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.

massive intimal hyperplasia compared to TABs with mild intimal hyperplasia ($p = 0.01$).

Conclusion: Based on the expression of markers of inflammation and tissue remodeling, different subsets of infiltrating macrophages can be distinguished. Macrophages in vascular lesions of GCA patients display a distinct spatial distribution pattern within the inflamed vessel wall (Figure 1). Adventitial macrophages express highest level of pro-inflammatory cytokines indicating pro-inflammatory functions of these macrophages. Colocalization of CD206 and MMP-9 along media borders indicates a role for CD206+ macrophages in collagen digestion and angiogenesis. Additionally, the association of increased numbers of FR- β + macrophages with extensive intimal hyperplasia suggests that FR- β + macrophages promote myofibroblast proliferation.

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SAT0233 VALIDATION OF A NOVEL DISEASE CLASSIFICATION IN HACETTEPE TAKAYASU ARTERITIS COHORT

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Background: Takayasu Arteritis (TAK) is a rare idiopathic granulomatous large vessel vasculitis and it is a clinically heterogeneous disease. Various classifications and subsets which based on distribution of arterial lesions were defined in the literature in order to exhibit and predict different disease courses.

Objectives: We aimed to validate a new disease subsets developed by Goel et al in our TAK series (1).

Methods: We retrospectively evaluated the medical records of 164 TAK patients followed at Department of Rheumatology and Department of Pediatric Rheumatology in Hacettepe University, Ankara, Turkey between August 2005 and October 2018. The pediatric-onset (< 18 years of age at disease diagnosis) and adult-onset patients fulfilled the Ankara 2008 and the American College of Rheumatology (ACR) 1990 criteria for TAK, respectively. All patients were assigned to three clusters (Cluster 1: Abdominal Predominant, Cluster 2: Aortic Arch Predominant, Cluster 3: Focal Disease) using a decision tree defined in the original study (1). Demographics, clinical and laboratory features, treatment regimens were compared between in three groups.

Results: There were forty-two (25.6%), sixty-three (38.4%) and fifty-nine (36%) patients in Cluster 1, 2 and 3, respectively. Demographic and clinical data was summarized in Table-1. Cluster 1 included approximately half of patients (45.8%) with pediatric-onset TAK ($p = 0.048$). Baseline acute phase reactants (Erythrocyte Sedimentation Rate:ESR and C-reactive protein:CRP) and age at disease diagnosis were slightly lower in Cluster 1, however none of them did not reach statistical significance ($p = 0.94$, $p = 0.99$ and 0.56 , respectively). In contrast, cerebrovascular accident rates were slightly higher in Cluster 2 ($p = 0.24$). Although anti-TNF biological agents were more frequently used for treatment in Cluster 1 ($p = 0.004$), cyclophosphamide and/or overall biological agents usage was similar in three groups ($p = 0.15$).

Conclusion: Decision tree which defined by Goel et al. was applied to our single center pediatric and adult TAK cohort. In contrast to original validation cohorts, there was no difference among three clusters of our cohort except pediatric-onset disease and anti-TNF usage. Dissimilarities among the cohorts in terms of new classification system may be caused either number of patients or ethnic/regional differences.

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Table 1. Demographics and Clinical Features of TAK Disease Clusters

	Cluster 1 (n=42) (Abdominal Predominant)	Cluster 2 (Aortic Arch Predominant) (n=63)	Cluster 3 (Focal Disease) (n=59)	P
Female, n (%)	37 (88.1)	58 (90.1)	53 (89.8)	0.791
Age at TA diagnosis, years, Mean $\pm$ SD	29.3 $\pm$ 15.5	32.0 $\pm$ 13.7	32.6 $\pm$ 13.8	0.56
Pediatric-Onset (< 18 years of age at disease diagnosis)	11 (26.1)	7 (11.1)	6 (10.2)	0.048
Median disease duration, years (IQR)	4.5 (5.1)	5.2 (6.8)	4.9 (6.9)	0.83
Median ESR at TA diagnosis, mm/h (IQR)	48.5 (59)	53.0 (59.0)	62.0 (49.0)	0.94
Median CRP at TA diagnosis, mg/dL (IQR)	2.4 (10)	3.8 (9.0)	3.6 (8)	0.99
Hypertension (HT), n (%)	18 (42.9)	17 (27)	15 (25.4)	0.13
Cerebrovascular Accident (SVA), n (%)	3 (7.1)	11 (17.5)	6 (10.2)	0.24
Death, n (%)	3 (7.1)	2 (3.2)	2 (3.4)	0.56
Methotrexate, n (%)	22 (52.4)	38 (60.3)	34 (57.6)	0.72
Azathiopurine, n (%)	16 (38.1)	18 (28.6)	19 (32.2)	0.59
Leflunomide, n (%)	4 (9.5)	5 (7.9)	2 (3.4)	0.42
Cyclophosphamide, n (%)	19 (45.2)	27 (42.9)	18 (30.5)	0.24
Interleukin-6 blockade, n (%)	9 (21.4)	14 (22.2)	10 (16.9)	0.75
TNF- α inhibitors, n (%)	13 (31.0)	7 (11.1)	5 (8.5)	0.004
Cyclophosphamide and/or biologics, n (%)	27 (64.3)	37 (58.7)	27 (45.8)	0.15
Endovascular intervention*, n (%)	8 (19)	3 (4.8)	7 (11.9)	0.069
Vascular surgery*, n (%)	4 (9.5)	10 (15.9)	4 (6.8)	0.26

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SAT0234 RNA SEQUENCING IDENTIFIES AN IGA VASCULITIS ASSOCIATED SERUM MICRORNA SIGNATURE, DISCRIMINATING PATIENTS WITH IGA VASCULITIS FROM AGE- AND SEX-MATCHED HEALTHY SUBJECTS

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Background: Immunoglobulin A vasculitis (IgAV) is a small vessel vasculitis with IgA deposits in the vessel wall, manifesting with skin purpura, arthralgia/arthritis, gastrointestinal (GI) and renal complications. Altered serum miRNA signatures can reflect disease-specific pathology, which makes serum miRNAs promising disease biomarkers. We have recently reported that miRNA expression is changed in the affected skin of IgAV patients. The profile of serum miRNAs in IgAV, however, remains unknown.

Objectives: To determine the serum miRNA signatures in adult patients with IgAV as compared with healthy controls (HC) and explore *in silico* their gene targets and pathways.

Methods: Small RNA were isolated from sera of IgAV patients with high disease activity (sera collected at the time of diagnosis) and from the age- and sex-matched HC (n=6 each). Small RNA libraries were sequenced with Illumina technology and reads were mapped onto human genome (GRCh38). One IgAV sample was omitted from analysis due to too low number of recovered miRNA reads. Unsupervised clustering was based on the expression of 150 miRNAs with the highest variance across samples. The miRrort platform was used to predict gene targets of differentially expressed (DE) miRNAs with log2 fold change $\geq |1|$ and $p_{adj} < 0.01$. The STRING protein networks platform was used for pathway enrichment analysis of these genes.

Results: Unsupervised clustering of serum miRNA reads clearly distinguished between IgAV patients and HC (Figure 1). Among 1918 annotated miRNA (miRBase), 446 miRNAs were detected in serum samples and 88 miRNAs were identified as DE between IgAV patients and HC (log2 fold change $\geq |1|$, $p_{adj} < 0.05$). Specifically, 28 miRNAs were elevated in the serum of IgAV patients (e.g. miRNA-22, -146a, -185, -320a/