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 safety 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 fever and treatment duration over time and ACR30/50/70 at week 2

Bars indicate time on study drug. Bullets indicate ACR90 response at the particular week. Patient 1 had sustained ACR70 response but not ACR90.

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

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AB1060

A RARE IGG4-REALTED DISEASE PHENOTYPE WITH BONE DESTRUCTIVE LESIONS

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Background: IgG4-related disease (IgG4-RD) is an immunomediated 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 fascial bones destruction due to maxillary sinus pseudotumor expansion and 2 patient with vertebral destruction lesions.

Patients 1, 2 and 3 with facial bone destruction were young men aged 42, 36 and 28 years. In all 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 (facial edema, eye pain and/or headache), but lacking in radiologic improvement.

Patient 4 is a 51-year-old man who developed retrosternal pain, mimicking angina pectoris. Chest CT showed a tumor of posterior mediastinum, infiltrating the right lung and Th6-Th8 bodies destruction. Biopsy of the lesion revealed lymphoplasmatic infiltrate with lymph follicles formation, few eosinophils, significant diffuse fibrosis. Immunohistochemical study, showed IgG:IgG4 ratio >40%. The serum IgG4 concentration was 1.94 g/l (normal range below 2.0 g/l). Combined treatment was administered: rituximab 500 mg weekly #4 and cyclophosphamide 1000 mg + methylprednisolone 250 mg IV every 2 weeks #6. After that - cyclophosphamide 200 mg per week intramuscularly, methylprednisolone 4 mg daily per os. Due to spinal instability the patient undergone surgery. Six months later at check-up examination CT has shown a dramatic decrease of the mediastinal infiltrate. The treatment with cyclophosphamide and oral metylprednisolone was tapered gradually during 2 years due to no sites of pathological hypermetabolic activity were found on PET.

Patient 5 is a 60-year-old woman who had spinal surgery for C2 odontoid destruction. Biopsy revealed chronic inflammatory with massive fibrosis. 1,5 years later she developed salivary glands enlargement. During evaluation she had elevated serum IgG4 4.9 g/l and IgM 4.1 g/l (normal range below 0.6-3.7 g/l), serum protein electrophoresis with immunofixation showed monoclonal IgMκ 3.2 g/l. Left submandibular salivary gland biopsy revealed significant fibrosis, dense lymphoplasmatic infiltrate forming lymph follicle with IgG:IgG4 ratio 50-80% and no pathological signs of lymphoma. The patient was treated with rituximab 500 mg per week #2 and cyclophosphamide 1000 mg + methylprednisolone 250 mg IV every 2 weeks #3. The treatment was discontinued because of toxic hepatitis. Unfortunately, no follow-up was available. The tissue specimens after spinal surgery were also unavailable.

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.

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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 immunomediated 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 homogenous 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.

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%) were women. Majority of patients had sialoadenitis (25 patients) and/or orbital disease (31 patients), 9 had retroperitoneal fibrosis (RPF). Other affected organs were lungs, pancreas, lymph nodes, paranasal 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 pathomorphological evaluation (lacking of the exact number and percentage (if >40%) of IgG4+ cells), lacking of multi-organ involvement or different patterns of involvement, e.g. in case of lungs involvement.

Conclusion: The new 2019ACR/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 pathomorphologists's approach to cell counting and percentage determination for IgG4-RD cases to get suitable protocols.

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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 539 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 MHz 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 the effect of pain pressing according to visual analogue scale (VAS pain), general blood and urine tests and ultrasound with elastography (USE) of the compaction. The main stages of control: initial (T0), after 14 days (T1), 1 month (T2) and 3 months (T3).

Results: before treatment 38 patients with LDS demonstrated asymmetric (83%) inflammation of SCF of the lower legs (100%) on its medial surface (91%). LDS regressed faster with normal body mass index ($p = 0.04$). In all patients of group I, after a course of physiotherapy a positive trend was registered, that is a decrease in VAS pain intensity (T0 50±18mm; T1 35±11mm), decrease in diameter (T0 6±2.2cm; T1 4.5±1, 7mm) 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 physiotherapy. In group II a positive clinical effect was registered for T2 in 14 patients (60.8%) and for T3 in 19 patients (83%) ($p < 0.05$). Over 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 1.05-17.8; $p = 0.037$). 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.

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AB1063

CASE SERIES OF PATIENTS WITH CHRONIC NON-TUBERCULOSIS 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 kanasii, 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.

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AB1064

IMMUNOGLOBULIN G4-RELATED DISEASE (IGG4-RD): CLINICAL AND LABORATORY CHARACTERISTICS, TREATMENT RESPONSES AND PROGNOSIS IN ONE HUNDRED FIVE PATIENTS.

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Background: IgG4-RD is a systemic fibro-inflammatory condition with incompletely understood that is capable of affecting multiple organs.

Objectives: We aimed to investigate clinical and laboratory findings in Japanese patients with IgG4-RD.

Methods: Dates on clinical characteristics, laboratory features, and treatment response from patients with IgG4-RD in our hospital were reviewed retrospectively from January 2004 to September 2019.

Results: Among 105 patients were diagnosed and treated in our hospital, 48% were female and 88% were biopsy-proven. The median age of the patients was 66 years, and female were younger at their diagnosed age ($p = 0.04$). Their median duration of follow-up was 45 months. 48% of the patients had allergic history (including sinusitis, asthma, hay fever), younger patients tended to have allergy history. Mean serum IgE was 303 IU/dL (2-4965 IU/dL). Salivary and lacrimal gland involvement (63%) and dacryoadenitis and ocular and orbital inflammatory disease (56%), autoimmune pancreatitis type 1 pancreatitis (18%), retroperitoneal fibrosis (16%), aortitis (15%) predominantly occurred. 84% of the patients had serum IgG4 higher than 135mg/dL, and high IgG and IgG4 concentration was associated with lower complements (CH50, C4) levels. Mean serum IgG was 1860mg/dL (861-8432mg/dL), and IgG4 was 449mg/dL (28-3210mg/dL). Male patients show higher serum IgG and IgG4 concentrations at baseline ($p < 0.01$). Younger patients and low serum C4 level were associated with necessity of treatment, 60 of them used steroid, and the mean dose of prednisone they used was 30mg. Most of them responded well and tapering steroid. Steroid sparing agents were used in 23% of them. Although 23% of patients relapse as tapering steroid, 15% of them could stop treat with steroid. Retreatment with glucocorticoids is not associated with any factors. There were 14 malignancies in 13 patients during the follow-up period.

Conclusion: Our study revealed that IgG4-RD occurred in middle age with allergic disease in Japanese patients. The pattern of head and neck was predominance. For the most part of the patient serum IgG and IgG4 concentrations was high. Serum low complement level could be associated with its diagnosis and necessity of treatment with steroid. Younger patients tend to treat with steroid and they responded well.

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