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Abstract AB1091 Table 1. The results of the quantitative evaluation of nailfold capillaroscopic findings

	Behçet's patients N=153	Healthy controls N=165	p
Irregularly enlarged capillary score*	0 (0-0.44)	0 (0-0.25)	<0.001
Giant capillary score*	0 (0-0.13)	0 (0-0.13)	0.517
Microhemorrhage score *	0 (0-0.63)	0 (0-0.19)	<0.001
Capillary number score *	0 (0-0.88)	0 (0-0.25)	<0.001
Capillary ramification score *	0 (0-0.75)	0 (0-0.25)	0.224
Disorganize capillary score*	0 (0-0.44)	0 (0-0.25)	<0.001
Total capillaroscopy score*	0.16 (0-1.75)	0 (0-0.50)	<0.001

*Data were given as median (minimum - maximum)

Abstract AB1091 Table 2. The relationship between clinical findings and total capillaroscopy scores

	Total Capillaroscopy Score		p
	Present	Absent	
Vascular involvement (n=54/99) *	0.19 (0-1.48)	0.13 (0-1.75)	0.814
Ocular involvement (n=64/89) *	0.19 (0-1.48)	0.13 (0-1.75)	0.358
Neurological involvement (n=12/141) *	0.19 (0-1.48)	0.13 (0-1.75)	0.222
Genital ulcer (n=131/22) *	0.19 (0-1.75)	0.13 (0-1.48)	0.121
Erythema nodosum (99/54) *	0.13 (0-1.75)	0.19 (0-1.48)	0.026
Populopustular eruption (n=119/34) *	0.19 (0-1.75)	0.13 (0-1.2)	0.238

N, number of patients, *Data were given as median (minimum - maximum)

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AB1093

INTERSTITIAL PNEUMONIA WITH AUTOIMMUNE FEATURES: A SINGLE CENTER EXPERIENCE AND THE IMPORTANCE OF A MULTIDISCIPLINARY APPROACH

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Background: Interstitial lung disease (ILD) remains a significant cause of morbidity and mortality in patients with connective tissue diseases (CTD). Interstitial pneumonia with autoimmune features (IPAF) is a subset of ILD with clinical features suggestive of but do not definitive for a CTD. IPAF is a new concept relatively unknown to rheumatologists. A multidisciplinary approach to diagnosis and management of IPAF patients is essential, involving close interaction between pulmonologists, rheumatologists, radiologists and pathologists.

Objectives: Revision of ILD patients followed at a specialized tertiary hospital's ILD department and description of their clinical characteristics and multidisciplinary approach.

Methods: The study was conducted according to the declaration of Helsinki. All patients who met the Fischer criteria for IPAF in 2000-2018 were identified. Clinical characteristics, comorbidities, ILD subtype, pulmonary function tests, baseline serologies and treatment strategies were collected. The consents from a multidisciplinary meeting and Rheumatology referral and evaluation were also recorded.

Results: We identified 8 cases fulfilling classification criteria for IPAF (4 [50%] female); mean age 64.9 years (range 34-83); past smoking was referred in 5 (62.5%) patients with an average of 54.15 smoking pack years. Overall, 4 (50%) patients were exposed to organic dusts and 2 (25%) to inorganic dusts. Arterial hypertension was the most frequently

recorded comorbidity (50%). Among the 8 patients, 6 (75%) had at least 1 feature from the serologic and morphologic domains, 1 patient had at least 1 feature from clinical and serologic domains and 1 patient had at least 1 feature from all 3 domains. From those meeting "suggestive radiology pattern based on high resolution chest CT (HRCT)", 2 had nonspecific interstitial pneumonia (NSIP) and 1 had organizing pneumonia (OP). Biopsy (3 transbronchial cryobiopsies and 1 transthoracic biopsy) were conclusive in 4 patients (2 NSIP, 1 lymphoid interstitial pneumonia and 1 OP). Usual interstitial pneumonia (UIP) pattern was observed in two patients. Antinuclear antibodies were positive in 5 (62.5%) patients. Overall, 5 (62.5%) clinical cases were discussed in a multidisciplinary meeting including revision of imaging and biopsies. A Rheumatology appointment was requested in 5 patients to investigate a possible CTD diagnosis. Six (75%) patients had pulmonary function tests (PFT) and diffusing capacity of the lung for carbon monoxide (DLCO) results recorded at baseline: 4 patients had a DLCO below 70% (33.8 – 61.8%), 3 patients had normal PFT, 1 had restriction pattern and 2 had small airways obstruction. During a median of 2.7 years of follow-up, none of the patients progressed to a definitive diagnosis of CTD. Pharmacological treatment was prescribed in 6 patients, including corticosteroids (5), DMARDs (3), antifibrotic therapy (2) and azithromycin (2).

Conclusion: IPAF is a relatively new and developing concept. Rheumatologists and pulmonologists should share their experience to uniformize terms and classifications, recognize relevant clinical patterns and optimize management of those affected with CTDs and ILD and IPAF.

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AB1094

ARTERIOSCLEROSIS MAY AFFECT PERIAORTIC/PERIARTERIAL LESIONS AND THEIR ANEURYSMAL CHANGES IN PATIENTS WITH IGG4-RELATED PERIAORTITIS/PERIARTERITIS

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Background: IgG4-related periaortitis/periarteritis (IgG4-PA) is a major organ manifestation of IgG4-related disease (IgG4-RD) [1]. In the affected aorta/artery, arteriosclerotic changes are frequently observed [2]. However, the influence of arteriosclerosis on IgG4-PA and aneurysmal changes of the affected lesions has not been well evaluated.

Objectives: This study aimed to clarify the relationship of arteriosclerosis with periaortic/periarterial lesions and their aneurysmal changes.

Methods: We retrospectively investigated the medical data, including the presence of IgG4-PA at diagnosis, new development of IgG4-PA during the clinical course, arteriosclerotic and aneurysmal changes of the affected lesions, and classic risk factors for arteriosclerosis in 130 patients with IgG4-RD at a single center in Japan. The relationship of arteriosclerosis with the periaortic/periarterial lesions and their aneurysmal changes was statistically analyzed.

Results: Of the 130 patients with IgG4-RD, 44 comprising 39 men and 5 women (mean age 67.4 years) were diagnosed with IgG4-PA. The mean follow-up period was 72.8 months, and the mean serum IgG4 level at diagnosis was 851 mg/dL. The proportions of males (88.6% vs. 55.8%, $P<0.001$), positive smoking history (81.4% vs. 47.6%, $P<0.001$), and history of malignancy (36.4% vs. 14.0%, $P=0.006$) were significantly higher in the 44 patients with IgG4-PA than in the 86 without it. Arterial wall calcification and/or mural thrombus in the affected lesions were detected in 86.0% of the IgG4-PA patients, some of whom had had these arteriosclerotic changes before the development of periaortic/periarterial lesions. Of the 44 with IgG4-PA, 9 had aneurysmal changes of the affected lesions at the time of the first diagnosis of IgG4-RD. Notably, these 9 patients with aneurysmal changes had a significantly higher incidence of hypertension (77.8% vs. 34.3%, $P=0.027$) and mural thrombus in the affected lesions (100% vs. 64.7%, $P=0.044$) than the remaining 35 without aneurysmal changes. However, diabetes mellitus, dyslipidemia, smoking history, C-reactive protein level elevation, or arterial wall calcification of the affected lesions did not differ significantly between the two groups.

Conclusion: The present study suggests that arteriosclerotic changes and the classic risk factors for arteriosclerosis may be related to periaortic/periarterial lesions and their aneurysmal changes in IgG4-PA.

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AB1095

THE GENETIC CHAMELEON IN RHEUMATOLOGY'S BACK GARDEN: A 2-CASE PRESENTATION

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Background: Adult Refsum disease is an autosomal recessive disorder, in which phytanic acid cannot be metabolized and it accumulates in plasma and lipid-containing tissues (1). Although its common clinical features are known, patients are displaying symptoms that can mimic an autoimmune pathology, making this disease a diagnostic dilemma.

Objectives: To highlight the overlap between the clinical and serological features of adult Refsum disease with those of a connective tissue disease, in order to achieve the correct diagnosis and make appropriate therapeutic decisions.

Methods: Clinical and laboratory data were evaluated for 2 patients with adult Refsum disease in a rheumatology tertiary department. The data was retrieved from patients' files.

Results: Both patients were females in their 50s with undiagnosed adult Refsum disease. A medical history revealed that both reported progressive nyctalopia, anosmia, ageusia, peripheral vision loss. In addition to these clinical features one of them reported muscle weakness, weight loss and fatigue, displaying periorbital oedema, heliotrope rash, periungual capillary changes. Her muscular enzymes were elevated (creatin-kinase was 3 times elevated and lactat dehydrogenase was 9 times above normal range), along with C-reactive protein (CRP = 2.4mg/dl). Due to clinical features, increased serum enzymes and a recently diagnosed myocarditis, a high suspicion of dermatomyositis was raised, but was disregarded after extensive investigations (immunology tests, muscular biopsy). The other patient had, in addition, sicca features (dry mouth, eyes, skin) with a Schirmer test of 4mm and an elevated CRP (3.23mg/dl). She was referred to the rheumatology department with a high suspicion of Sjogren's syndrome. Immunology and minor salivary gland biopsy were negative. Interestingly, physical examination showed bilateral brachymetacarpia and brachymetatarsia in both patients, suggesting pseudopseudohypoparathyroidism but, after investigations, the diagnosis was excluded.

Both patients underwent ophthalmological examination to further investigate progressive nyctalopia and peripheral vision loss. Specific changes for pigmentary retinopathy were described, which, together with the above neurological changes and skeletal deformities, were highly suggestive of adult Refsum disease (2). Plasma phytanic acid was 41 and 42 times above normal range respectively, confirming the diagnosis.

After dietary restrictions which excluded phytanic acid containing foods, there was an improvement concerning both clinical and serological manifestations. All connective tissue disease relating symptoms, slowly and gradually started to subside. Muscular enzymes and CRP values returned within normal range (3).

Conclusion: High levels of phytanic acid have triggered a series of uncommon symptoms and serological changes resembling a connective tissue disease, making the diagnosis of adult Refsum disease a very tangled, costly and lengthy process. Although this disease affects many body systems, a case of myositis or sicca syndrome hasn't yet been acknowledged in relation to this condition.

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AB1096

CLINICAL SPECTRUM AND TREATMENT OF THE RELAPSING POLYCHONDritis: A SERIES OF 12 CASES

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Background: Relapsing polychondritis (RP) is a systemic autoimmune disease characterized by inflammation of the cartilaginous tissues associated with the appearance of symptoms caused by the involvement of other non-cartilaginous tissues and the presence of systemic symptoms.

Objectives: To describe the clinical characteristics and treatment of patients diagnosed with RP in a third level university hospital.

Methods: Data from RP patients were obtained from the computerized medical history of our center, including demographics, disease phenotype, treatment, laboratory and imaging studies.

Results: A total of 12 patients were included; 66.7% were women, age at diagnosis was 36 [34-52.5] years (median [p25-p75]). The main manifestation observed was auricular involvement (75%) followed by nasal (66.7%), joint (66.7%), and laryngo-tracheo-bronchial disease (50%). All patients were on DMARD's treatment and two thirds of the patients were receiving biological therapy at the time of analysis. Infliximab was the first biological line in all the cases (100%). Other biological therapies used, in order of frequency, were: adalimumab (25%), rituximab (25%) and tocilizumab (12.5%). 50% of the patients associated another autoimmune disease diagnosed before or during RP follow-up: Crohn's disease, epidermolysis bullosa, primary biliary cholangitis, autoimmune hypothyroidism, systemic lupus erythematosus, granulomatosis with polyangiitis and ulcerative colitis. Only one patient (8.33%) associated hematologic disease (myelodysplastic syndrome) and another one died during follow-up.

Conclusion: RP is a rare disease with a broad spectrum of clinical expression, which often requires intensive management to control its manifestations, potentially serious when there is involvement of the tracheo-bronchial tree or is associated with other autoimmune diseases. A large part of our patients require biological therapy at some point in their evolution.

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AB1097

RETROPERITONEAL FIBROSIS- A SINGLE CENTER EXPERIENCE

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Background: Idiopathic retroperitoneal fibrosis (iRPF) is a rare, chronic, progressive disorder of unknown etiology and characterized by the presence of inflammatory and fibrous retroperitoneal tissue that often encompasses the ureters or abdominal organs.