Objectives: To evaluate in the real clinical practice the frequency of prescribing, efficacy and safety of IL-1 therapy in pts with mAIDS and sJIA according to the Federal Rheumatology Center.

Methods: A retrospective study from 2013 to 2021 included 66 pts who were prescribed IL-1. Among them 45 pts with mAIDS: FMF - 8, CAPS - 25, TRAPS - 10, HIDS/MKD - 2, 21 pts had sJIA. Canakinumab (CAN) was administered subcutaneously at the dose of 2-5mg/kg or 150mg every 4-8 weeks, anakinra (ANA) - subcutaneously at the dose of 1-5mg/kg or 100mg daily.

Results: Among pts with mAIDS 45 received IL-1. The age of pts ranged from 1.5 to 44 years, the median (Me) age was 5.9 [6.9; 17] years. Female pts predominated (60%). The median age at onset was 0.5 [0; 4] years (0 - 35). The median duration of the disease was 7.5 [3; 9; 15] years (3 months to 44 years). 35 pts received CAN, 10 - ANA. Both drugs showed significant positive dynamics with a complete response in 40 (88.9%), a partial response in 5 (11.1%), mainly due to serious neurological and cognitive impairments in 2 pts with CANOMID, sensorineural hearing loss in 3 adult pts with MWS, as well as amyloidosis in 1. In 7 pts with CAPS who received ANA as first biologic treatment, after achieving a reliable positive response, switching to CAN was performed while maintaining full efficacy. It was possible to discontinue the glucocorticoids (GC) in all pts. In 2 pts with CANICA/NOMID (1), MWS (1) due to insufficient efficiency the interval between injections of CAN was reduced from 8 to 4 weeks. The duration of use of CAN in pts with mAIDS ranged from 6 months to 12 years, ANA – 2 months to 12 years, 13 pts (28.9%) have been receiving therapy for 5 years or more. The age of pts with sJIA ranged from 3 to 17 years, Me 8.15 [5.3; 12.7] years. The age of the onset varied from 4 months to 12 years, Me 3.1 [8.5; 5.7] years. Female pts predominated (61.9%), 20 pts received CAN (18 after secondary inefficiency/infusion reaction of tociluzumab (TCZ), 2 had previous experience of 3 B. TCZ-etanercept-adalimumab, TCZ-sabatacept-adalimumab). 1 patient received ANA. The duration of CAN treatment was from 5 months to 4.5 years. Among pts with sJIA who received CAN, secondary inefficiency with discontinuation was observed in 6 (28.6%) (10-25 months after treatment initiation), and in 1 patient who received anakinra for 8 months. The tolerability of therapy was generally good. We have not observed any SAE in the treatment of IL-1. All pts are continuing taking medications.

Discussion of Interests: Svetlana Salungia Speakers bureau: Novartis, Sobi, Maria Kalea Salungia Speakers bureau: Pfizer, Roche, Novartis, Evgeny Fedorov Speakers bureau: Novartis, Sobi, MEDAC, Irina Nikishina Speakers bureau: Pfizer, MSD, Roche, Sobi, Anna Torgashina Speakers bureau: Novartis DOI: 10.1136/annrheumdis-2022-eular.3195

AB1298

AL-AMYLOIDOSIS MIMICKING IgG4-RELATED DISEASE

B. Chalcev1, A. Torgashina1, E. Sokol1, J. Khvan1 on behalf of Laboratory of Rare Rheumatic Diseases and Primary Sjögren’s Syndrome. 1V.A. Nasonova Research, Laboratory of Rare Rheumatic Diseases and Primary Sjögren’s Syndrome, Moscow, Russian Federation

Background: Hematological diseases such as multiple myeloma (MM) and POEMS-syndrome can be accompanied by high serum IgG4 level and thus mimic an IgG4-related disease (IgG4-RD) [1, 2]. However, there are no descriptions of AL-amyloidosis with increased secretion of IgG4 in the literature.

Objectives: to present a clinical case of AL-amyloidosis mimicking IgG4-RD.

Methods: At the age of 42, patient S. developed nasal congestion, and 2 years later bilateral symmetrical swelling of submandibular salivary glands developed. Chest CT scan revealed hydrothorax and pericarditis, interstitial changes in both lungs and intrathoracic lymphadenopathy. According to blood tests, ANF, RF and CRP were within normal values, an increase in the serum IgG4 (32.9 g/l) and IgG (34.1 g/l) levels was detected, and the patient was admitted to our clinic with suspicion of IgG4-RD.

Results: On examination, we noticed a symmetrical enlargement of submandibular salivary glands and macroalgosis. The patient also complained of tingling in the fingers, performed electromyography and revealed bilateral carpal tunnel syndrome. According to the immunofixation of blood and urine proteins, IgG4-lambda paraproteinaemia (18.3 g/l) and a trace amount of Bence-Jones-lambda protein in the urine were detected. AL-amyloidosis was suspected and a biopsy of the submandibular salivary gland was performed, followed by Congo red staining and darkfield microscopy that confirmed amyloidosis (Figure 1). There were no signs of IgG4-RD (storiform fibrosis or obliterator phlebitis) in the biopsy specimens. Echocardiography revealed thickening of the interventricular septum and hypertrophy of the ventricular myocardium, MM and other malignancies were excluded on the basis of bone marrow trephine biopsy and PET-CT (also revealed severe hepatomegaly). The diagnosis of IgG4-secreting AL-amyloidosis affecting salivary glands, tongue, heart, lungs, liver, nervous system was made and polychemotherapy was started, the patient’s further life remained unknown.

REFERENCES:

Disclosure of Interests: None declared

Figure 1. Congo red staining and darkfield microscopy demonstrating amyloid deposits.

AB1297

EVALUATION OF POSSIBLE RISK FACTOR OF PULMONARY EMBOLISM IN SARCOIDOSIS PATIENTS

B. Ruan1, P. Contalsoneri1, C. Torregiani1, S. Picherei2, M. Contalsoneri1, F. Salton1. 1Department of Pulmonology, University of Trieste, Trieste, Italy; 2Department of Pulmonology, University Hospital of Cattinara, Trieste, Italy

Background: Some studies reported a correlation between sarcoidosis and an increased risk of pulmonary embolism (PE) [1, 2].

Objectives: To assess possible risk factors of PE in sarcoidosis patients.

Methods: We enrolled 260 sarcoidosis patients (170 females and 90 males; mean age at diagnosis 46 ± 9), after giving written informed consent. We performed clinical evaluations, laboratory tests and radiology features.

Results: Our study population included 20 sarcoidosis patients with PE (14 females and 6 males; mean age at diagnosis 45 ± 10), diagnosed by lung scintigraphy and 240 sarcoidosis patients without PE (170 females and 70 males; mean age at diagnosis 46 ± 11). There was a significant increase of the presence of antiphospholipid antibodies in the sarcoidosis group with pulmonary embolism (55%) than in group without PE (9%) (p-value<0.01). There was no statistically significant difference between the two groups on smoking habit, obesity, treatments and hereditary thrombophilia frequency (p > 0.05, respectively).

Conclusion: This study demonstrated an increased of antiphospholipid antibodies positivity in sarcoidosis patients with pulmonary embolism. Furthermore, we propose screening for these antibodies in all sarcoidosis patients.

REFERENCES:

Disclosure of Interests: None declared