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Background: Coronary periarteritis is one of the clinical manifestations of IgG4-related disease. It can cause serious conditions such as angina and ruptured aneurysms. Therefore, it is important to recognize the clinical and radiological characteristics, which was little known.

Objectives: We report four patients with IgG4-related coronary periarteritis with a systematic literature review.

Methods: We identified four patients with IgG4-related coronary periarteritis at the St. Luke's International Hospital in Tokyo, Japan from 2014 to 2020. A systematic literature review was conducted for English articles on IgG4-related coronary periarteritis cases with a full text or abstract available. We summarized patient demographics, IgG and IgG4 titers, the site and morphological type of coronary lesion, and other organ involvements.

Results: Our 4 cases and 38 cases identified by the literature review were assessed. Coronary artery lesions were detected by a coronary CT in all but two cases. Wall thickening was the most common type of the lesion. Moreover, there were 32 (76.1%) patients with other organ involvements. The commonest other lesion was peri-aortitis in 21 (50.0%) patients. In cases with peri-aortitis, IgG and IgG4 titers were significantly higher than those without peri-aortitis (IgG4; 1540 [705.0, 2570.0] vs 246.0 [160.0, 536.3]; $p = 0.001$, IgG; 3596.5 [2838.3, 4260.0] vs 1779.0 [1288.3, 1992.8]; $p = 0.040$). In addition, 15 (71.4%) patients of them had three or more IgG4 related organ involvements.

Conclusion: Coronary CT was a useful imaging modality for the diagnosis of IgG4-related coronary periarteritis, and wall thickening was the most common lesion. Moreover, about half cases coexisted with peri-aortitis. Peri-aortitis and other organ involvements should be screened in those with higher IgG and IgG4.

Table 1. Characteristics of our cases and the literature review cases.

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	All cases (n=42)	The number of organ involvements	Lesion distribution	Total number of coronary lesions (n=126)
Age (years)	64.0 (58.6, 69.6)			
Male (%)	35 (83.3)	3-three (%)	20 (47.6)	45 (35.7)
Female (%)	7 (16.7)	1-one (%)	12 (28.6)	44 (38.1)
IgG4 (mg/dL)	625.0 (214.0, 2090.0)	2-two (%)	44 (38.1)	
IgG (mg/dL)	2843.5 (1672.5, 4138.6)	3-three (%)	35 (29.2)	
Diagnostic criteria		With other organ involvements	Lesion type	
possible (%)	10 (23.8)	all cases (%)	stenosis (%)	28 (22.2)
probable (%)	9 (21.4)	peri-aortitis (%)	wall thickening (%)	70 (55.6)
definite (%)	23 (54.8)	gland (%)	aneurysm (%)	28 (22.2)
		pancreas (%)		
		lymph (%)		
		kidney (%)		
		retroperitoneal (%)		
		pulmonary (%)		
		colon (%)		
		Coronary branch lesion counts		
		none (%)	14 (33.3)	
		double (%)	31 (23.8)	
		triple (%)	14 (33.3)	

RCA: right coronary artery, LAD: left anterior descending artery, LCx: left circumflex artery

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2021-eular.1293

AB0764 MANIFESTATIONS OF BEHCET'S DISEASE

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Background: Behcet's disease (BD) is a systemic vasculitis of unknown etiology, characterized by recurrent oral and genital erosions and ulcerations, as well as involvement of joints, ocular, vascular, gastrointestinal (GI), and central nervous system (CNS).

Objectives: To study the influence of such factors as patient's gender, age at the disease onset and initial manifestations of the disease on the severity of BD.

Methods: 95 patients with confirmed BB were examined. The majority of patients were males (70 men, 25 women) with the 2.8:1 M:W ratio. Patients' mean age (median [interquartile range]) was 29 years [22; 34], from 16 to 51 years, the average age at disease onset was 20 years [14; 24]; and mean disease duration was 8 years [4; 15].

Results: The following clinical manifestations of BD were documented at disease onset (%): recurrent aphthous stomatitis – 63.1; skin lesions – 12.6; genital ulcers – 4.2; ocular lesions – 6.3; joint syndrome -7.4; CNS lesions -2.1; gastrointestinal tract involvement -2.1; thrombosis -1; epididymitis -1. 11.6% of all patients had signs of organ system involvement by the time of establishing BD diagnosis. Recurrent aphthous stomatitis was more common in female patients at BD onset– 76% vs. 58.6%, $p=0.01$, while organ system involvement was more common in males-14% vs. 4% in females, $p=0.02$.

Progression from initial symptoms to the mature BD phenotype allowing to verify the diagnosis occurred within 4.3 years on average [1; 6], although 30.5% of patients progressed to a full-blown disease within 1 year.

Analysis of potential predisposing or precipitating factors prior to BD onset identified such in only 12 patients, including frequent episodes of tonsillitis in 3 patients, cold exposure with hypothermia in 2, dental procedures in 2, stressful events - in 2, and one case of each - herpes labialis, frontal sinus puncture and appendectomy. Other patients could not recall any potential precipitator of BD.

Mean age at BD onset was 20 years [14; 24], including 30.5% of patients with disease onset before 16 years, 22.1% aged from 16 to 21 years, 36.8% aged from 21 to 30 years at disease onset, and in remaining 10.5% the disease manifested after 30 years of age. BD onset in men and women was documented at approximately the same time. There were no signs or correlations indicating that patient's age at disease onset could be associated with specific clinical symptoms in males or females. However, analysis of gender-associated differences in disease severity based on Krause's Clinical Severity Scoring yielded the following results: mild BD was found in 15.7% men and 44% women ($p=0.009$), moderate disease - in 7.1% men and 16% women, ($p>0.05$), and severe BD - in 77% men and 40% women, respectively, ($p=0.001$). The risk of developing severe BD was 5-fold higher in men vs women [OR = 5.06, 95% CI 1.7-15.1].

Conclusion: First BD symptoms manifested in the majority of patients before the age of 30 years, with only 10% of patients with BD onset at the age of > 30 years. In more than 50% of all patients aphthous stomatitis was the first manifestation of the disease, and was more common in women. Prognostically unfavorable BD signs were found in 11.6% of patients already at the initial stages of the disease in men. No correlation was found between the age at BD onset and clinical symptoms of the disease in both males and females.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2021-eular.1364

AB0765 PHENOTYPES OF BEHCET'S DISEASE IN DIFFERENT ETHNIC GROUPS

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Background: Early diagnosis and identification of predominant organ system involvement in Behcet's disease (BD) is crucial for identifying most optimal treatment. The study evaluates 6 described phenotypes of BD - cutaneous-mucous, articular, ophthalmic, vascular, neurological and intestinal in different ethnic groups.

Objectives: To assess the occurrence of Behcet's disease (BD) phenotypes in different ethnic groups.

Methods: The study included 202 patients with BD from the 5 most common ethnic groups.

The male-female ratio was 2.4:1. Patients' mean age was 31 years [24;37], mean age at the disease onset was 21 years [15;28]; and mean disease duration was 7 years [3;14]. The severity of BD (mild, moderate and severe) was assessed based on the I. Krause's Clinical Severity Scoring for BD.

Results: Severe BD was more often diagnosed in Azerbaijanis and indigenous residents of Dagestan compared to Russians (75 and 70.4% vs. 36.2%), in Armenians - 50% and Chechens - 54.5% out of all BD cases.

Russians were significantly more likely to have a neurological phenotype (15.5% vs. 0-9.4% in all other ethnic groups) and intestinal phenotype (36.2% vs. 13.8-22.7% in all other ethnic groups). Azerbaijanis demonstrated higher prevalence of ocular involvement (68.7% versus 36.2% in Russians, 50% - in Chechens and Armenians, and 57% - in Dagestanis). Dagestanis were more likely to have a vascular phenotype (40.7% versus 15.6% in Azerbaijanis and 18.9% in Russians). The male/female ratio among Russian patients was 1:1, among Dagestanis 4.4:1, Azerbaijanis 3.5:1, Chechens and Armenians 2.6: 1.

Conclusion: BD phenotypes vary and demonstrate significant association with the patient's ethnic affiliation therefore, ethnicity should be viewed as the prognostic marker of specific organ-system involvement in case of a disease.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2021-eular.1373

AB0766 SUCCESSFUL TREATMENT OF ANKYLOSING SPONDYLITIS ASSOCIATED AA AMYLOIDOSIS WITH SECUKINUMAB: A CASE SERIES WITH THREE PATIENTS

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