FRI0230 CLINICAL PRESENTATION OF 475 PATIENTS WITH PRIMARY SJÖGREN'S SYNDROME WHO WERE SEEN BY THE RHEUMATOLOGISTS

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Background

Objectives Study the clinical presentation of primary Sjögren's syndrome (pSS) patients in a rheumatology unit.

Methods 475 patients with pSS have been studied. European criteria was used for the diagnostic. Patients with primary biliary cirrhosis and hepatitis C were excluded. Demographic, clinical and laboratory data were taken following the protocol of the unit. Minor salivary biopsy was carried out on 90% of the patients.

Results 94.5% were women. Age was 63 ± 12 . Years of evolution were 10 ± 13 . More prevalent extraglandular manifestations were: Carpal tunnel's syndrome (CTS), 40%; fibromyalgia, 36%; intermittent non inflammatory arthritis, 36%; inflammatory osteoarthritis, 10%; thyroid's dysfunction, 25.4%; photosensitivity, 20%; peripheral neurologic manifestations without CTS, 5%; central neurologic manifestations, 5%; haematological manifestations, 8; renal manifestations, 5%; vasculitis, 13%.

Rheumatoid factor (RF) was positive in 50% of the patients, ANA in 70%, antithyroidal antibodies in 27%, SS-A/Ro in 23% and SS-B/La in 11%.

37% of them come because of rheumatic manifestations, (14% fibromyalgia, 14% arthritis, 5% inflammatory osteoarthritis, 4% Raynaud's phenomenon, another 5% are arthralgia and low back pain.) 30.5% come because of oral or ocular dryness and parotidomegalia. Other reasons for consultation were: autoimmune cytopenia (5%), thyroidal dysfunctions (3%), vasculitis (2%), and RF or ANA (2%).

Conclusion The most common reason for consultation were rheumatic manifestations. The second reason was the oral and ocular dryness. Analytical alterations (Cytopenias, RF, ANA) can be other reasons for consultation.

FRI0231 ASSOCIATION OF SYSTEMIC SCLEROSIS WITH PULMONARY SARCOIDOSIS IN TWO CASES

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Background

Objectives

Methods A 42-year-old man has been followed up due to mediastinal lymphadenopathy and pleural effusion parallel with Raynaud syndrome, sclerodactylia and bibasal pulmonary fibrosis. He was diagnosed as having systemic sclerosis by the clinical symptoms and skin biopsy and sarcoidosis which was confirmed by the biopsy of mediastinal lymph nodes. A 24-year-old woman has been followed up due to sarcoidosis proved by the biopsy of mediastinal lymph nodes, too) for 3 years before the development of the clinical symptoms of systemic sclerosis.

Results In both patients sarcoidosis were successfully treated with corticosteroids (in the second case corticosteroid treatment was completed with cyclophosphamide because of fibrotising alveolitis). We analysed the clinical symptoms, organ manifestation, prognosis in both cases together with the examination of

cytokine profile (interferon- γ , IL-1, IL-2, IL-4, IL-6, TNF- α) of CD4 positive lymphocytes from peripheral blood.

Conclusion Our results showed Th1 dominance in both cases, which may play an important role in the coexistence of sarcoidosis and systemic sclerosis.

OP0065 ABNORMALITIES IN THE PKC SIGNALLING SYSTEM DOWNSTREAM TO MUSCARINIC RECEPTORS IN AN EXPERIMENTAL MODEL FOR SJÖGREN'S SYNDROME (SS)

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Background Postganglionic parasympathetic nerve fibers¹ release acetylcholine, which binds to acinar cell muscarinic receptors. Short-term stimulation leads to activation, but long-term stimulation leads to a negative feedback in form of proteolytic degradation. Salivary glands acinar cells in SS patients lack PKC β II and are low in PKC α isoform² as if they had been consumed/proteolytically degraded.

Objectives Our aim was to study, if also NOD mice have PKC involvement, which isoforms are involved and if this involvement is of the early activation type or like is seen as a result of a long-term, autoantibody-mediated stimulation in human disease.

Methods Conventional (α , β , γ), unusual (δ , ϵ , θ , ι) and atypical (λ , ζ) PKC isoforms were studied using immunohistochemical staining and Western blotting.

Results Control BALB/c mice had PKC α and β like their human counterparts. They require calcium, DAG and phosphatidylserine for their activation. In contrast, NOD mice with sialadenitis had a strong expression of PKC α and β . Transgenic NOD x IFN- γ mice had an early involvement, but similar pattern to that of NOD mice. Acinar cells in NOD scid mice did not express any of the PKC isoforms studied.

Conclusion Findings suggest that the calcium-dependent conventional PKC isoforms are involved also in NOD mice. In contrast to the human disease, mice with a short life span display upregulation and do not reach the stage with proteolytic degradation/loss of PKC. Interestingly, the altered PKC status modulates the susceptibility of cells to apoptosis.

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OP0066 EFFECTS OF TNF BLOCKADE WITH INFlixIMAB IN PATIENTS WITH PRIMARY SJÖGREN'S SYNDROME: AN OPEN PILOT STUDY

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Background Tumour necrosis factor α (TNF α) is a proinflammatory cytokine involved in the pathogenesis of Sjögren's syndrome (SS), and blockade of TNF α may reduce the activity of the disease.