The course of the disease was mostly recurrent (54%) with asymptomatic periods between flare-ups. About half of the patients (44%) in follow-up currently have active disease.

Conclusion: Our cohort presented axial involvement like other series and significant peripheral involvement. The most frequent skin condition was PPP. Half of the patients in our cohort required bDMARD, with ADL being the most widely used.

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Clinical cases

POS1575 ASSOCIATION OF IGG4-RELATED DISEASE AND SYSTEMIC RHEUMATIC DISORDERS

Keywords: Rare/orphan diseases

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Background: Autoimmune disorders can occur together especially in genetically predisposed individuals.

Objectives: We here aimed to assess the occurrence of IgG4-related disease (IgG4-RD) in association with other systemic immune-mediated conditions.

Methods: We retrospectively analyzed the clinical records of our routinely followed patients with IgG4-RD for pre-existing or concomitant immune-mediated disorders. IgG4-RD was diagnosed based on histological findings and on the 2011 Comprehensive Diagnostic criteria. Associated immune-mediated disorders were diagnosed based on available classification and/or diagnostic criteria.

Results: Two-hundred and thirty-four patients with a definitive diagnosis of IgG4-RD were included in this study. A pre-existing immune-mediated connective tissue disease was reported in 6/234 patients (3%): one case each of sarcoidosis, Takayasu arteritis (TA), eosinophilic granulomatosis with polyangitis (EGPA), and rheumatoid arthritis; and two cases of granulomatosis with polyangitis (GPA). Organs involved by IgG4-RD included the lungs, the pancreas, the peritoneum, lacrimal glands, meninges and orbits. Sarcoidosis, EGPA, and TA preceded the onset of IgG4-RD. GPA preceded IgG4-RD onset in one case and occurred simultaneously in the other case. Rheumatoid arthritis occurred together with IgG4-RD in one case.

Conclusion: IgG4-RD can present in the context of pre-existing systemic immune-mediated disorders. Our observation suggests that "secondary" IgG4-RD may preferentially complicate chronic granulomatous conditions including large- and small-vessels vasculitis as well as sarcoidosis.

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POS1576 AN UNUSUAL INITIAL PRESENTATION OF VEXAS SYNDROME

Keywords: Rare/orphan diseases

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Background: VEXAS (Vacuoles, E1 enzyme, X-linked, Autoinflammatory, Somatic) syndrome is an adult-onset multisystem autoinflammatory disease caused by a somatic mutation in the ubiquitin-activating enzyme (UBA1) gene in myeloid or erythroid precursor cells, causing a variety of symptoms including fever, cytopenia, vacuoles in myeloid and erythroid progenitors, dysplastic bone marrow, neutrophilic dermatosis, pulmonary infiltrates, chondritis and/or vasculitis.

Table 1 Characteristics of IgG4RD and other immunological diseases

Patient		#1	#2	#3	#4	#5	#6
Sex (age)	M (69)	F (16)	M (67)	F (34)	F (51)	M (65)	
IgG4-related disease	**Organs involved**	Salivary glands, pancreas, biliary tract, lung	Peritoneum	Pancreas	Eyelid	Periorbital region, lacrimal gland	Meninges, abdominal aorta
	ACR/EULAR CC	36	26	26	20	21	24
	2011 CDC [13]	definite	definite	definite	probable	definite	definite
	Organ biopsy	Lung	Peritoneal lesion	Pancreas	Periorbital tissue	Periorbital tissue, lacrimal gland	Meninges
	Therapy	PDN + RTX	Surgery + PDN + MTX	Surgery	PDN	MPDN + RTX	PDN + MTX
	Response	Y	Y	Y	Y	Y	Y
Systemic Rheumatic Disease	**Diagnosis**	**Sarcoidosis**	**Takayasu**	**RA**	**GPA**	**GPA**	**EGPA**
	Clinical manifestations	Lymph nodes	Subclavian, common carotid, anonymous, and right pulmonary arteries + artery, thoracic and suprarenal abdominal aorta	Wrists	subglottic stenosis	Lung nodules, scleritis	Respiratory tract (asthma), skin, peripheral nervous system, blood eosinophilia
	Fulfillment of disease specific CC	NA	Y[#]	Y^	Y*	Y*	Y[§]
	Autoantibody	none	none	FR, ACPA	c-ANCA	c-ANCA	p-ANCA
	Definite histological diagnosis	Lymph nodes	NA	NA	NA	Lung nodule	Skin
	Onset vs IgG4-RD	Prior	Prior	Concomitant	Prior	Concomitant	Prior
	Years before IgG4-RD	1	14	-	13	0	3
	Therapya	PDN + MTX	IFX → TCZ	PDN + MTX	PDN + AZA	MPDN + RTX	PDN + CYC
	Response	Y	Y	Y	Y	Y	Y

[#]Modified diagnostic criteria for Takayasu arteritis. Shanna et al. 1995; *ACR/ EULAR 2017 provisional classification criteria for GPA; ^ACR/EULAR 2010 Classification criteria for Rheumatoid Arthritis; [§]ACR 1990 Classification criteria for EGPA **Abbreviations**: ACPA: Anti-citrullinated protein antibodies; ANCA: Anti-neutrophil cytoplasmic antibodies; RA: Rheumatoid Arthritis; AZA: Azathioprine; CYC: Cyclophosphamide; EGPA: Eosinophilic Granulomatosis with Polyangiitis; ENG: electroneurography; GPA: Granulomatosis with Polyangiitis; IgG4-RD: IgG4-Related Disease; IFX: Infliximab; TCZ: tocilizumab; F: female; M: male; MPDN: Methylprednisolone; MTX: Methotrexate; N: No; PDN: Prednisone; RTX: Rituximab