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.

REFERENCES

- Umehara H, Okazaki K, Masaki Y, et al. Comprehensive diagnostic criteria for IgG4-related disease (IgG4-RD), 2011[J]. Japanese Journal of Rheumatology, 2012, 22(1):21-30.
- Okazaki K, Kawa S, Kamisawa T, et al. Japanese clinical guidelines for autoimmune pancreatitis[J]. Pancreas, 2009, 38(8):849-866.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.7493

AB0629 CIRCULATING ENDOTHELIAL CELLS MAY BE A MARKER FOR VASCULAR INVOLVEMENT IN BEHCET DISEASE

Nazife Sule Yasar Bilge¹, Eren Gunduz¹, Meltem Akay², Timuçin Kaşifoğlu¹.

¹Eskişehir Osmangazi University, Eskişehir, Turkey; ²Koç Üniversitesi, İstanbul, Turkey

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.

REFERENCES

- Woywodt A Circulating endothelial cells in vasculitis and transplantation. Pathophysiol Haemost Thromb. 2003;33:500-2
- Kutlay S, Calayoglu R, Boyvat A, Turkcapar N, Sengul S, Keven K, Nergizoglu G. Circulating endothelial cells: a disease activity marker in Behçet's vasculitis. Rheumatol Int 2008;29:159-162

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.6101

AB0630 CLINICAL CHARACTERISTICS ANALYSIS AND LITERATURE REVIEW OF 55 CASES OF PANNICULITIS

¹Hui Zheng^{*}, Li Rong¹, Guozhu Che¹, Liyun Zhang¹, Huiqin Hao². ¹Shanxi Da Yi Hospital, Rheumatology, Tai Yuan, China; ²Shanxi University of Chinese Medicine, Jin Zhong, China

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