glucocorticoids. Currently, only limited data on dynamics of peripheral leukocytes and leukocyte subset composition before, during and after treatment is available.

Objectives: To gain insight into the dynamics of leukocytes during the entire disease course and to determine whether patients in treatment-free remission resemble healthy controls.

Methods: Newly-diagnosed, treatment-naïve GCA (N=42) and PMR (N=31) patients were prospectively followed for up to 7 years. Absolute numbers of leukocyte subpopulation, both myeloid (neutrophils, monocytes) and lymphoid (CD4 and CD8 T-cells, B-cells and NK-cells) were measured at fixed time points. Laboratory inflammation markers (CRP, ESR, platelets and haemoglobin) were assessed as well. Values were compared with age-matched healthy controls (N=51) and infection controls (N=13). A stable treatment-free remission group for at least 6 months was defined.

Results: Neutrophil and monocyte numbers were higher in baseline GCA and PMR patients compared to healthy controls, whilst NK-cell numbers were reduced. In baseline GCA patients, CRP correlated positively with monocyte numbers. In baseline PMR patients, B-cell numbers were lowered and B-cell and NK-cell numbers correlated negatively with CRP. During glucocorticoid treatment, fluctuations in numbers of all leukocyte subsets were observed but neutrophil and monocyte numbers remained elevated and NK-cells numbers remained lowered during the entire treatment period (fig. 1). CRP and ESR were reduced by GC treatment and only moderately elevated during relapses. GCA patients in treatment-free remission still had elevated neutrophil and monocyte numbers. In addition, ESR and platelet numbers, were elevated whilst haemoglobin was lowered. PMR patients in treatment-free remission still had elevated monocytes while NK-cell and CD8+ T-cell numbers were decreased. Inflammatory markers and haemoglobin was normal.

Conclusion: In baseline GCA and PMR patients a shift towards the myeloid lineage is observed. This myeloid bias persisted in spite of GC treatment and also in treatment-free remission, suggesting a long-term effect of inflammation (an inflammatory imprint) on the peripheral blood compartment. Only GCA patients showed persistently elevated inflammatory markers; whether this reflects ongoing subclinical disease remains to be investigated.

Figure 1. Dynamics in leukocyte subset numbers during follow-up in GCA (A) and PMR (B) patients. Numbers at baseline (GCA n=42, PMR n=31), treatment phase I (GCA n=38, 69 samples; PMR n=25, 54 samples), phase II (GCA n=32, 43 samples; PMR n=23, 33 samples) and phase III (GCA n=29, 65 samples; PMR n=19, 56 samples) are expressed as median fold-change compared to healthy controls (n=51). #: sign difference between HC and baseline, *: sign difference between HC and treatment phase I, #: significant differences between HC and phase II and #: significant differences between HC and phase III (Mann-Whitney U test, P<0.05).

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SAT0229 INTESTINAL PERMEABILITY IN IGA-VASCULITIS IN ADULTS

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Background: Primary IgA-vasculitis and inflammatory bowel diseases (IBD) share many clinical signs, such as abdominal syndrome, skin purpura and IgA-nephropathy. It may indicate a common pathogenetic background for both pathological conditions. One of the hypotheses to explain a common IgA abnormalities and clinical similarities between two conditions is rupture of intestinal barrier resulting in defective antigen exclusion at mucosal level and increased antigen absorption into systemic

circulation. While increased intestinal permeability has been well established in IBD there are few reports addressing this issue in IgA-vasculitis. **Objectives:** Evaluation of small intestinal permeability in IgA-vasculitis. We have also looked for correlation between this parameter and clinical activity of the disease.

Methods: Intestinal permeability was assessed in 34 adult patients having primary IgA-vasculitis. EULAR/PRINTO/PRES criteria [1] were used for making of diagnosis after thorough laboratory and instrumental screening aiming to exclude a secondary nature of disease, including upper and lower endoscopy. Skin purpura was present in all patients being mandatory diagnostic criterion. Arthritis/arthralgia were seen in 20 patients (58%). There were no patients with evident abdominal damage just before and during investigation, although 15 (44%) patients had transient abdominal pain in the history. Mild to moderate signs of renal involvement (hematuria and/or proteinuria) were seen in 16 (47%) patients without renal impairment.

We have evaluated the gut permeability using ovalbumin (OVA) which has been measured in serum 3 hours after fasting oral intake of the two raw chicken eggs. The level of OVA in serum was measured using bivalent solid-phase immune-enzyme assay ("Sandwich-ELISA"). An increase of OVA in serum above 4 ng/ml was considered as the evidence of increased intestinal permeability.

Results: Totally, intestinal permeability to OVA was found to be increased in 20 of 34 patients (59%). In seventeen of twenty patients (85%) with increased intestinal permeability there were clinical signs of disease activity (skin purpura and/or arthritis and/or urinary abnormalities).

To find out which pattern of clinical activity is best associated with increased intestinal permeability we have separately analyzed the level of OVA in patients with skin purpura, arthritis and urinary abnormalities correspondingly during remission and exacerbation of the process.

Statistically significant difference (p<0,05) of OVA level was found only in patients selected on the basis of skin purpura activity: at the moment of clinically evident skin purpura the median OVA level was much higher (50 ng/ml) than during periods with no skin purpura (5 ng/ml).

The difference between the rate of increased intestinal permeability in patients with and without abdominal syndrome in the history was not statistically significant (p=0.87).

Conclusion: Results of our study may indicate a subclinical abdominal involvement in considerable part of patients with IgA-vasculitis, characterized with transient episodes of increased intestinal permeability. This phenomenon is chronologically related to disease activity, particularly to flare-up of the skin purpura. It could be speculated that this transient increase of intestinal permeability may result from mucosal inflammation which needs to be clarified in further investigations.

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SAT0230 GIANT CELL ARTERITIS WITH INTRACRANIAL VASCULITIS: A CASE SERIES

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Background: Giant cell arteritis (GCA) is a large vessel vasculitis that primarily affects the aorta and its branches. Extracranial branches of the carotid artery are frequently affected; however, intracranial involvement in GCA is rare [1].

Objectives: To determine the clinical features and outcome of patients with intracranial giant cell arteritis (IC-GCA).

Methods: A retrospective medical record review was performed to identify all patients with IC-GCA from 1/1996 through 5/2018. Demographic, clinical, laboratory, radiographic, and treatment data at baseline and subsequent follow-up visits were collected. Data was summarized using descriptive statistics.

Results: The study cohort included 9 patients with IC-GCA (78% male, mean [±SD] age 72.1 [±7.9] years). 8 of 9 patients had positive temporal artery biopsies. One patient with negative biopsy had evidence of large-vessel GCA on thoracic CT angiography.

The mean time from onset of GCA to intracranial involvement was 17 months (± 38). All patients had neurologic symptoms, 33% (n:3) had a stroke and 22% (n:2) had a transient ischemic attack. IC-GCA was diagnosed by cranial imaging in 8 patients and by autopsy in one. Cranial imaging modalities used included magnetic resonance angiography (n=8), CT angiography (n=2) and cerebral angiography (n=2). Intracranial vasculitis most commonly affected the internal carotid artery 78% (N=7) followed by the, vertebral artery 56% (n=5), posterior cerebral artery 56% (n=5), middle cerebral artery 44% (n=4), anterior cerebral artery 33% (n=3) and posterior inferior cerebral artery 11% (n=1). Stenosis was present in 89%, occlusion in 33%, dilatation in 11%, and wall thickening in 11%. All patients received glucocorticoids. For treatment of intracranial disease, additional agents included: cyclophosphamide (67%), Tocilizumab (22%). Despite treatment, outcomes for patients with IC-GCA were poor. Five of nine patients died during a mean length of follow-up of 2.4 months. **Conclusion:** Although rare, IC-GCA is associated with significant morbidity and mortality. It occurs predominantly in men, presenting with ischemic cerebrovascular events. Current treatment strategies appear to be of limited efficacy for IC-GCA.

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SAT0231 AGE AND THE CLINICAL FEATURES OF ADULT IGA VASCULITIS

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Background: IgA vasculitis (IgAV) is a common vasculitis of adult population, yet the disease in adults is still poorly defined.

Objectives: The aim of our study was to evaluate the potential influence of age on the presentation of adult IgAV.

Methods: We analyzed medical records of adult, histologically proven IgAV cases, diagnosed and followed at our secondary/tertiary rheumatology center between January 2010 and December 2018. We stratified IgAV cases into four groups, based on the patient age at presentation (18-39, 40-59, 60-79 and ≥ 80 years). The clinical features were compared between the groups.

Results: During the 108-month observation period we identified 265 new IgAV cases (60.0% males, median (IQR) age 64 (45–77) years). Skin, joint, gastrointestinal tract (GIT) and renal involvement developed in 265, 103 (38.9%), 80 (30.2%), and 118 (44.5%) cases, respectively. Younger adults (≤ 40 years) more frequently reported an infection prior to IgAV, and developed arthritis or GIT involvement. Severe renal involvement was less common under the age of 40 years. The incidence of skin limited IgAV and of necrotic purpura increased with patient age. Follow-up data were available for 189 patients. During a median (IQR) follow-up of 12 (6–24) months, 29 (15.3%) patients relapsed. Relapsing disease was more frequent in younger adults (≤ 40 years), compared to older ones.

Table 1. Age and clinical features of adult IgAV

Clinical characteristics	Age interval			
Age (years)	18-39	40-59	60-79	≥ 80
Number of cases	49	69	96	51
Male gender (%)	63.3	68.1	62.5	41.2
Prior infection (%)	57.1	36.2	28.1	21.6
Generalized purpura (%)	49.0	52.2	50.0	45.1
Skin necroses (%)	14.3	43.5	55.2	62.7
Skin limited IgAV	16.3	18.8	35.4	41.2
Joint involvement (%)	63.3	49.3	30.2	17.6
Arthritis (%)	28.6	14.5	14.6	2.0
GIT involvement (%)	42.9	34.8	21.9	27.5
Severe GIT involv. (%)	14.3	7.2	7.3	2.0
Renal involvement (%)	36.7	43.5	50.0	43.1
Severe renal involv. (%)	2.0	15.9	14.6	11.8
Elevated serum IgA	22.6	45.6	56.5	48.6
Relapses (% and number)	25.0 (9/36)	13.2 (7/53)	18.6 (13/70)	0 (0/30)

Legend: Generalized purpura: purpura extending above the waistline; GIT: gastrointestinal tract; severe GIT involvement: bloody diarrhea or ileus or bowel perforation; severe renal involvement: nephritic syndrome with acute renal failure or nephrotic syndrome;

Data on age related variations in the clinical presentation of IgAV are presented in Table 1.

Conclusion: We found subtle age related differences in the presentation and during follow up of adult IgAV.

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SAT0232 DISTRIBUTION OF MACROPHAGE SUBSETS IN TEMPORAL ARTERY BIOPSIES OF PATIENTS WITH GIANT CELL ARTERITIS

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Background: Giant cell arteritis (GCA) is a granulomatous vasculitis characterized by recruitment of T-cells and monocytes to the vessel wall. Upon recruitment, monocytes are activated and differentiate into macrophages. These lesional macrophages take on different phenotypes depending on cues from the microenvironment. These macrophages are known to produce pro-inflammatory cytokines, chemokines, matrix-metalloproteinases (MMPs), and growth factors contributing to amplification of the inflammatory response, vessel wall injury and remodeling. However, current knowledge regarding macrophage subsets in GCA lesions is limited.

Objectives: To examine the distribution of macrophage subsets in GCA lesions.

Methods: Immunohistochemistry was performed on consecutive sections of paraffin-embedded temporal arteries of biopsy-confirmed GCA patients (n = 11) with antibodies to selected markers of inflammation and tissue remodeling (Table 1). Positive cells were scored in three layers of the vessel wall (adventitia, media-intima, inner-intima) using a semi-quantitative scoring system. Expression by macrophages was confirmed by double staining with the macrophage transcription factor PU.1.

Table 1. Immunohistochemistry panel. AB019

Pro-inflammatory	Tissue remodeling
IL-6	CD206 (MR)
IL-1 β	IL-10
IL-12	FR- β
IL-23	MMP-12
CD64	MMP-9
	MMP-2

Results: The pro-inflammatory marker CD64 and pro-inflammatory cytokines (IL-12, IL-23 and IL-1 β) were strongly expressed by macrophages throughout the vessel wall, most prominently in the adventitia. However, subsets of these CD64+ macrophages also showed concomitant expression of the tissue remodelling markers CD206 and FR- β in specific compartments of the lesions. More specifically, CD206+ macrophages were mainly found along the media borders which co-localized with positive MMP-9 staining. Interestingly, FR- β positivity was also found in the inner-intima of TABs with a high degree of intimal hyperplasia. FR- β expression was significantly higher in the inner-intima region in TABs with

Figure 1. Spatial distribution of macrophage subsets in temporal artery of GCA affected vessel.