SECONDARY HEMOPHAGOCYTIC SYNDROME: RETROSPECTIVE STUDY ACCORDING TO THE UNDERLYING DISEASE

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Background: Secondary Hemophagocytic Syndrome (SHS) is associated with hematologic (HO), autoimmune (AI) and solid tumors (Tum) and it has a poor prognosis. The presence of uveitis is a predictor of mortality. We aim to analyze patients with SHS and determine variables that predict mortality.

Objectives: To determine specific features that predict mortality in SHS patients.

Methods: Retrospective analysis of patients diagnosed with SHS identified at the NCI and our hospital from 2005 to 2018. The primary outcome of interest was mortality. The variables analyzed were: sex, age, diagnosis of the underlying disease, fever, organomegaly, laboratory findings, days of hospital stay, days from admission to B.M.O. and mortality.

Results: A total of 27 patients were found. Table 1 shows the characteristics of the groups. AI diseases found were: 5 SLE, 2 AS, 1 Rheumatoid Arthritis and 1 Serositis. The clinical presentation of cases was reported in 205 patients for osteoarticular manifestations. 5 patients were female, age 67 years on average, with fever (55.5%) and organomegaly (55.5%). The most frequent diagnosis was lymphoma (33.3%), followed by leukemia (22.2%). The average number of days of hospital stay was 17 days. The average number of days from admission to B.M.O. was 5 days.

Conclusions: Patients with SHS to HO disease had a high mortality and a longer hospital stay compared to the rest of the groups. Practically all patients met all diagnostic criteria, the most frequent were fever, pancytopenia and hyperferritinemia.

Disclosure of Interest: None declared


SAT0605

THE PRESENCE OF UEVITIS PREDICTS THE RESPONSE TO THE INTERLEUKIN (IL)-1 INHIBITORS ANAKINRA AND CANAKINUMAB IN BEHÇET’S DISEASE

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Background: In recent times IL-1 inhibition has been proposed as an intriguing therapeutic option in Behçet’s disease (BD) patients with multi-drug resistant manifestations. However, despite the good clinical results obtained during the last years, cases of BD nonresponsive to anakinra (ANA) and canakinumab (CAN) have also been identified. This evidence has induced to suspect the existence of specific subsets of patients characterised by a more pronounced IL-1 driven pathogenesis.

Objectives: To identify predictive factors of response to interleukin (IL)-1 inhibitors among demographic, clinical and therapeutic data in patients with BD.

Methods: BD patients treated with ANA or CAN were enrolled. Patients were divided into 2 groups according to the clinical response: group 1 included subjects showing a treatment duration of at least 52 weeks and no secondary inefficacy during the first follow-up year; the remaining patients were included in the group 2. Demographic, clinical and therapeutic data were analysed to identify significant differences between groups.

Results: Eighteen patients (50%) were included in group 1 and 18 (50%) in group 2. A better response to IL-1 inhibitors was significantly more common among patients with BD-related uveitis (p<0.006) and patients with a longer disease duration (p<0.03).

Conclusions: IL-1 blockade is effective in BD, especially in the subset of patients presenting ocular involvement and in those with long-lasting disease.

REFERENCES:

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SAT0606

DISEASE MODIFYING ANTI RHEUMATIC DRUGS IN THE TREATMENT OF SAPHO SYNDROME: SYSTEMATIC LITERATURE ANALYSIS

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Background: SAPHO (Synovitis Acne Pustulosis Hyperostosis Osteitis) Syndrome is a rare, heterogeneous clinical entity with cutaneous and osteoarticular expression. The therapeutic management is not codified and there is no validated evaluation tool for SAPHO syndrome.

Objectives: To perform a systematic analysis of the literature in order to evaluate the effects of DMARDs in SAPHO syndrome.

Methods: Biphosphonates, conventional and targeted synthetic DMARDs, anti-TNF alpha, and other biologics have been subjected to advanced Pubmed research. Treatment was considered effective when the patient validated the response criteria defined in the study or if at least partial benefit was obtained for a minimum of three months. The different treatments were ranked according to their effectiveness rate in three interest groups and then grouped by therapeutic class to determine an overall response rate. These rates led to the calculation of an efficacy index weighted by the number of patients treated in the subgroup (molecule or therapeutic class) to provide the total number of patients in our study.

Results: Treatment efficacy was evaluable in 284 of the 292 patients analysed. The clinical presentation of cases was reported in 205 patients for osteoarticular