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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)

Disclosure of Interests: None declared

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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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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.