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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)				Total number of coronary lesions (n=126)
Age (years)	64.0 [58.0, 69.8]	The number of organ involvements		Lesion distribution	
Male (%)	35 (83.3)	≥ three (%)	20 (47.6)	RCA (%)	45 (35.7)
IgG4 (mg/dL)	625.0 [214.0, 2090.0]	two (%)	12 (28.6)	LAD (%)	48 (38.1)
IgG (mg/dL)	2841.5 [1672.3, 4138.8]	one (%)	10 (23.8)	LCx (%)	33 (26.2)
Diagnosis criteria		With other organ involvements		Lesion type	
possible (%)	10 (23.8)	all cases (%)	32 (76.1)	stenosis (%)	28 (22.2)
probable (%)	9 (21.4)	peri-aortitis (%)	21 (50.0)	wall thickening (%)	70 (55.6)
definite (%)	23 (54.8)	gland (%)	12 (28.6)	aneurysm (%)	28 (22.2)
		pancreas (%)	9 (21.4)		
		lymph (%)	6 (14.3)		
		kidney (%)	4 (9.5)		
		retroperitoneal (%)	4 (9.5)		
		pulmonary (%)	2 (4.8)		
		colon (%)	1 (2.4)		
		Coronary branch loison counts			
		mono (%)	14 (33.3)		
		double (%)	11 (23.8)		
		triple (%)	16 (38.1)		

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 tonsilitis 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 TEATMENT OF ANKYLOSING SPONDILITIS ASSOCIATED AA AMYLOIDOSIS WITH SECUKINUMAB: A CASE SERIES WITH THREE PATIENTS

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Background: Systemic AA amyloidosis is a serious and life-threatening complication of chronic inflammatory diseases such as rheumatoid arthritis, spondyloarthritis (SpA), and periodic fever syndromes. While most common cause of AA amyloidosis is Familial Mediterranean Fever; Ankylosing Spondylitis (AS) is another frequent cause of AA amyloidosis in Turkey.
Objectives: We aimed to evaluate the response of secukinumab (SEC) treatment in three patients with AS and AA amyloidosis (AS-AA) in our tertiary referral centre.
Methods: We retrospectively evaluated three AA amyloidosis patients who fulfilled Modified New York Criteria for diagnosis of AS in our AA amyloidosis cohort with 163 patients. Diagnosis of AA amyloidosis was confirmed by Congo red stain and by monoclonal AA-specific antibodies.
Results: *Patient 1:* 61-year-old male patient with inflammatory back pain (IBP) and peripheral arthritis for 14 years was evaluated in our clinic. After methotrexate (MTX) failure, he used adalimumab (ADA), etanercept (ETA) and certolizumab (CZP). Nephrotic range proteinuria was detected when he was on CZP, and rectum biopsy documented AA amyloidosis 3 years ago. After the diagnosis, CZP treatment was switched to infliximab (IFX). IFX was ineffective in controlling inflammatory findings. SEC was started 15 months ago and he responded partially. The dose of SEC was increased to 300 mg monthly, which resulted in a sustained improvement in clinical and laboratory findings.
Patient 2: 69-year-old woman was admitted to our clinic with peripheral arthritis in addition to the history of IBP for 19 years in 2005. MTX, NSAID and prednisolone were started. Because of inefficacy to conventional treatments and development of nephrotic range proteinuria, ETA was added to treatment. The patient responded to ETA and was followed-up for 13 years without symptoms of AS and proteinuria. ETA was switched to IFX due to secondary inefficacy two years ago. On the third month of IFX treatment, she developed demyelinating polyneuropathy. IFX treatment was switched to SEC and she is still being followed-up on SEC without any findings of AS and proteinuria.
Patient 3: 49-year-old woman who was on sulphasalasine for 24 years for treatment of ulcerative colitis (UC) was evaluated for recent onset IBP and peripheral arthritis in 2007. After failure of MTX, she started to receive IFX. She did not respond to first IFX and then ADA and CZP, and she developed nephrotic range proteinuria when she was on anti-TNF. Her serum creatinine increased progressively, and haemodialysis (H/D) was started six months later. Due to ongoing IBP and elevated acute phase response with CZP treatment, SEC was started. Significant improvement was observed in both clinical and laboratory findings with no worsening of UC.

Table 1. Clinical characteristics, laboratory findings and treatment responses of patients

	Patient 1	Patient 2	Patient 3
Age (year)	61	69	49
Sex	Male	Female	Female
Age of AS onset (year)	47	35	25
Age of amyloidosis diagnosis (year)	57	50	46
Amyloidosis duration (months)	42	168	30
Family history	No	AS and amyloidosis in siblings	No
HLA-B 27 status	Positive	Positive	Not available
MEFV status	Negative	Negative	M694V heterozygous
Organ involvement of amyloidosis	GIS and kidney	Kidney, liver, heart, bone marrow	GIS, kidney
Secukinumab duration (months)	15	18	24
CRP (mg/L)			
Before	27	15	23
After	2.4	4.5	2.8
Creatinine (mg/dl)			
Before	0.5	1.6	H/D
After	0.5	1.3	H/D
Proteinuria (g/day)			
Before	4.2	5.5	H/D
After	2.4	0.9	H/D
Albumin (g/dL)			
Before	2.7	2.5	3.1
After	3.1	4.3	4.2
ASDAS			
Before	4.1	1.6	4.6
After	1.8	1.1	1.7

GIS: Gastrointestinal System, ASDAS: Ankylosing Spondylitis Disease Activity Score

Conclusion: AA amyloidosis is a rare complication of SpA, and SEC treatment was found to be safe and effective in our three patients with AS-AA. Although anti-TNF agents have previously used successfully in treatment of AS-AA, SEC may be a new option especially in patients who are resistant or intolerant to anti-TNFs.

Disclosure of Interests: None declared
DOI: 10.1136/annrheumdis-2021-eular.1469

AB0767 SEVERE BREAST ULCERATION IN BEHÇET'S DISEASE TREATED WITH CYCLOPHOSPHAMIDE: A CASE REPORT

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Background: Behcet's disease is a systemic autoimmune vasculitis that typically involves mucocutaneous surfaces, the eyes and the skin with varying manifestations. Skin ulceration often secondary to pathergy is a common feature.
Objectives: To raise awareness of breast ulceration as a clinical manifestation in Behcet's disease.
Methods: We report a case of complex Bechet's disease associated with severe refractory breast ulceration successfully treated with cyclophosphamide.
Results: A 27 year old female patient presented to the rheumatology department with a non-healing lesion on her face secondary to a cat scratch. She reported previous episodes of ulceration usually as a result of minor trauma such as IV cannulation, as well as a two year history of severe oral and genital ulceration. She had also been diagnosed with 'colitis' requiring iliocaecal resection two years prior to presentation. Previous treatments for her bowel included azathioprine, infliximab, adalimumab and vedolizumab. Over the next three years she developed recurrent eye involvement in the form of scleritis with ongoing oral and genital ulceration. Further treatment under our care included ustekinumab, entanercept, benepali, certolizumab and tacrolimus with background prednisolone. She then developed a small area of broken skin on her left breast which continued to extend and ulcerate despite treatment with IV steroids. Tissue biopsy showed deep focal vasculitis with intravascular thrombi. Anticoagulation was commenced and she was treated with tocilizumab IV for 3 months with no clinical improvement. Involvement extended to include 80 percent of the breast surface and the nipple self-amputated. Due to the severity and extent of the ulceration which was extremely painful and distressing, she was given 3 months of IV cyclophosphamide at a dose of 15mg/kg every 2-3 weeks. The ulceration showed rapid clinical improvement (see Figure 1).

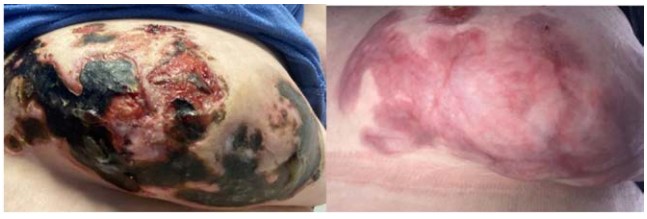

Figure 1. Breast ulceration before and after 3 months of cyclophosphamide treatment

Conclusion: Involvement of the breast in systemic vasculitides (such as GPA or PAN) has been reported, although usually manifesting as a palpable mass with diagnosis on biopsy [1]. Similar presentations in Behcet's disease have been recognised [2], although much less commonly. Our case demonstrates that severe breast ulceration can be a clinical manifestation of this condition and can be refractory to usual therapies.
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Disclosure of Interests: None declared
DOI: 10.1136/annrheumdis-2021-eular.1639

AB0768 NEUROLOGICAL MANIFESTATIONS IN GRANULOMATOSIS WITH POLYANGIITIS: ABOUT 11 CASES

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Background: Granulomatosis with polyangiitis (GPA) is a rare systemic vasculitis, affecting mainly the superior airways, lungs and kidneys. Neurological impairment is frequent, described in 22 to 54% of cases, and rarely inaugurates the disease.