ophthalmoplegia is provoked by intra-axial lesions within the higher part of the cerebral trunk, above mesencephalic oculomotor centres; however, a supra-nuclear lesion during pSS has never been reported so far.

Through a literature review, we discuss relevance, aetiology and treatment of CNS involvement in pSS. Data are imprecise and even contradictory. Frequency of CNS involvement in pSS could vary from 0% to 25% and might display various patterns: focal or multi focal lesions entailing encephalon, cerebral trunk, cerebellum or medulla, but also more diffuse processes as an aseptic meningo-encephalitis or as a cognitive impairment.

It's pathogenesis is poorly understood. Two mechanisms have been proposed: the first being of an ischaemic nature resulting from a vasculitis of small brain vessels, the second through an autoimmune response (humoral by anti-neuronal antibodies or cytotoxic).

There is no clear consensus concerning the treatment of neurological complications of pSS. Corticosteroids are usually initiated first; immuno-suppressive agents may be necessary in the absence of improvement.

Conclusion A supranuclear ophthalmoplegia can be observed in pSS; it reinforces the thesis of a CNS involvement in this connective disease, in so far such a characteristic symptom cannot be explained by a Steele-Richardson disease, a multiple sclerosis or vascular lesions. Be aware of CNS involvement in pSS would lead to realise a minor salivary gland biopsy that is the only exam useful to establish the diagnosis.

AB0119 PRIMARY SJOGREN'S SYNDROME ASSOCIATED WITH ANTERIOR HORN CELL DISORDER

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Background Central nervous system (CNS) involvement in primary Sjogren's syndrome (pSS) is controversial with regard to incidence, aetiology and pathogenesis. In the literature, the incidence of CNS-SS varies widely, ranging from rare to incidence rates of 20% to 25%. The clinical picture is diverse, ranging from mild cognitive symptoms to fatal cerebrovascular accidents. **Objectives** We describe a patient with pSS presenting with dysphagia and progressive respiratory failure resulting from anterior horn cell disorder. To the best of our knowledge this is the first case reported in medical literature. The disease responded to corticosteroids and hydroxychloroquine treatment.

Methods A 58-year-old woman had a one year history of dry eyes and dry mouth. She experienced progressive dysphagia and dyspnea ten days prior to admission. She developed progressive respiratory muscle weakness which resulted in respiratory failure and received mechanical ventilation during admission. Examination revealed symmetrically decreased muscle power without sensory impairment and positive Schirmer test. Blood investigations revealed positive ANA, anti-SS-A and elevated IgG level up to 2820 mg/dl. Full blood cell count and creatine kinase level were normal. Sjogren's syndrome was confirmed by minor salivary gland biopsy. Electromyography, nerve conduction velocity and evoked potential studies showed anterior horn cell disorder. MRI of brain revealed negative finding. The disease improved with administration of corticosteroids and hydroxychloroquine.

Results

Conclusion Sjogren's syndrome should be considered in the differential diagnosis of patients with motor neuron diseases. Prompt diagnosis and treatment are required to prevent further morbidity and mortality.

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AB0120 PRIMARY SJOGREN'S SYNDROME ASSOCIATED WITH ANTERIOR HORN CELL DISORDER

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Objectives We describe a patient with pSS presenting with dysphagia and progressive respiratory failure resulting from anterior horn cell disorder. The disease responded to corticosteroids, hydroxychloroquine and azathioprine treatment. To our knowledge this is the first case reported in the medical literature.

Methods A 58-year-old woman had a one year history of dry eyes and dry mouth. She experienced progressive dysphagia and dyspnea ten days prior to admission. She developed progressive respiratory muscle weakness which resulted in respiratory failure and received mechanical ventilation during admission. Examination revealed symmetrically decreased muscle power without sensory impairment. Schirmer test was positive. Blood investigations revealed positive ANA, anti-SS-A and elevated IgG level up to 2820 mg/dl. Full blood cell count and creatine kinase level were normal. Sjogren's syndrome was proved by minor salivary gland biopsy. Electromyography, nerve conduction velocity and evoked potentials studies showed anterior horn cell disorder. MRI of brain revealed negative finding. The disease improved with administration of corticosteroids, hydroxychloroquine and azathioprine.

Results

Conclusion Sjogren's syndrome should be considered in the differential diagnosis of patients with motor neuron diseases. Prompt diagnosis and treatment prevent further morbidity and mortality.

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AB0121 CYTOSTATICS EFFECT ON THE DEVELOPMENT OF MALIGNANT AND LYMPHATIC PROLIFERATIVE FORMATIONS DURING SJÖGREN'S DISEASE (SS)

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Background Treatment of rheumatological pts by alkylating cytostatics is undoubtedly of positive effect on the process of the disease but in a long term can stimulate the development of neoplasms and lymphatic proliferative diseases.

Objectives To detailise the degree of risk of above mentioned pathologies in SS pts who for a long time took low doses of chlorobutin or cyclophosphane – from 5 to 15 years.

Methods In the 1st group of 300 pts aged 15–75 years (46.4 (1.7) with disease duration of 5–25 years (8.7(1.9) 200 pts took 5 mg of Prednisolone daily, 1.8 g for a year, 0.9–1.8 g for the subsequent years combined with chlorobutin by 2 mg daily; 720 mg for a year and 360–720 mg for the next years. 100 pts had cyclophosphane by 200 mg twice a month i.v.; 4.8 g for a year and in the next 5 years yearly dosage was 2.4–4.8 g. 2nd group included 100 female pts who took only Prednisolone according the same scheme. 3d group of 100 female pts had no antirheumatic therapy. Age of pts in 2nd and 3d groups was 18–78 years (46.2(2.3) and disease duration of 4–26 years (9.2(2.1). Minimal frequency of blastomas and lymphatic proliferative diseases (1.6%) development was in the 1st group (pseudolymphoma in 1 pt, lymphoma – 2 cases, cancer of stomach in 1 and of skin in 1 pt). In the group treated only by GCS 7 cases of lymphoma were found and 1 case of stomach cancer (8%). Pts who had no antirheumatic therapy had 7 pseudolymphomas 9 lymphomas, 2 cases of Valdenstrem crioglobulinemia, 2 cases of stomach cancer and 1 pulmonary cancer (21%). Out of them pseudolymphomas and lymphomas were diagnosed in 18%.

Results

Conclusion Long term of taking of low doses of cytostatics (chlorobutin, cyclophosphane) combined with low doses of GCS does not promote the development of blastoma process in SS pts and decreases the frequency of transformation of the disease in malignant lymphomas.

FRI0212 CORRELATION BETWEEN HISTOPATHOLOGICAL FINDINGS, SERO-IMMUNOLOGICAL PROFILE AND MARKERS OF DISEASE ACTIVITY IN SICCA SYNDROME

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Background We evaluated sero-immunological parameters and markers of disease activity in ninety-two patients all female who underwent lip biopsy for histological assessment of sicca syndrome, according new preliminary criteria proposed for classification of Sjogren Syndrome (S. S).

Objectives The aim of our study was to compare the histopathological score index according to Chisolm and Mason classification with twenty-two parameters of disease activity.

Methods We have classified thirty-seven patients (group A) with focus score I-II with probable S. S (positivity of 3/6 item) and

fifty-five patients (group B) with focus score III-IV with defined S. S (positivity of 4/6 item). The mean age of two group was: group A 60 ± 11 years, group B 64 ± 9.8 years. The statistical comparison between the two group was performed with T-Student for 20 parameters and chi-square method for ANA and SSA/SSB positivity.

Results The comparison between the two groups show statistically significant differences for sero-immunological parameters such as gamma globulin ($p < 0.01$), SSA/SSB ($p < 0.001$). Moreover the group B shows higher levels of ESR ($p < 0.005$), fibrinogen ($p < 0.02$) and alpha1-antitrypsin ($p < 0.005$).

Conclusion This data show the important role of lip biopsy in the diagnosis of S:S and suggest the possible correlation between high histopathologic focus score and immunological markers of disease activity.

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FRI0213 RAYNAUD'S PHENOMENON IN PRIMARY SJÖGREN'S SYNDROME. PREVALENCE AND CLINICAL CHARACTERISTICS IN A SERIES OF 320 PATIENTS

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Background

Objectives To determine the prevalence of Raynaud's phenomenon (RP) in a large series of patients with primary Sjögren's syndrome (SS) and to identify the clinical and immunological features related to its presence.

Methods In a cross-sectional study, we investigated 320 consecutive patients with primary SS (294 women and 26 men; mean age at onset 60 years; range 16–87 years). All patients fulfilled 4 or more of the diagnostic criteria for SS proposed by the European Community Study Group in 1993. Diagnosis of RP was done in SS patients with intermittent attacks of digital pallor and/or cyanosis, in the absence of any other associated disease or anatomical abnormalities.

Results Raynaud's phenomenon (RP) was present in 40 (13%) patients. All were women, with a mean age of 57 years (range, 18 to 78 years). RP precedes onset of sicca symptomatology in 18 (45%) patients. The main triggering factor was cold exposure, which induced RP in all patients, emotional stress in 12 and 2 patients had labour predisposition. Fifteen (38%) patients required pharmacological treatment with calcium channel blockers (12 patients) or angiotensin-converting enzyme inhibitors (2 patients) in the cold seasons. One patient required treatment with intravenous prostacyclin for ischaemic complications. When compared with SS patients without RP, those with RP showed a higher prevalence of articular involvement (50% vs 31%, $p = 0.031$), cutaneous vasculitis (30% vs 11%, $p = 0.003$), antinuclear antibodies (95% vs 65%, $p < 0.001$), anti-Ro/SS-A (59% vs 31%, $p < 0.001$) and anti-La/SS-B antibodies (44% vs 20%, $p = 0.003$).

Conclusion We found RP in a 13% of patients with primary SS. Interestingly, RP was the first autoimmune symptomatology in almost 50% of these patients. These patients constituted a subset of SS with a higher frequency of some extraglandular features and positive immunological markers. In contrast, RP have a