Table 1. Demographic and clinical characteristics of the patients

	MEFV gene mutations	Treatment cycles (months)	Attacks per year before TCZ	Attacks per year after TCZ
44/Female	M694V homozygote	96	24	5
49/Male	M694V homozygote	48	30	0
59/Male	M694V/N heterozygote	8	12	1
79/Male	M694V/M680I heterozygote	67	12	0
40/Female	M694V homozygote	2	14	1
38/Male	M694V homozygote	3	48	3
44/Male	M694V homozygote	18	30	10
49/Male	M694V homozygote	3	36	0
34/Male	M694V homozygote	23	24	0
39/Male	MEFV/- heterozygote	33	20	0
28/Male	M694V homozygote	19	18	0
24/Male	M694V homozygote	12	30	2
27/Male	M694V homozygote	3	36	36
27/Male	M694V/M680I heterozygote	12	12	0
50/Male	M694V homozygote	12	6	0

MEFV:Mediterranean Fever, TCZ:Tocilizumab

Conclusion: Tocilizumab is found to be effective on improvement of amyloidosis. In the current analysis, we found that TCZ also decreases the frequency of recurrent attacks in patients with FMF. Besides, TCZ is well tolerated among the patients. Further and prospective studies with larger sample are needed to support these results.

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FRI0587 IGG4-RELATED DISEASE IN ITALY: RESULTS FROM A MONOCENTRIC COHORT OF 150 PATIENTS (2013–2018)

Marco Lanzillotta, Emanuel Della Torre, Corrado Campochiaro, Enrica Bozzolo, Lorenzo Dagna. *San Raffaele Scientific Institute, Milan, Italy*

Background: IgG4-related disease (IgG4-RD) is a recently recognized fibro-inflammatory disease characterized by fibrotic lesions and elevated IgG4 serum level. Epidemiological and clinical data in our country have not been precisely assessed yet

Objectives: The aim of this work is to present the data from the largest monocentric Italian IgG4-RD cohort.

Methods: We conducted a retrospective analysis searching for IgG4-RD patients referred to our tertiary care centre between 2013 and 2018. Diagnosis were made according to the "Comprehensive Diagnostic Criteria 2011", and patients were clustered on the basis of a definite, probable and possible, diagnosis. Each patient underwent clinical, serological, and radiological evaluation according to the organ involvement. IgG4-RD activity was measured with IgG4-RD Responder Index (IgG4-RD RI). All the data are reported in median and interquartile range.

Results: 150 patients were included in this work (111 male, 39 female, M:F ratio 2.8:1). 75 cases received a definite IgG4-RD diagnosis, while 6 patients and 49 patients were diagnosed as *probable* and *possible* IgG4-RD, respectively. Median age at diagnosis was 60 (SD 13) years old. The most involved organ was the pancreas (66 cases; 44%), followed by lymph nodes (26 cases; 17%), biliary tree (25 cases; 17%), salivary glands (21 cases; 14%), aorta and retroperitoneum (12 cases; 8%), meninges (9 cases; 6%), nasal sinuses and nasal septum (5 cases; 3%). 18 patients (12%) had a positive history of malignancies that occurred ten years earlier or after IgG4-RD diagnosis. At baseline, IgG4 level was

elevated in 92 patients (61%), while eosinophil count and IgE level were elevated in 35 (23%) and 52 (35%) patients, respectively. Circulating plasmablast were measured in 88 and elevated in 66 (1859 cell/mL, 600-3950). Median IgG4-RD RI at diagnosis was 6 (6-9). 55 patients were treated with glucocorticoids alone, whereas immunomodulator (Mycophenolate mofetil, azathioprine, cyclophosphamide) were associated in 31 cases. Rituximab was administered in 32 patients. 15 patients underwent surgical excision of the fibrotic lesion. 15 patients were candidate to a watchful waiting strategy, due to low disease burden.

Conclusion: Our study reports clinical and epidemiological data from the largest monocentric cohort of patients with IgG4-RD in Italy. Our data reflect those reported in other international cohorts of IgG4-RD patients.

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FRI0588 THE SCALE OF ACTIVITY IN IDIOPATHIC LOBULAR PANNICULITIS

Olga Egorova, Boris Belov, Svetlana Glukhova. *V.A. Nasonova Research Institute of Rheumatology, Moscow, Russian Federation*

Background: Idiopathic lobular panniculitis (ILP) refers to chronic systemic connective tissue lesions (M 35.6). The disease develops with typical clinical symptoms and is characterized by alternating periods of stable condition (remission) and active manifestations (exacerbations). Assessment of the activity and identification of the disadvantageous predictors is the main task in monitoring patients with ILP, however, the "gold standard" has not been developed yet.

Objectives: to create a rating scale for the inflammatory process activity in ILP on the basis of clinical and laboratory parameters

Methods: We examined 67 patients (9 men and 58 women) with the ILP diagnosis verified in the V.A. Nasonova Research Institute of Rheumatology in 2007-2017. The age of patients ranged from 20 to 76 years, the average duration of the disease was 78.91[48;50] months. At the time of treatment at the institute only 16 patients (23,88%) were diagnosed with ILP. All the patients underwent comprehensive clinical examination and laboratory and instrumental examination of biochemical and immunological parameters, radiography or computed tomography (CT) of the chest organs, as well as pathomorphological examination of skin and subcutaneous fat biopsies from the node area.

Results: Analysis of clinical manifestations allowed to identify four forms of ILP: nodular (30 patients), plaque (10), infiltrative (15) and mesenteric (12), which had specific clinical and laboratory features. Based on the obtained data we developed a scale of the ILP activity (SA), which includes a description of the state of the 7 organ systems. The maximum score for individual systems is from 1 to 3 points depending on the number of estimated parameters. The total maximum possible score is 42 points. The score includes all types of damage since the onset of the disease (caused directly by ILP or developed as a result of therapy), while taking into account only signs that persist for 6 months. Score <5 shows inactive disease, 5-10 - low disease activity; 11-20 - moderate activity and >20 - high activity of the disease. Thus, low disease activity is characterized by the predominance in the clinical picture of limited moderately painful indurations in the absence of any significant changes in laboratory tests. At the moderate disease activity, changes in various localization with a predominance of proliferative disorders (common painful compaction of the 1st and/or 3rd stage, moderate area of damage to the trunk and limbs, subfebrile temperature, cognitive impairment, mesenteric disorders, etc.) are revealed against the background of less pronounced laboratory tests. ILP with high activity is characterized by the presence of fever and other common signs of the disease, the predominance of the 2nd stage with a pronounced VAS pain intensity of the induration, the common nature of the defeat of the pancreas, involvement of the lungs, heart and gastrointestinal tract in the pathological process.

Conclusion: In fact, SA is an analysis of the organ damage, assessment of the symptoms severity, and it takes into account the development of exacerbations with the involvement of new organs in the process of the disease. The proposed ILP SA is of practical importance. Further

research and possibly search for new parameters of the ILP activity are needed.

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FRI0589

IS THERE AN OVERLAP OF ANTINEUTROPHIL CYTOPLASMIC ANTIBODY-ASSOCIATED VASCULITIDES WITH IGG4-RELATED DISEASE OR NOT?

Abdulsamet Erden, Ertuğrul Çağrı Bölek, Gözde Kübra Yardımcı, Şule Apraş Bilgen, Omer Karadag, Hacettepe University Vasculitis Centre, Hacettepe University Vasculitis Centre. *Hacettepe University Vasculitis Center, Ankara, Turkey*

Background: Pseudotumor orbita, pachymeningitis, periaortitis could be seen in both ANCA-associated vasculitis and IgG4-RD. Sometimes it may be difficult to differentiate these two entities. The co-occurrence/concurrence of Antineutrophil cytoplasmic antibody (ANCA)-associated vasculitides (AAV) and IgG4-related disease (IgG4-RD) was recently published by a collaborative EUVAS group [1].

Objectives: Firstly, we aimed to investigate ANCA positivity of our IgG4-RD cohort. Secondly, a literature review of co-occurrence/concurrence of AAV and IgG4-RD was done.

Methods: Data of totally 62 patients with IgG4-RD in Hacettepe Vasculitis Center Database was used. Patients were diagnosed with IgG4-RD according to comprehensive diagnostic criteria [2]. Dataset of patients including demographic data, clinical characteristics, and imaging and laboratory findings of IgG4-RD was re-evaluated in terms of AAV and ANCA test.

At next step, we performed a systematic literature review of the PUBMED database covering the time period until April 2018. Relevant publications were searched using the MeSH terms "IgG4-related disease and Eosinophilic Granulomatosis with Polyangiitis", "IgG4-related disease and Anti-Neutrophil Cytoplasmic Antibody-Associated Vasculitis" and "IgG4-related disease and Granulomatosis with Polyangiitis".

Results: Totally 29 (46.7%) of our patients had ANCA results. Out of 29 patients 15 (51.7%) were considered as probable, 10 (34.5%) as definite and 4 as possible (13.8%) for IgG4-RD. Three (10.3%) of these patients had ANCA positivity. All of these ANCA titers were in low degree positivity (MPO-ANCA 1/100, MPO-ANCA 1/32 and PR3 ANCA 1/100). These three patients didn't have any findings of vasculitis and didn't have granuloma in their biopsy. When we evaluate these three patients with regards to meeting the Ig G4- RD criteria, 1 was definite, 1 was probable and 1 was possible.

In literature review, we found 17 cases that having both features of IgG4-RD and AAV (Table). These cases were re-evaluated according to the 'Comprehensive Diagnostic Criteria for IgG4-RD'. Diagnoses of IgG4-RD were definite in 11 cases (64.7%), probable in 2 cases (11.8%) and possible in 4 cases (23.5%). ANCA were positive in 15 of 17 patients (88%). ANCA were directed against proteinase 3 (PR3-ANCA) in 6 patients and were directed against myeloperoxidase (MPO-ANCA) in 5 patients. Other four cases had both MPO-ANCA and PR3-ANCA. All PR3-ANCA positive cases have high titers of ANCA, whereas only one MPO-ANCA positive case has high titers of ANCA.

Conclusion: None of our IgG4-RD patients have any overlap with ANCA-associated vasculitis. Only in 3 patients (10.3%), ANCA positivity was detected without any histopathologic evidence. Just two patients of literature review, seemed to be full compatible with both diseases. Even though ANCA-associated vasculitis and IgG4-RD share clinical features, we think this might be as co-occurrence instead of a histopathologic link.

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Disclosure of Interests: None declared

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Table: Distribution of Demographic, Clinical, Features in Seventeen Patients at Literature

Author	No. of Patients	Age (mean)	Sex (M/F)	ANCA	ANCA Titers	ANCA Specificity	ANCA Positive Cases	ANCA Negative Cases
Erden et al.	29	51.7	15/14	PR3-ANCA	1/100	1/32	1/100	1/100
...	...	...	...	...	...	...	...	...

FRI0590

UVEITIS SECONDARY TO CHECKPOINT INHIBITORS

JAVIER GARCÍA, Eugenio Perez-Blazquez. *HOSPITAL 12 DE OCTUBRE, MADRID, Spain*

Background: The introduction of immunotherapy (immune checkpoint inhibitors, ICI) has led to a revolution in oncological treatments. The inhibitors of CTLA4 (ipilimumab), of PD1 (pembrolizumab, nivolumab) and of the ligand of PD1 (atezolizumab, avelumab, durvalumab) regulate T activation and its effector function, being effective for the treatment of various types of cancer¹. However, this effect leads to a series of immune-mediated adverse events, among which uveitis of autoimmune mechanism have been described in about 1% of treatments².

Objectives: Methods: descriptive study of retrospective review of the cases of a tertiary hospital with about 1000 treatments between the beginning of 2014 and the end of 2018.

Results: A series of 4 cases of uveitis of autoimmune origin associated with ICI is presented (see table). The series described has characteristics similar to the information previously reported in the literature², with an incidence of around 0.4%, according to the previously described, with frequencies of ocular toxicity around 1%, being the uveitis is the most frequent form of presentation. The time of presentation of uveitis since the beginning of treatment has been in all cases in the first 6 months. The forms of presentation described before, ranges from anterior uveitis (AU) to panuveitis and often papilledema, usually bilateral, as are all those described in our series. In addition, 50% are AU (grade 2) and the other 50% panuveitis with papilledema (grade 3). In all the cases described, treatment was interrupted, a half it could be reintroduced without recurrence of uveitis. All the patients received topical treatment, the more severe systemic corticosteroids at doses of 0.5-1 mg/kg in accordance with the recommendations. The final prognosis was good, with preservation of VA in all cases, and topical and systemic treatment could be withdrawn in all patients. A patient treated with Ipilimumab presented synchiae as a complication.