Objectives: Summarizing the clinical characteristics of IgG4-related diseases to improve the understanding of clinicians and achieve early diagnosis and reasonable treatment.

Methods: The clinical data of 15 patients with IgG4-related diseases diagnosed in our hospital from 2014 to 2018 were retrospectively analyzed.

Results: The ratio of male to female was 2:1 in 15 patients. The average age of onset was 56.8 years (34 to 84 years of age). From the first symptom to the diagnosis of 8 days to 4.75 years, the most common first symptom in 15 patients was Submandibular gland swelling, pain, anorexia, cough, the most common affected organs are the pancreas, lungs, submandibular glands. There were 11 cases of histopathological examination by surgery or needle biopsy. The histopathology showed interstitial fibrous tissue hyperplasia with more lymphocyte and plasma cell infiltration. Immunohistochemistry showed that the number of IgG4-positive cells was >50/HPF, and IgG4/IgG was >40%. Two of the 15 patients underwent surgical resection of the affected organ occupying lesions before definitive diagnosis. One patient was treated with mycophenolate mofetil and one patient was given a medium dose of hormone combined with tamoxifen. The remaining 13 patients were treated with simple hormones or combination immunosuppressive agents, including hydroxychloroquine, mycophenolate mofetil, azathioprine, and cyclophosphamide.

Conclusion: The organs involved in IgG4-related diseases are diverse, the clinical manifestations are not specific, and the hormone combined immunosuppressive therapy is effective.

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AB0629

CIRCULATING ENDOTHELIAL CELLS MAY BE A MARKER FOR VASCULAR INVOLVEMENT IN BEHCET DISEASE

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Background: Circulating endothelial cells (CEC) are defined in conditions which vascular damage is seen in course of disease such as systemic vasculitis, coronary artery disease and chronic renal failure. CEC is thought to be an indicator of vascular damage (1).

Behcet disease (BD) is a systemic vasculitis mostly known with recurrent oral and genital ulceration, uveitis and mucocutaneous lesions. On the other hand vascular involvement such as deep venous thrombosis, cerebral sinus thrombosis and pulmonary artery aneurysm, is an important clinical finding of disease which may cause mortality (2).

Objectives: Our aim in this study was to analyse CEC levels in patients with Behcet disease, to compare them between patients with vascular (Group 1) and mucocutaneous (Group 2) involvement. Also we have compared the results of Behcet patients with patients with thrombosis due to other causes (Group 3) and healthy controls (Group 4). Each group involved 20 participant.

Methods: Blood samples of the patients and healthy controls are drawn into tubes containing ethylene-diamine-tetra-acetic acid (EDTA). A panel of monoclonal antibodies, including anti-CD45 to exclude hematopoietic cells, anti-CD31, -CD34, -CD36, -CD105, -CD106, -CD133, and -CD 146 and appropriate analysis gates were used to enumerate resting and activated CECs and circulating and endothelial progenitor cells (CEP).

A hundred microlitre complete blood was added and incubated for 20 minutes at room temperature in the dark. After incubation for 10 minutes with erythrocyte lysing solution at room temperature, centrifugation at 1,800 rpm for 5 minutes was performed. Supernatant was removed and washed with phosphate buffer saline (PBS) for two times. Pellet was resuspended with PBS and 300.000–400.000 cells were counted with FACSCalibur flow cytometry device.

Results: Mean age, sex distribution and duration of disease were similar in all groups (Table 1).

Table 1. Demographic features of the study population

	Group				p
	Group 1	Group 2	Group 3	Group 4	
	mean ± standard deviation				
Age	43.55 ± 8.31	46.85 ± 9.43	42.50 ± 15.22	41.65 ± 6.54	0.323**
Duration of disease	14.90 ± 8.69	11.30 ± 8.80	-	-	0.109*
	n (%)				
Sex					
Female	9 (% 45.0)	12 (% 60.0)	11 (% 55.0)	14 (% 70.0)	0.447***
Male	11 (% 55.0)	8 (% 40.0)	9 (% 45.0)	6 (% 30.0)	

* Mann Whitney U Test

** Kruskal Wallis H Test

**** Pearson Chi Square Test

CEC levels did not show a statistically significant difference between all groups. CEPs, activated CECs (aCEC) and resting CECs (rCEC) were also compared between groups. CEPs were higher in Behcet patients with thrombosis similar to patients with thrombosis due to other causes (p:0.042). Activated CECs levels did not show a difference between groups (p>0.05).

Resting CECs are higher in Groups 1 and 3 than Groups 2 and 4. The detailed analysis of CEC, CEP, activated and resting CECs between groups is listed in Table 2.

Abstract AB0629 Table 2. Comparison of CEC, CEP, aCEC and rCECs between groups

	Group				p	Multi analysis
	Group 1	Group 2	Group 3	Group 4		
	mean ± standard deviation					
CEC	5.09 ± 4.7	2.52 ± 2.55	4.89 ± 3.83	4.09 ± 4.9	0.077**	-
CEP	13.49 ± 9.38	7.62 ± 4.02	15.6 ± 11.21	13.71 ± 10.1	0.042**	1-2: 0.015 2-3: 0.009
aCEC	4.39 ± 5.78	2.24 ± 2.14	8.17 ± 13.26	8.78 ± 13.21	0.054**	-
rCEC	6.44 ± 5.4	5.43 ± 3.49	9.03 ± 7.79	3.52 ± 2.34	0.007**	1-4: 0.034 3-4: <0.001

Conclusion: Increased levels of aCECs may be an indicator of active vascular involvement in BD. But in the current study aCEC levels did not show a difference between groups but none of the patients had active vascular involvement which may cause of this sameness. CEP and resting CEC levels were elevated in both groups of patients with thrombosis. So CEC may be a marker for vascular damage but it is not specific for BD.

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AB0630

CLINICAL CHARACTERISTICS ANALYSIS AND LITERATURE REVIEW OF 55 CASES OF PANNICULITIS

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Background: Panniculitis is a heterogeneous inflammatory disease involving subcutaneous fat. It can be divided into different subtypes according to the clinical characteristics and pathological changes of the disease. Because the cause of panniculitis is unclear, clinical manifestations are diverse, lack of specificity, early diagnosis is difficult, and misdiagnosis and missed diagnosis are prone to occur.

Objectives: To improve the clinical understanding of the disease by retrospective analysis of 55 cases of patients with panniculitis.

Methods: The hospitalized patients with panniculitis were collected from December 2011 to October 2018 in the Shanxi Dayi Hospital Affiliated to Shanxi Medical University. The demographics, clinical manifestations, auxiliary examinations and treatments were analyzed and summarized.

Results: The proportion of males and females in the 55 patients was 1:2.23, with an average of 53.3 years (18-82 years). A total of 52 cases

of nodular panniculitis (including 14 cases of mesenteric involvement) and 3 cases of special type of panniculitis were collected. Patients with nodular panniculitis often find subcutaneous nodules or masses. According to the affected parts, they are divided into skin type and system type. Among them, 10 cases are skin type and 42 cases are system type. The most common parts is mesenteric and kidney involvement. 9 of 55 patients was combined with tumor. Patients with mesenteric panniculitis mainly present abdominal pain and bloating, which are mainly diagnosed according to CT. Patients with systemic panniculitis can be significantly relieved after treatment with hormones and immunosuppressive agents.

Conclusion: Panniculitis is an inflammatory disease of unknown etiology. It is common in middle-aged and elderly people. It has various clinical manifestations and lacks specificity. The diagnosis is mainly based on pathological results. It is easy to be combined with tumors. When subcutaneous nodules are found, pathological examination should be improved in time in order to avoid misdiagnosis.

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AB0631 INTERSTITIAL LUNG DISEASE – A RARE MANIFESTATION OF MICROSCOPIC POLYANGIITIS

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Background: Interstitial lung diseases (ILD) are a group of diffuse inflammatory and/or fibrotic lung disorders with similar clinical, radiologic and histopathologic features. The coexistence of ILD and ANCA-associated vasculitis has been reported in case reports and small case series.

Objectives: The aim of this study was to assess the prevalence, clinical and radiological characteristics of ILD in MPA patients admitted to the Rheumatology Department of our hospital.

Methods: This retrospective single center cohort study included 102 patients diagnosed with MPA according to CHCC 2012 and EMA algorithm. We assessed Birmingham Vasculitis Activity Score (BVAS) at disease onset and VDI (Vasculitis Damage Index) at the end of the follow up in each patient. ANCA type and titer were assessed by enzyme-linked immunosorbent assay (ELISA). The results of repeated high resolution computed tomography (HRCT) of the chest were analyzed and interstitial changes (focal ground-glass opacity, reticular changes, honeycombing, traction bronchiectasis) were classified in four patterns: non-specific interstitial pneumonia (NSIP), typical interstitial pneumonia, possible interstitial pneumonia, and “inconsistent” usual interstitial pneumonia (UIP).

Results: ILD-features on HRCT was found in 11 (11.8%) of 102 patients with MPA. Their median age was 55 [53; 63] years. All of them were ANCA-positive (Table 1). In 5 cases interstitial pneumonia was the first and the sole manifestation that preceded the occurrence of overt systemic vasculitis for a median of 3 years. The radiologic patterns included non-specific interstitial pneumonia in 5 cases, usual interstitial pneumonia in 3 cases and unclassifiable interstitial pneumonia in 3 cases. The most common clinical manifestations were non-productive cough (82%), progressive dyspnea (82%) and crepitation (27%). The most common extrathoracic manifestations of MPA in patients with ILD were glomerulonephritis with decreased renal function (91%), fever (100%) and arthritis (91%). All patients received induction therapy with glucocorticosteroids combined with cyclophosphamide, rituximab, azathioprine or methotrexate.

Abstract AB0631 Table 1.. Characteristics of patients with MPA and ILD

	n = 11
Male gender, n (%)	7 (64)
Age of the onset of ILD, Me, IQR, years	55 [53; 63]
Age of the onset of MPA, Me, IQR, years	59 [54; 65]
ILD onset	
Before systemic manifestations of MPA, n (%)	5 (45)
At MPA onset, n (%)	6 (55)
ANCA specificity	
MPO-ANCA, n (%)	6 (55)
PR3-ANCA, n (%)	2 (18)
unidentified ANCA, n (%)	3 (27)
Clinical symptoms	
Fatigue, n (%)	11 (100)
Fever, n (%)	11 (100)
Weight loss, n (%)	4 (36)
Arthralgia, n (%)	10 (91)
Skin vasculitis, n (%)	5 (45)
Renal manifestations, n (%)	10 (91)
Ear, nose and throat involvement, n (%)	4 (36)
Chronic cough, n (%)	9 (82)
Hemoptysis, n (%)	2 (18)
Dyspnea, n (%)	9 (82)
Crackles, n (%)	3 (27)
Laboratory and instrumental findings	
Increasing of ESR and/or CRP, n (%)	10 (91)
Anemia, n (%)	7 (64)
24-hours proteinuria, g/day	0.61 [0.25; 1.95]
Estimated GFR, ml/min/1.73 m ²	39 [19; 60]
Radiological patterns	
UIP, n (%)	3 (27)
NSIP, n (%)	5 (45)
Unclassified IP, n (%)	3 (27)
Mediastinal lymphadenopathy, n (%)	3 (27)
Emphysema, n (%)	2 (18)
Remission induction therapy	
Glucocorticosteroids, n (%)	11 (100)
Cyclophosphamide, n (%)	7 (64)
Rituximab, n (%)	1 (9)
Azathioprine, n (%)	1 (9)
Methotrexate, n (%)	1 (9)

Conclusion: ILD is a rare manifestation of MPA, which can precede systemic manifestations. ANCA-associated vasculitis should be included in the spectrum of differential diagnosis in patients with ILD.

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AB0632 MAINTENANCE RITUXIMAB IN ANCA ASSOCIATED VASCULITIS – OBSERVATIONAL DATA FROM UNIVERSITY HOSPITAL COVENTRY, UK

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Background: Rituximab is considered an effective¹ maintenance therapy in anti-neutrophil cytoplasmic antibody (ANCA) associated vasculitis (AAV), with reduced relapse rates post induction therapy and comparable adverse event profile to other agents such as Azathioprine².

Objectives: To describe the clinical outcomes of all patients receiving maintenance Rituximab for AAV in University Hospital Coventry in UK.

Methods: Records from the Renal and Rheumatology units were used to identify patients receiving maintenance Rituximab. Electronic patient records (EPR) were used to collate laboratory and clinical data.