25-7.59], p=0.015), persistent salivary gland swelling (OR 3.98 [1.63-9.76], p=0.003), purpura/cutaneous vasculitis (OR 2.98 [1.10-8.03], p=0.031) and CNS affection (OR 7.32 [1.78-30.16], p=0.006). In turn, CNS disease was only associated with PNS involvement (OR 5.29 [1.10-25.35], p=0.037).

Conclusion:

In the Portuguese cohort of SjD, 5.3% of patients had confirmed neurological involvement. Predictors of these manifestations included male sex, purpura, monoclonal gammopathy and a higher non-neurological baseline ESSDAI. Moreover, a bidirectional association was observed between CNS and PNS affection. In addition, PNS disease was specifically associated with purpura, older age at SjD symptom onset, low C4 and persistent salivary gland swelling.

Table 1 – Demographic and clinical characteristics of SjD patients with and without PNS and/or CNS involvement.

	SjD patients with PNS and/or CNS involvement N=65*	SjD patients without PNS or CNS involvement N=1168**	p
Female	**57 (87.7)**	**110 (96.5)**	**0.029**
Caucasian / White	52 (92.9)	915 (92.9)	1.000
Age at diagnosis	52.7±15.2	52.8±14.5	0.960
ACR/EULAR 2016	41 (64.1)	676 (60.0)	0.517
ANA	57 (90.5)	957 (90.6)	0.969
anti-Ro	51 (81.0)	920 (80.6)	0.939
anti-La	23 (38.3)	489 (44.6)	0.343
Rheumatoid factor	27 (49.1)	485 (48.2)	0.899
Cryoglobulin	**10 (27.8)**	**52 (14.2)**	**0.031**
Low C3	12 (20.0)	177 (17.5)	0.615
Low C4	**10 (16.7)**	**88 (8.7)**	**0.037**
Persistent salivary gland swelling	**9 (14.8)**	**75 (7.2)**	**0.043**
Purpura / cutaneous vasculitis	**10 (16.7)**	**57 (5.5)**	**0.002**
Lymphopenia	16 (25.4)	205 (19.9)	0.288
Monoclonal gammopathy	**8 (13.6)**	**52 (5.3)**	**0.016**
Focus score ≥1	25 (52.1)	405 (48.5)	0.629
Baseline ESSDAI	**4.5 [11.5]**	**2.0 [4.0]**	**<0.001**
Baseline ESSDAI without neurological domains	**3.0 [7.0]**	**2.0 [4.0]**	**0.005**
Baseline ESSPRI	6.0 [5.3]	5.3 [4.66]	0.324

Results presented as mean±standard deviation or median [interquartile range] or n (%)

* Caucasian / white: N=56; age at diagnosis: N=61; ACR/EULAR 2016: N=64; ANA: N=63; anti-Ro: N=63; anti-La: N=60; Rheumatoid factor: N=55; Cryoglobulin: N=36; Low C3: N=60; Low C4: N=60; Persistent salivary gland swelling: N=61; Purpura / cutaneous vasculitis: N=60; Lymphopenia: N=63; Monoclonal gammopathy: N=59; Focus score≥1: N=48; Baseline ESSDAI: N=58; Baseline ESSDAI without neurological domains: N=58; Baseline ESSPRI: N=41

** Caucasian / white: N=985; age at diagnosis: N=1060; ACR/EULAR 2016: N=1127; ANA: N=1056; anti-Ro: N=1142; anti-La: N=1097; Rheumatoid factor: N=1006; Cryoglobulin: N=367; Low C3: N=1014; Low C4: N=1014; Persistent salivary gland swelling: N=1040; Purpura / cutaneous vasculitis: N=1034; Lymphopenia: N=1032; Monoclonal gammopathy: N=988; Focus score≥1: N=835; Baseline ESSDAI: N=927; Baseline ESSDAI without neurological domains: N=927; Baseline ESSPRI: N=541

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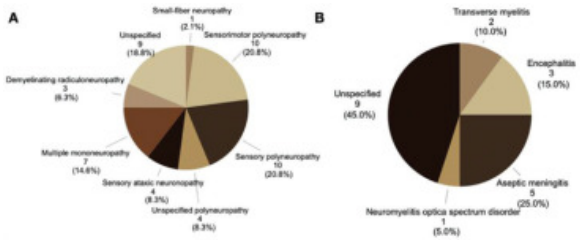

Figure 1 - Frequency distribution of the types of manifestations of the peripheral (A) and central (B) nervous systems.

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REFERENCES:

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Disclosure of interests:

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