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AB1048 RHUPUS SYNDROME IN A TERTIARY HOSPITAL

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Background: Rhupus syndrome (RhS) is a rare combination of rheumatoid arthritis (RA) and systemic lupus erythematosus (SLE). Different studies describe RhS cases that begin with erosive arthritis and the presence of rheumatoid factor (RF) and/or anti CCP and then the SLE symptoms.

Objectives: Despite the fact that RhS shows a low prevalence, it would be useful to know clinical characteristics of RhS patients since their therapy and outcome differ from those having RA or SLE alone.

Methods: Retrospective study with systematic revision of electronic clinical records of RhS patients was performed. Demographic, clinical and immunological data were collected.

Results: Eight RhS patients were included (all fulfilled SLICC 2012 criteria for SLE and ACR 2010 for RA). Mean age was 67.3 (45-84) years (7 were female). In 3 cases RA was the first diagnosis with a mean evolution of 4.5 years until SLE diagnosis. In contrast, in 5 cases SLE was the first diagnosis with a mean evolution of 7.2 years until RA diagnosis. Photosensitivity and arthritis were the predominant clinical manifestations. One patient presents pericarditis and other case showed rheumatoid nodules in elbows. Renal, pulmonary or neurological affection was no reported.

4 patients were under biological/JAK inhibitors therapies (2 abatacept, 1 rituximab and 1 baricitinib) with favorable response of treatment.

Conclusion: In contrast to other series, only the 37.5% of our RhS cases begins with polyarticular seropositive arthritis. The 62.5% started with SLE symptoms as haematological alterations, cutaneous and serological manifestation, and showed longer progression to have polyarticular affection. Thus, RhS diagnosis is earlier in patients that begin with RA symptoms. 4 RhS patients were refractory to DMARD treatments, where biological/JAK inhibitors therapies are needed.

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AB1049 CLINICAL SPECTRUM AND THERAPEUTIC MANAGEMENT OF AUTO-IMMUNE MYELOFIBROSIS: A NATION-WIDE STUDY OF 30 CASES

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Background: Little is known about autoimmune myelofibrosis (AIMF), a rare entity that can occur alone or in association with another autoimmune disease (AID) and is responsible for bone marrow (BM) failure and life-threatening complications.

Objectives: We conducted a nationwide retrospective observational study of AIMF cases to better characterize the epidemiology, clinical presentation and evolution of this rare entity.

Methods: The aim of the study was to analyze the characteristics of AIMF and the nature and indication of treatments currently used. Response to treatment was evaluated by the revised Tefferi et al. response criteria.

Results: Among 30 cases of AIMF, the sex ratio (F/M) was 4:1 and the median age at diagnosis was 37 years (interquartile range 30–49). AIMF was diagnosed after the onset of an associated AID in 12 cases and concomitant to an AID in the remaining 18 cases. The most frequently associated AID was systemic lupus erythematosus, followed by Sjögren syndrome. All cases consisted of reticulin fibers, and no collagen fibrosis was described. More than 50% of cases showed complete response after first-line therapy, with glucocorticoids (GC) in 28 cases. Half of the cases had treatment complications mainly related to GC therapy.

Conclusion: Diagnosis of AIMF remains challenging in the absence of a validated set of diagnosis criteria, and must always be searched in the presence of hematological abnormalities at onset or during follow-up of AID. Clinical context, search for mutations and pathology findings can help differentiating this rare disease from a clonal pathology. GC is currently an effective first-choice therapy for AIMF, but a high rate of GC dependency and long-term complications indicate the need to find new sparing drugs.

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AB1050 CLINICAL IMPLICATIONS OF ULTRASONOGRAPHY (US) IN DIAGNOSIS AND MONITORING DISEASE ACTIVITY OF RELAPSING POLYCHONDRIITIS (RP) AND COMPARATIVE INVESTIGATION BY US BETWEEN AURICLE OF RP, REPEATED TRAUMA, CELLULITIS AND HEALTHY SUBJECT

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Objectives: To assess the clinical implications of ultrasonography (US) in monitoring disease activity and diagnosis of relapsing polychondritis (RP).

Methods: Firstly, auricular (n=5) and nasal (n=1) chondritis of six patients with RP were assessed by US before and after treatments. The relationship between US findings and serum markers were evaluated. Moreover, the comparisons of US findings between the auricle of patients with RP (n=5), repeated trauma (n=5), cellulitis (n=2) and healthy subjects (n=5) were also assessed.

Results: US finding before treatment showed low-echoic swollen auricular and nasal cartilage with increased power Doppler signals (PDS) in all cases of RP. US findings corresponded to biopsy findings. After treatment, the swollen ear and nose completely resolved. Then, US findings also showed dramatic reductions in swollen cartilage with the decrease in PDS. Although serum markers completely improved, US finding remained in 1 of 6 cases, and this case showed flare due to PSL tapering. Finally, RP could be differentiated from the damage of repeated trauma and cellulitis by the presence or absence of PDS and subperichondrial serous effusion.

Conclusion: US of auricular and nasal cartilage in RP possibly facilitates evaluation of auricular lesions and monitoring of disease activity, especially when we consider the treatment response and the timing of drug tapering.

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AB1051 KIKUCHI FUJIMOTO DISEASE, IS IT SLE?

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