ACR30/50/70/90 responses in the anakinra group were sustained throughout the study period. Patients in the anakinra group had a prompt and persistent decrease in CRP and ferritin levels at Week 1, which was not observed in the placebo group. There were no unexpected adverse findings. All anakinra patients developed anti-drug antibodies (ADAs) at some timepoint during the study. ADAs were persistent throughout the treatment period, except in one patient. Titers were low to moderate. One placebo patient had low ADA titers at one occasion. No neutralizing antibodies were observed and the ADAs did not appear to impact clinical efficacy or safety.

**Conclusion:** Anakinra is superior to placebo in the treatment of Still’s disease. ADAs occur frequently but do not appear to adversely impact efficacy or safety. These results confirm the benefits of anakinra treatment in patients with active, newly diagnosed Still’s disease across ages.

**Figure 1:** Individual ACR90 response with absence of four and treatment duration over time and ACR50/70 at week 2

**Figure 2:** Time to study drug discontinuation, Kaplan-Meier plot

**Disclosure of Interests:** Laura Schanberg Grant/research support from: Sobi, BMS, Consultant of: Aurinia, UCB, Sanofi, Peter Nigrovic Grant/research support from: Novartis, BMS, Pfizer, Consultant of: Novartis, BMS, Pfizer, Miach Orthopedics, Simcere, XBiotech, Quench Bio, Ashley Cooper: None declared, Winn Chatham Grant/research support from: Sofi, Consultant of: Sofi, Shoghik Akoghlanian: None declared, Namrata Singh: None declared, Egla Rabinovich: None declared, Alysha Taxter: None declared.

**AB1060**

**A RARE IGG4-REALTED DISEASE PHENOTYPE WITH BONE Destructive LESSIONS**

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**Background:** IgG4-related disease (IgG4-RD) is an immunemediated fibroinflammatory condition with systemic course that can affect almost any organ in the body. In the majority of cases it is a benign condition with slow progression. The clinical symptoms are usually determined by compression of nearby anatomic structures by tumefactive lesions, but not by invasive growth of the pseudotumor with the destruction of the organs. Bone destruction always raises suspicion of malignant tumor, but it should be considered in the course of IgG4-RD as well.

**Objectives:** To report a very rare bone destructive phenotype of IgG4-RD.

**Methods:** We report 5 cases of biopsy proven IgG4-RD with bone destructive lesions.

**Results:** In our cohort of patients there were 3 patients with multiple fusal bone destruction due to maxillary sinus pseudotumor expansion and 2 patient with vertebral destruction lesions. Patients 1, 2, 3 with facial bone destruction were young aged 42, 36 and 28 years. In cases the primary lesion was located in the maxillary sinus with expansion to the fascial soft tissues and orbit. Two patients had probable and 1 possible IgG4-RD diagnosis according to consensus diagnostic criteria, 2011. All patients were treated with rituximab and low doses of glucocorticoids with improvement of clinical symptoms (fascial edema, eye pain and/or headache), but lacking in radiologic improvement.

**Conclusion:** IgG4-RD can manifest by bone, including vertebral, destructive lesions and thus should be included when considering differential diagnosis in patients with bone destruction.

**Disclosure of Interests:** None declared

**AB1061**

**2019 ACR/EULAR CLASSIFICATION CRITERIA FOR IgG4-RELATED DISEASE IN RUSSIAN COHORT OF PATIENTS.**

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**Background:** IgG4-related disease (IgG4-RD) is a systemic immunemediated fibroinflammatory condition that can affect almost any organ in the body. This is the reason for dramatic variety of clinical symptoms and complexity of diagnostics. 2011 Comprehensive diagnostic criteria (CDC) for IgG4-RD are used to establish the diagnosis for all lesions (except autoimmune pancreatitis type 1). In 2019 the new ACR/EULAR classification criteria for IgG4-RD were proposed to facilitate the formation of more homogeneous groups of patients primarily for clinical trials inclusion purpose. They also provide a framework for clinicians considering diagnosis of IgG4-RD.

**Objectives:** To evaluate 2019 ACR/EULAR classification criteria for IgG4-RD in Russian cohort of patients with IgG4-RD.

**Disclosure of Interests:** None declared

**DOI:** 10.1136/annrheumdis-2020-eular.6391
Methods: 59 patient with IgG4-RD according to CDC with biopsy proven diagnosis were included.

Results: The mean number of affected organs was 2.1 ± 31 patients (52.5%), women. Majority of patients had saliadenitis (25 patients) and/or orbital disease (31 patients), 9 had retroperitoneal fibrosis (RPF). Other affected organs were lungs, pancreas, lymph nodes, paranalal sinuses, thyroid and low urinary tract. Twenty five (25) patients (42.4%) had definite, 14 (23.3%) probable and 20 (34.3%) possible diagnosis of IgG4-RD. Twenty three (23 patients) (39%) didn’t fulfill the 2019 ACR/EULAR classification criteria for IgG4-RD. Among them were the majority of patients with RPF (7 patients) who were lacking other organ involvement and IgG4 hypersecretion either in the tissue or serum. The majority of excluded cases were due to inadequate morphological evaluation (lacking of the exact number and percentage in >40%) of IgG4+ cells), lacking of multi-organ involvement or different patterns of involvement, e.g. in case of lungs involvement.

Conclusion: The new 2019 ACR/EULAR classification criteria for IgG4-RD are very useful in evaluation of typical organ involvement and systemic course of IgG4-RD. It is essential to adjust Russian pathomorphologist’s approach to cell counting and percentage determination for IgG4-RD cases to get suitable protocols.

Disclosure of Interests: None declared
DOI: 10.1136/annrheumdis-2020-eular.6327

AB1062 LIPODERMATOSCLEROSIS AS A TYPE OF LOBULAR PANNICULITIS: THE EFFECTIVENESS OF NON-PHARMACOLOGICAL TREATMENT METHODS

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Background: In medical practice lobular panniculitis-lipodermatosclerosis (LDS) is becoming more and more common. It is manifested by degenerative-dystrophic changes in subcutaneous fat (SCF) and occurs more often in middle-aged women affected by chronic venous insufficiency.

Objectives: to evaluate the effectiveness of mesotherapy (MT) and shockwave ultrasound therapy (UST) for LDS

Methods: among 538 patients referred to the V.A. Nasonova Research Institute of Rheumatology with the referral diagnoses of erythema nodosum or panniculitis 8.5% (46) of patients (44 women, 2 men) aged 18 to 82 with overweight (32) LDS with the disease duration of 11,8±6,4 months was verified. Patients were randomized into two groups of 23 patients each: group I received daily MT (10 sessions) therapy with drugs that have antioxidant, anti-inflammatory, lymphatic drainage and lipolytic effects, and 3 kHz UST of the node area twice a week (5 sessions). In group II MT was performed daily with 9% Natrii chloridum solution at a dose comparable to group I. The control methods included general clinical examination (characterization of induration on the lower legs with an assessment of pain intensity (T0 50±18 mm; T1 35±11 mm), decrease in diameter (T0 6±2.2 cm; T1 4.5±1, 7 mm) and color intensity of the node (p=0.002), SCF thickening which results in “lumping” with macrovascularization according to USE, and decrease in ESR and CRP. In 44% of cases the treatment effect increased to T2 (p<0.05). After 3 months of observation, 15 patients required a second course of therapy. In group II a positive clinical effect was registered for T2 in 14 patients (60.8%) and for T3 in 19 patients (83%) (p<0.05). The entire observation period LDS recurrence was registered in 19 patients (41%), the median of recurrence was 3 [1; 6] months, mainly in patients of group I. Recurrence was associated with node fusion into conglomerates (OR 4.33, 95% CI 1.05-17.6; p = 0.037). MT and UST were tolerated well, no side effects were detected.

Conclusion: the use of MT with 9% Natrii chloridum solution allowed us to achieve positive dynamics in patients with LDS, which significantly reduced the cost of treatment. Further studies are needed to evaluate the significance of these techniques.

Disclosure of Interests: None declared
DOI: 10.1136/annrheumdis-2020-eular.1443

AB1063 CASE SERIES OF PATIENTS WITH CHRONIC NON-TUBERCULOUS MYCOBACTERIA ACCOMPANYING WITH ANTI-INTERFERON GAMMA ANTIBODY

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Background: Anti-interferon (IFN) gamma antibody was a rare autoantibody which mainly been reported in Asia. It interferes the interferon pathway and eradication of intracellular pathogens, such as Salmonella or Mycobacteria. This rare disease should be raised for more awareness.

Objectives: To analyze clinical presentations of the patients with anti-interferon (IFN) gamma antibody.

Methods: We checked anti-IFN gamma antibody when patient fulfilled multiple NTM infection (especially bone marrow and lymph nodes). There are 6 cases of non-tuberculosis mycobacteria (NTM) infection accompanying with anti-IFN gamma antibody in our hospital from 2015 to 2019 of hospitalist ward, and the antibody titer is recorded.

Results: Among these cases, patients had initial presentations of fever (100%), elevated CRP and ESR (100%), lymphadenopathy (67%), body weight loss (50%), and elevated LDH (50%). All the 6 cases had negative results of TB-PCR test, but 1 had positive result of interferon gamma release assay (IGRA, 17%). NTM was identified from lymph node biopsy (50%), sputum (33%), skin (33%), and bone marrow (33%). Different NTM was identified, including Mycobacterium abscessus, Mycobacterium fortuitum group complex, Mycobacterium kansasi, and Mycobacterium avium complex. Prolonged antibiotics treatment was used, but all patients had recurrent or persisted NTM infection under medications. 4 cases had different NTM strain after 1 year of treatment from the initial involved organ. One case had an episode of Salmonella bacteremia with septic shock. Another case had positive result of ANA, and other cases had negative auto-antibodies.

Conclusion: Careful history taking and physical examinations are crucial in diagnosis this acquired immunodeficiency disease. This rare but unique disease should be taken into consideration.

Disclosure of Interests: None declared
DOI: 10.1136/annrheumdis-2020-eular.61610