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Background: Giant cell arteritis (GCA) and polymyalgia rheumatica (PMR) are closely related inflammatory conditions affecting people aged over 50 years.
Objectives: We present our experience of using tocilizumab (TCZ) therapy for management of GCA/PMR aggravated by severe concurrent pathologies that potentially increase the risk of side effects of glucocorticoids (GCs).
Methods: 22 patients were recruited into the prospective study: six patients with GCA, 13- PMR, and three- with both GCA and PMR, 95.5% were females, mean age 72.8±6.5 years. Mean disease duration was 3.5 (0.5-19) months. All patients had active GCA/PMR with mean CRP 30.3±32.7 mg/l. Seven patients had visual ischemic complications, and another one- aortitis. All patients had serious comorbidities, 59% of patients had three and more severe concurrent diseases. All patients were administered TCZ i/v 2.3-8.8 mg/kg Q4W. 50% patients were also treated with prednisone at mean 20 (10-70) mg/day. The follow-up period was 24 (6-60) months.
Results: All patients demonstrated good clinical response to TCZ i/v 2.3-8.8 mg/kg Q4W given for average 4,5 (2-11) months, achieving remission in 100% of cases. Some patients showed a very rapid improvement after initiation of treatment, including TCZ monotherapy. Prednisone dose was discontinued in 6/11, or was reduced to 2.5 (2.5-10) mg in 4/11. There was one relapse after TCZ discontinuation, although this patient managed to regain the remission after resumption of TCZ i/v 4 mg/kg. There was one (4.6%) serious complication (septic olecranon bursitis 1 month after TCZ discontinuation), one patient died of myocardial infarction 12 months later after TCZ discontinuation. Three remaining complications included one case of peripheral artery disease (claudication), one- psoriasis, and one- sural lipodermatosclerosis.
Conclusion: Interleukin-6 inhibitors should be considered as potentially effective and relatively safe treatment for GCA/PMR patients with serious comorbidities, intolerance or contraindications to standard therapy. More data is necessary to identify the optimal dosing regimen and duration of TCZ therapy, as well as cost-effectiveness aspects.
Disclosure of Interests: None declared
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AB0461 ANCA-ASSOCIATED VASCULITIS: CLINICAL FEATURES, RELAPSE, ORGAN DAMAGE AND SURVIVAL IN 197 PATIENTS

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Background: ANCA-associated vasculitis (AAV) is a multisystemic autoimmune disease with high mortality and morbidity.
Objectives: We aimed to present the long-term follow-up results of our cohort.
Methods: Data of patients who fulfilled Chapell Hill Consensus Criteria and followed up at least 6 months between 1999-2019 were analyzed. A standard form including vasculitis damage index (VDI) was used. Multivariable analysis was performed by using logistic regression.
Results: Long-term data was available for 197 patients (%53.8 female) from 208 patient records. Mean age at diagnosis was 49.4 years and mean follow-up was 80.7 months. Granulomatosis with polyangiitis (GPA); microscopic polyangiitis (MPA), eosinophilic GPA (EGPA) were 117 (64.5%), 52 (26.4%), 17 (8.6%), respectively. Relapses are observed in 31.6% of patients. Disease relapses were higher in GPA compared to MPA and EGPA (p = 0.014). Relapse rate was higher in patients with *s.aureus* carriage (p = 0.037). Cyclophosphamide (CYC) (76.6%) was most commonly used drug for induction, whereas azathioprine (57.3%) was used mostly in maintenance. In multivariate analysis relapse was found to be associated with maintenance treatment with rituximab (p <0.001), venous thrombosis (p=0.046) and serious infection (p<0.004). There was no significant association between relapse and mortality. Five-year survival rates were 98.5% for GPA, 88.5% for MPA and 100% for EGPA. Nineteen patients died during follow-up (9.6%). In univariate analysis mortality were high in MPA patients. Low hemoglobin and increased creatinine at baseline, subglottic stenosis, polyneuropathy, and cerebrovascular events (CVE) were associated with increased mortality. In multivariable analysis, mortality was associated with CVE (p=0.047) and anti-MPO positivity (p=0.014). Malignancy was developed in 9 patients (M / F: 7/2; two lung, three bladder, one cervix, one thyroid papillary, one kidney and one of unknown primary). There was no association between malignancy and cumulative dose of CYC. Venous thromboembolism was developed in 12 (6 %) and avascular necrosis (AVN) was detected in 30 patients (15.4%). Most (88.7%) patients developed damage during follow-up. Mean VDI score was 2.6 and VDI score was found to be higher in GPA (p= 0.035). There was no association between VDI score and mortality.
Conclusion: In our AAV cohort, GPA was most frequent. Although survival was improved, permanent organ damage was detected in the majority of patients. Relapse and organ damage were found to be increased in patients with GPA. Relapses are frequent and maintenance with rituximab could not prevent relapses. Also relapses were associated with venous thrombosis and severe infections. Patients should be screened for malignancies especially of the genitourinary tract.

Table 2. Damage findings of AAV patients according to VDI

Organ/system	Number(%)
Steroid myopathy	23 (%11.7)
Osteoporosis	31 (%15.9)
AVN	30 (%15.4)
Cataract	30 (%15.4)
Partial loss of vision	6 (%3.1)
Blindness (one eye)	2 (%1)
Subglottic stenosis	9 (%4.5)
Hearing loss	18 (%9.1)
Nasal septum perforation	21 (%10,7)
Chronic nasal crusting	9 (%4,6)
Chronic ashtma	28 (%14,2)
Chronic dispnea	1 (%0,5)
Hypertension	60 (%30,5)
Coronary artery disease / Angioplasty	10 (%5,1)
Cardiomyopathy	6 (%3)
Valvular heart disease	5 (%2,5)
Myocardial infarction	7 (%3,6)
Deep vein thrombosis	12 (%6)
Chronic renal failure (GFR <50 ml/min)	51 (%26)
End stage renal disease	22 (%10.8)
Cerebrovascular accident	9 (%4,4)
Peripheric neuropathy	39 (%19.8)
Malignancy	9 (%4.5)
Diabetes mellitus	24 (%12.2)
Gonadal failure	2 (%1)

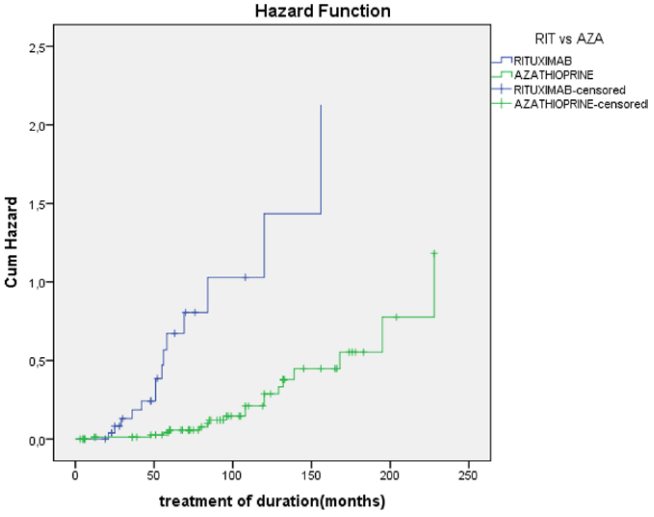

Figure 3. Cumulative Relapse Rate: Hazard ratio of patients treated with Rituximab versus Azathioprine (Log Rank: p<0.001)

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AB0462 BEHCET'S DISEASE: CLINICAL FEATURES AND OFF-LABEL BIOLOGIC TREATMENT STRATEGIES

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Background: The treatment of Behçet's disease (BD) is still mainly based on the evidence derived from case reports, case series, retrospective analyses, and few clinical trials suggesting the safety and potential efficacy of off-label use of biologic agents in refractory cases. [1]
Objectives: To describe clinical manifestations and their management, with particular focus on treatment indications, outcomes and safety of biologic therapy, in a cohort of patients with BD.

Methods: Patients with a diagnosis of BD who visited our outpatient clinic until December 2019 were included in the study. Clinical data were recorded since diagnosis until the latest follow-up visit, analyzing clinical features, flares and therapeutic strategies adopted.
Results: A total of 95 patients were included in the study with a medium follow-up of 108.54 ± 169.59 months. 20 of them (21. 05%) were treated with biologic agents. Patients treated with biologic therapy compared to those on conventional non-biologic therapies had a higher proportion of musculoskeletal (80% vs 46.67%, p = 0.008), neurological (30% vs 10.67%, p = 0.031), intestinal involvement (40% vs 12%, p = 0.004), and they were treated with a higher dose of glucocorticoids at diagnosis (16.84 mg ±14.01 vs 8.89 mg ± 11.76, p = 0.012). The most frequent indications for biologic step-up therapy were musculoskeletal involvement (40%), eye involvement (25%), neurological involvement (15%) and intestinal involvement (10%). Most patients initiated a biologic treatment within the first year of follow-up. TNF-inhibitor (TNFi) were more frequently prescribed (95%) and one patient was treated with 8 therapeutic cycles of Rituximab (500 mg/weekly for 4 infusions to be repeated after at least 6 months) because of recurrent pancytopenia. All patients experienced non-biologic therapy before starting a TNFi. The preferred first-line TNFi was infliximab (50%), followed by adalimumab (40%) and etanercept (5%). As second line treatment were also prescribed certolizumab (10%) and golimumab (5%). 10 patients switched to a second line treatment because of inefficacy of the first biologic agent, mainly because of refractory arthritis, intestinal and mucocutaneous involvement. One patient switched from infliximab to certolizumab during pregnancy with subsequent worsening of arthritis.
85% of patients treated with biologic agents reached a clinical remission by the time of the latest follow up visit without any safety or tolerability issues.
Conclusion: A relevant proportion of patients in our BD cohort were treated with biologic therapy, because of severe or refractory manifestations. The most frequent indications were musculoskeletal, neurological or intestinal involvement. Biologic agents were a generally effective and safe therapeutic approach.
References:
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Table 1. General characteristics and disease involvement at diagnosis

	Biologic therapy		No biologic therapy		p value
	20 (21.05%)		75 (78.95%)		
General characteristics	**Media**	**SD**	**Media**	**SD**	
Age at disease onset (years ± SD)	34.5	± 10.49	38.64	± 13.18	p = 0.1976
Diagnostic delay (months ± SD)	45.28	± 67.48	28.09	± 48.42	p = 0.1996
Glucocorticoids at diagnosis (mg prednisone ± SD)	16.84	± 14.01	8.89	± 11.76	**p = 0.0115**
Glucocorticoids at latest follow up visit (mg prednisone ± SD)	6.38	± 7.76	3.83	± 4.81	p = 0.0707
	N	**%**	**N**	**%**	
F / M	12 / 8	60 / 40	54 / 41	72 / 28	p = 0.3030
Disease involvement at diagnosis					
Oral ulcers	20	100	75	100	
Genital ulcers	11	55	37	49,33	p = 0.6540
Cutaneous lesions	15	75	50	66,67	p = 0.4787
Eye involvement	6	30	27	36	p = 0.6184
Musculoskeletal involvement	16	80	35	46,67	**p = 0.0082**
Neurological involvement	6	30	8	10,67	**p = 0.0311**
Intestinal involvement	8	40	9	12	**p = 0.0039**
Thrombosis	2	10	18	24	p = 0.1747

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AB0463 ETIOLOGY OF PALPABLE PURPURA; A SINGLE CENTER EXPERIENCE

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Background: Purpura are lesions that occur after bleeding on the skin, mucous or serosal surfaces. Purpura can be classified into 2 subgroups; which are linked to thrombocytopenia and non-trombocytopenic purpura. While thrombocytopenic purpura often occurs due to a hematological disease; in non-trombocytopenic purpura etiological causes are very variable, and systemic vasculitis has an important place among them.
Objectives: The demographic features of the patients applying with purpura and the underlying causes and diseases were aimed to be revealed.
Methods: 44 consecutive patients (22 women, 22 men) who were admitted to the hospital in the last 6 months, due to purpura were evaluated. Average age of patients was 49.6 ± 19.6 years. Patients were questioned about a recent infection, drug use, concomitant or underlying diseases. The serological tests and other laboratory tests for etiology were performed and biopsy was taken from the skin lesions which are appropriate.
Results: While 4 (9%) patients had widespread purpura on the trunk-upper-lower extremities, in 22 (50%) patients purpuric lesions were limited only to the lower limb. The period between the onset of the first symptom and the admission to the hospital was longer than 4 weeks in 35 (79.5%) patients, and shorter than 1 week in 9 (20.5%) patients. 24 (54%) patients had an anamnesis of infection 2-3 weeks before purpura, and 20 (45%) patients had an anamnesis of drug use. The most common accompanying symptom was abdominal pain and was present in 15 (34%) patients. Biopsy was performed from the skin lesion in 37 patients. Histopathological examination of all was compatible with leukocytoclastic vasculitis. In indirect immunofluorescence staining, 17 were found to be IgA positive. 2 (4.5%) patients were diagnosed PR3-ANCA positive granulomatosis with polyangiitis. 1 patient had Hepatitis B virus infection was detected in 1 patient (2.2%), HIV infection was detected in 1 patient (2.2%) and malignancy was detected in 1 patient (2.2%).
Conclusion: In our study, the most common reason was found as IgA vasculitis in patients presenting with palpable purpura. Although vasculitic involvement was limited to the skin in most patients, organ-threatening systemic vasculitis was detected in a few patients. Patients applying with Purpura should be questioned for infection and drug use, should be examined for underlying diseases including systemic vasculitis, and closely monitored for organ involvement.
Disclosure of Interests: None declared
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AB0464 THE ROLE OF CYCLOPHOSPHAMIDE CHEMOTHERAPY IN THE TREATMENT OF EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS

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Background: Eosinophilic granulomatosis with polyangiitis (EGPA) is a rare type of ANCA associated Vasculitis (AAVs). Cyclophosphamide (CYC) is generally recommended for the induction of remission in life/organ threatening AAVs in combination with glucocorticoids. However, due to its rarity, randomized controlled trials regarding the efficacy of treatment modalities in EGPA are hard to perform. Therefore, the level of evidence for the use of CYC in the treatment of EGPA is lower when compared to other AAVs (1).
Objectives: The aim of this study is to investigate common therapeutic agents used for the treatment of patients with EGPA.
Methods: Medical records of patients who were followed-up with the diagnosis of EGPA between 2007-2020, in rheumatology clinics of Ankara and Adana Hospitals of Başkent University, were analyzed retrospectively. Treatment outcomes were assessed.
Results: Records of 11 patients (six females) were analyzed. The median age was 47 (19-77) years. The median follow-up time of the patients was 24 (9-156) months. Six patients were diagnosed with asthma. The median time between the diagnosis of asthma and EGPA was 4.5 (1-3) years. Five patients had tissue biopsies. Biopsy locations were terminal ileum, lung, myocardium and nerve. The most common forms of involvement were asthma, eosinophilic pneumonia and / or nodule, cardiovascular involvement, mononoritis multiplex, vasculitic skin rash, arthritis and bowel involvement, respectively. P-ANCA was positive in 8 patients. Three patients had myocarditis and cardiomyopathy, and two patients had isolated valve problems. The median BVAS value at the time of diagnosis and the third month of treatment was 17 (6-27) and 4 (2-7), respectively.
Nine patients used oral 1mg/ kg methylprednisolone (MP) and 500mg CYC every two weeks as an induction therapy. The cumulative median CYC dose was 4.5 g (1.5-8). Neither of the patients developed CYC related side effects. MP was tapered to 2 mg in five patients, and was quited in two patients. Azathioprine (AZA) was used in remission treatment following CYC therapy. Rituximab (RTX) therapy 1 g twice, 2 weeks apart was initiated in two patients due to unresponsiveness to CYC. While RTX was effective in one patient, newly developed renal involvement was detected after the third cycle of RTX therapy in other patient. Two patients had pregnancy plan therefore they used AZA plus MP as induction. A patient had mycophenolate mofetil plus MP due to AZA allergy. All patients are currently in remission except one patient.
Conclusion: In seven out of 11 EGPA patients, long-term remission was achieved with CYC treatment. CYC appears to be an effective and inexpensive method of first-line treatment for organ threatening EGPA.