in tissue (on frozen tissue section or paraffin embedded) by PCR analysis of immunoglobulin V-D-J genes heavy chain rearrangements (FR1, FR2, FR3); monoclonal secretion was tested in serum protein electrophoresis.

Results: 40 patients with IgG4-ROD had orbital lesions at the onset of the disease and 13 developed them after 2 - 9 years from the onset. The frequency of different orbital anatomic structures involvement see in table 1. 15 (28%) patients had isolated orbital lesions while 38 (72%) had systemic IgG4-RD with simultaneous involvement of 2 - 7 other organs. 6 (14%) patients had unilateral orbital lesions. 23 (53%) patients with IgG4-ROD had associated IgG4-related sialoadenitis and nasal lesions. Most common pathomorphological features of IgG4-ROD see in table 2. The most common laboratory features of IgG4-ROD were elevation of: total serum IgG (44%), serum IgG4 (88%), serum IgE (61%) and monoclonal serum secretion (23%).

Table 1. Prevalence of different orbital anatomic structures affection in IgG4-ROD (n=43)

Anatomic structure	N, %
Lacrimal gland	93
Extraocular muscles	85,5
Fibroinflammatory lesions of eyelids	32,5
Retrolbulbar infiltration	32,5
Optic nerve thickening	32,5
Trigeminal involvement	21
Nasolacrimal duct	4,5

Table 2. Pathomorphological features in IgG4-ROD (n=43)

Pathomorphological feature	N, %
Lymphoplasmatic infiltrate	93
Dense fibrosis	93
Lymphoid follicle formation	69
Tissue eosinophilia	32,5
Obliterative phlebitis	14
Vasculitis	7
IgG4/IgG > 40%	90,5
MALT-lymphoma of lacrimal gland	2,3
AL-amyloidosis	2,3

Conclusions: In our cohort of IgG4-RD patients orbit is the leading site of the disease (64.5% of patients) and the disease onset (75% of patients). The majority of patients have bilateral orbital lesions (89%) and systemic course of the disease with other organ involvement (72%). Most often affected orbital anatomic structures are lacrimal glands, extraocular muscles, retrolbulbar infiltration with optic nerve thickening and fibroinflammatory lesions of eyelids. MALT-lymphoma of lacrimal gland and local AL-amyloidosis can complicate the long history of IgG4-ROD. Monoclonal serum secretion and B-cell clonality in the tissue in 23% of patients can potentially act as a background for lymphoma formation.

References:

- [1] Uchida K, Masamune A, Shimosegawa T et al. Prevalence of IgG4-related disease in Japan based on nationwide survey in 2009. *IntJ.Rheumatol.* 2012;2012:358371.
- [2] Umehara H, Okazaki K, Masaki Y et al. A novel clinical entity, IgG4-related disease (IgG4RD): general concept and details. *ModRheumatol.* 2012; 22: 1-14.
- [3] Stone JH, Khosroshahi A, Deshpande V et al. Recommendations for the Nomenclature of IgG4-Related Disease and Its Individual Organ System Manifestations. *Arth.Rheum.* 2012; 64:3061-3067.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2017-eular.4283

FRI0622 A RETROSPECTIVE COHORT STUDY ABOUT THE CLINICAL CHARACTERISTICS OF IGG4-RELATED DISEASES

X. Wu, C. Wang, Y. Miao, Y. Huang, L. Dong. *Department of Rheumatology and Immunology, Tongji Hospital of Tongji Medical College, Huazhong University of Science and Technology, Wuhan, Hubei, China*

Background: Immunoglobulin (Ig) G4-related disease (IgG4-RD) is an increasingly recognized systemic autoimmune disease in the past decade, and encompasses a wide spectrum of disorders previously considered to be unrelated, but with common pathologic, serologic, and clinical features. There have been several hundred literatures on IgG4-RD published in the past few years, however, we still cannot well characterize this complicated disease, including clinical features, treatments as well as mechanisms, which promotes us to further describe this disease.

Objectives: To describe the clinical presentations, laboratory features, imaging manifestations, treatments and evolutions in a cohort of 34 Chinese patients with immunoglobulin G-related diseases (IgG4-RD).

Methods: A retrospective study was performed in Wuhan Tongji Hospital. Electronically stored records of clinical, laboratory, imaging, and histological features were reviewed. The diagnoses were made according to comprehensive diagnostic criteria for IgG4-RD. Disease activity was assessed by IgG4-RD Responder Index (RI). This study was approved by the Ethical Committee of Tongji Hospital (TJ-C20150603). All patients were contacted, and consents were obtained from all the participants.

Results: We found that among the whole 34 patients enrolled in this study (median

age=51 years, male/female ratio=1.4/1), 65.2% tested patients had a declined HDL level, 82.3% tested patients had an elevated IgG4 level, and lymph node was the most frequently involved organ. 18F-FDG PET/CT had high sensitivity in providing information about the organ distribution of IgG4-RD. The IgG4 level and IgG4-RD RI in the multi-organs affected patients were significantly higher than that in the localized ones. Most patients were prescribed with glucocorticoids (GCs) alone or combined with disease modifying antirheumatic drugs (DMARDs), the majorities responded well, while some relapsed. Patients given rituximab (RTX) obtained good responses.

Table 1. The overall prognosis of 19 patients who were followed up

Prognosis	GCs	GCs + DMARDs	Surgery + GCs	Surgery + DMARDs	Antifungal therapy	MMF	Total, n
Remission, n	5	1	2	0	0	1	9
Recurrence, n	3	4	0	0	0	0	7
Worsen/Persistence, n	0	1	0	1	1	0	3
Total, n	8	6	2	1	1	1	19

Abbreviations: GCs, glucocorticoids; DMARDs, disease-modifying anti-rheumatic drugs; MMF, mycophenolate mofetil.

Fig 1: Analysis of IgG4, IgG4/IgG(%), IgG4-RD RI, and ESR levels between patients of single organ involvement and multiple organs involvement.

Conclusions: It was concluded that 18F-FDG PET/CT can be an effective tool to detect involved lesions of IgG4-RD. The addition of DMARDs to the initial GCs therapy in the remission induction stage may not bring much benefit. The most appropriate dosage schedule of RTX needs to be further investigated. Patients with younger age, longer duration of disease, higher IgG4 level and IgG4/IgG ratio may more commonly experience recurrence.

References:

- [1] Hamano H, Kawa S, Horiuchi A, et al. High serum IgG4 concentrations in patients with sclerosing pancreatitis. *N Engl J Med.* 2001;344(10):732-738.
- [2] Islam AD, Selmi C, Datta-Mitra A, et al. The changing faces of IgG4-related disease: Clinical manifestations and pathogenesis. *Autoimmun Rev.* 2015;14(10):914-922.
- [3] Martinez-de-Alegria A, Baleato-Gonzalez S, Garcia-Figueiras R, et al. IgG4-related Disease from Head to Toe. *Radiographics.* 2015;35(7):2007-2025.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2017-eular.3623

FRIDAY, 16 JUNE 2017

Diagnosics and imaging procedures

FRI0623 [18F]FLUORIDE PET-CT IMAGING OF BONE FORMATION IN ANKYLOSING SPONDYLITIS BEFORE AND AFTER 12 WEEKS OF ANTI-TNF TREATMENT

S. Bruijnen¹, N. Verweij¹, L. van Duivenvoorde¹, N. Bravenboer², D. Baeten¹, C. van Denderen¹, I. van der Horst-Bruinsma¹, A. Voskuyl¹, P. van de Ven³, J. Bot⁴, B. Boden⁴, A. Lammertsma⁴, O. Hoekstra⁴, P. Rajmakers⁴, C. van der Laken¹. ¹Rheumatology, Amsterdam Rheumatology and immunology Center; ²Clinical Chemistry; ³Epidemiology and Biostatistics; ⁴Radiology & Nuclear Medicine, VU University Medical Center, Amsterdam, Netherlands

Background: Excessive bone formation is an important hallmark of ankylosing spondylitis (AS). Recently, it has been demonstrated that axial bony lesions in AS patients can be visualized using [¹⁸F]fluoride PET-CT. This technique may also provide opportunities to monitor changes in bone formation in axial AS lesions during therapy.

Objectives: To assess (1) changes in [¹⁸F]fluoride uptake in axial lesions of AS patients before and 12 weeks after treatment with TNF-inhibitors (TNFi) and (2) whether [¹⁸F]fluoride uptake on PET is related to focal bone formation in spine biopsies.