Results: TAB specimens from patients with biopsy-proven GCA, biopsy-negative GCA and controls were positive for anti-IL-6 staining in 59%, 13% and 48% of cases, respectively, the difference between biopsy-proven and biopsy-negative GCA patients being significant (p=0.04). In non-inflamed TABs, IL-6 was mainly expressed by mesenchymal cells in media and intima layers, while in inflamed TABs IL-6 was mainly expressed by mononuclear inflammatory infiltrating cells.
IL-6 grade 2–3 expression was observed in all 6 patients with visual loss compared to 25 (43.9%) of 57 patients without (p=0.011). Blindness was recorded in 2 patients with biopsy-proven GCA and 4 controls (all with a final diagnosis of non-arteritic ischaemic optic neuropathy). No associations were found between IL-6 expression and demographic characteristics, GCA signs/symptoms, laboratory and histopathological TAB findings. However, there was a statistical trend (p=0.055) of increased frequency of the halo sign at temporal artery CDS in patients with IL-6 expression grade 2–3 compared to those with IL-6 expression grade 0–1. No significant differences for the expression of IL-6 were observed between patients with and without PMR (5/8%–62.5% - versus 6/15%–40% -, p=0.400) and between patients with isolated PMR and those with TAB positive GCA (62.5% vs 59%, p=1.000).
Conclusions: Our study provides evidence that IL-6 expression does not increase the sensitivity of TAB in patients with morphologically uninflamed arteries. A search for further markers that may increase the sensitivity of TAB is warranted.
Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2018-eular.6729

THU0458 INVESTIGATION OF THE ROLE OF M-TOR PATHWAY IN KIDNEY NEEDLE BIOPSIES OF PATIENTS WITH ANTINEUTROPHIL CYTOPLASMIC AUTOANTIBODY-ASSOCIATED VASCULITIS

Z. Soypacaci[1], O. Cakmak[2], F. Cakalagaoglu[3], O. Gercik[4], I. Ertekin[5], A. Uzum[1], R. Ersoy[1], S. Akar[4]. *[1]Nephrology; [2]Internal Medicine, Izmir Katip Celebi University School of Medicine; [3]Pathology, Izmir Katip Celebi University School of Medicine Ataturk Research Hospital; [4]Rhemautology, Izmir Katip Celebi University School of Medicine; [5]Nephrology, Izmir Katip Celebi University School of Medicine Ataturk Research Hospital, Izmir, Turkey*

Background: Anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV) frequently affect the kidneys and renal involvement is an important factor regarding morbidity and mortality. Kidney lesion in AAV is characterised by necrotizing and crescentic glomerulonephritis by little or no immune deposition, and hence it was called pauci-immune glomerulonephritis (PIGN). The underlying mechanisms in the formation or progression of crescent formation need further investigations. Mammalian target of rapamycin (mTOR) is a serine/threonine kinase and plays role in the regulation of cell growth and proliferation.
Objectives: We aimed to evaluate the role of mTOR, which might be a potential therapeutic target, in kidney biopsies of patients with AAV.
Methods: The patients diagnosed as PIGN at an outpatient nephrology clinics of a tertiary hospital, between $^{\text{May 2009}}$ and June 2016, were retrospectively reviewed and those patients who had a renal biopsy before receiving an immunosuppressive treatment were included in the study. Renal biopsy specimens were immunohistochemically stained with antibodies of mTOR, phosphatase and tensin homolog (PTEN) and transforming growth factor- β (TGF-β) and scored by an experienced renal pathologist.
Results: In total 54 patients with AAV ([52%] female) were included in the study. Twenty-five (46%) patients were diagnosed as granulomatosis with polyangiitis, 6 (11%) patients as microscopic polyangiitis, 16 (30%) patients as renal-limited disease, one (2%) patient as eosinophilic granulomatosis with polyangiitis. Six (11%) patients with PIGN could not be classified definitely. According to the histopathologic examination; 22% of the biopsies were classified as focal, 33% crescentic, 22% mixed and 22% as sclerotic. The mTOR was expressed in substantial percentages of glomeruli of patients with PIGN. However we observed PTEN expression in all samples and mTOR in all tubulointerstitial areas. mTOR expression was found to be related with the presence of crescentic and sclerotic changes observed in glomeruli and the degree of fibrosis in interstitial areas. In our study serum creatinine level or response to treatment were not found to be associated with mTOR pathway expression
Conclusions: Our study showed that glomerular or interstitial expression of PI3K/Akt/mTOR pathway may play a role in the pathogenesis of PIGN and mTORC1 inhibitors might provide a viable alternative for this disease.
Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2018-eular.1920

THU0459 SURVIVAL OF BIOPSY PROVEN GIANT CELL ARTERITIS IN NORTHERN ITALY: CORRELATION WITH CLINICAL, LABORATORY AND HISTOPATHOLOGICAL FINDINGS

P. Macchioni[1], L. Boiardi[1], F. Muratore[1], M. Catanoso[1], A. Cavazza[2], P. Mancuso[3], L. Cimino[4], G. Restuccia[1], C. Salvarani[1]. *[1]Rheumatology Unit; [2]Pathology Unit; [3]Epidemiology Service; [4]Ophthalmology Unit, Azienda USL-IRCCS di Reggio Emilia, Reggio Emilia, Italy*

Objectives: To correlate survival with clinical, laboratory and histopathological findings in a population based cohort of patients with biopsy-proven giant cell arteritis (GCA) living in the Reggio Emilia area during a 26 years period.
Methods: In this population-based study, all patients living in the Reggio Emilia area who underwent temporal artery biopsy (TAB) for suspected GCA from January 1, 1986 to December 31, 2012 were identified. A pathologist with expertise in vasculitis and blinded to clinical data and final diagnosis reviewed all TABs. Based on the localization of the inflammation, positive TABs were classified into 4 categories: small vessel vasculitis (SVV), with inflammation limited to small periadventitial vessels devoid of muscular coat; vasa vasorum vasculitis (VVV), with inflammation surrounding the adventitial vasa vasorum; inflammation limited to adventitia (ILA), with inflammation spreading from vasa vasorum to the adventitia without extension to the media; transmural inflammation (TMI), with external elastic lamina disruption and extension of the inflammation to the media. Histopathologic features evaluated were: the severity of inflammation and intimal hyperplasia, both graded on a semiquantitative scale (mild=1, moderate=2 severe=3), the presence of intraluminal acute thrombosis, calcifications, giant cells, fibrinoid necrosis and laminar necrosis. Information about clinical manifestations, laboratory findings, treatment and disease course were collected. Patients were followed from GCA diagnosis to death, migration or December 2013.
Results: 281 patients (206 female, 73.3%) with biopsy-proven GCA were identified in the study period. 120 patients (84 female, 70%) died during a median follow-up period of 96 (IQR 55, 143) months. At univariate analysis, the presence of polymyalgia rheumatica (PMR) (HR 0.54, 95% CI 0.37–0.79, p=0.002), higher level of haemoglobin (HR 0.84, 95% CI 0.74–0.96, p=0.011) at disease onset, long-term remission (HR 0.47, 95% CI 0.26–0.86, p=0.015) and ILA or VVV at TAB (HR 0.48, 95% CI 0.24–0.97, p=0.041) were associated with lower mortality, while the evidence of large vessel involvement at imaging studies performed at diagnosis was associated with increased mortality (HR 5.84, 95% CI 1.57–21.8, p=0.009). Multivariate analysis confirmed the association between lower mortality and PMR (HR 0.54, 95% CI 0.36–0.81, p=0.003), higher level of haemoglobin (HR 0.83, 95% CI 0.69–0.99, p=0.049) at disease onset, and ILA or VVV at TAB (HR 0.38, 95% CI 0.17–0.82, p=0.014), and between increased mortality and large vessel involvement at imaging studies performed at diagnosis (HR 5.31, 95% CI 1.39–20.26, p=0.014).
Conclusions: PMR at diagnosis and only adventitial inflammation at TAB seem to identify subsets of biopsy-proven GCA patients with more benign disease, while large vessel involvement at diagnosis a subset with reduced survival.
Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2018-eular.6752

THU0460 FULLY INTEGRATED 18F-FDG PET/MR IN LARGE VESSEL VASCULITIS

R. Padoan[1], F. Crimi[2], M. Felicetti[1], F. Padovano[3], L. Punzi[1], C. Lacognata[2], R. Stramare[2], D. Cecchin[3], F. Bui[3], P. Zucchetta[3], F. Schiavon[1]. *[1]Operative Unit of Rheumatology, Department of Medicine DIMED; [2]Operative Unit of Radiology, Department of Medicine DIMED; [3]Operative Unit of Nuclear Medicine, Department of Medicine DIMED, University of Padova, Italy*

Background: Positron emission tomography (PET) is a non-invasive imaging method that detects ^{18}F-fluorodeoxyglucose (FDG) uptake in vessel's walls. Its simultaneous combination with magnetic resonance (MR) would offer not only a more detailed morphological analysis of the vessels but also a reduction of the radiation, simplifying the clinical workflow and being logistically easier for the patient.
Objectives: To evaluate, for the first time up to now, the usefulness of a fully integrated 18F-FDG PET/MR in a series of large vessels vasculitides (LVV) patients.
Methods: We performed a controlled non-randomised prospective study. Images were acquired on a fully integrated PET/MR scanner (Siemens Biograph mMR), consisting in a complete MR protocol and FDG-PET whole body imaging. We evaluated vessel's standard uptake value (SUV) maximum and wall thickness (WT), defined as the mean of 4 measures (at 12, 3, 6 and 9 o'clock) at the inferior margin of D5, D9, D12, L3 and at thickest point (max WT).

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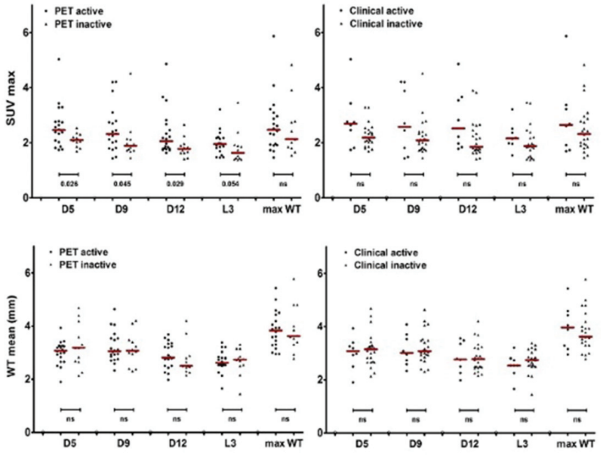
	All (n 23)	*GCA (n 13)*	*TAK (n 8)*	*p*
Female, n (%)	19 (82.6)	12 (92.3)	6 (75)	*ns*
Age at diagnosis	63 (51–68)	68 (63–73)	43.5 (30.5–57)	*0.003*
Diagnostic latency (months)	4.5 (2–12)	3 (2–10)	8 (3.5–12)	*ns*
ESR at disease onset (mm/h)	49 (38–68)	52.5 (45.5–59.7)	42 (40–61.5)	*ns*
CRP at disease onset (mg/L)	61.8 (13–132.5)	89 (32.5–106)	60.5 (9.3–132.5)	*ns*
Disease duration at PET/MR (months)	27 (18–36)	24 (13–29.5)	36.5 (14.75–129.3)	*ns*
ESR at examination (mm/h)	18 (9–35)	16 (7–31)	20.5 (11–44.5)	*ns*
CRP at examination (mg/L)	4.5 (2.55–8.9)	3.9 (3.48–4.72)	4.55 (2.05–10.4)	*ns*

Results: 23 LVV patients were included, 56.5% GCA, 34.8% TAK and 8.7% isolated aortitis, all Caucasian, mostly females (82%). We considered 55 PET scans, 32/55 in LVV group (from min. 1 to max. 3 scans/patient) mainly during follow-up (29/32 scans), and 23/55 in control group. Considering patients with abdominal aorta involvement, we found higher SUV max compared to controls, in all sites, regardless of disease activity. Mean WT resulted higher in patients than controls, but did not significantly differ between PET active or inactive patients (figure 1). Mean WT positively correlated with age in both cohorts, inversely correlated to disease duration in LVV patients, while no correlation with SUV max was observed. Despite clinical assessment was suggestive of remission in 24 (75%) cases before PET/MR acquisition, a normal uptake was present only in 12 (50%) of them. On the contrary, all patient with active disease at clinical examination (8, 25%) had also a positive PET/MR. Cohen's K coefficient between clinical assessment and imaging was poor (K Cohen=0, 33, 0.11–0.55). Finally, we found no significant correlation between SUV max and acute phase reactants.
Demographic and clinical data of LVV patients.



Conclusions: PET/MR is a safe imaging technique capable of detecting vasculitic inflammation, similar to PET/CT, but with a greater anatomical definition. The low radiological exposure represents a valid alternative to PET/CT for disease monitoring, especially in young women.

Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2018-eular.4773

THU0461 COMPARISON BETWEEN CLINICAL PROFILE AND OUTCOME OF PATIENTS WITH JUVENILE ONSET AND ADULT ONSET TAKAYASU ARTERITIS

R.A Goel[1], D. Danda[1], G. Joseph[2], A. Nair[1], R. Ravindran[1], V. Jeyaseelan[3].
[1]Clinical Immunology and Rheumatology; [2]Cardiology; [3]Biostatistics, Christian Medical College, Vellore, Tamil Nadu, Vellore, India

Background: There is a paucity of data comparing juvenile onset Takayasu arteritis (jTA) and adult onset TA (aTA).
Objectives: We aimed to compare differences in clinical profile and outcome of patients with jTA and aTA attending our centre during 1998–2017.
Methods: Details of demography, clinical presentation, laboratory results, angiography and treatment response were collected prospectively for 252 and retrospectively for the rest of patients with TA. Disease activity was defined by Indian Takayasu Activity Score- A (ITAS-A)(CRP).[1] Complete remission (CR) was defined as ITAS-A=0 with no angiographic progression. Patients with onset of disease at ≤16 years of age were classified as jTA while the rest as aTA.

Results: Among 602 patients with TA during this period, 119 (19.8%) were jTA, while 483 were aTA. Female predominance was less striking in jTA (71.4%) than aTA (79%), p=0.047. Patients with jTA had presented more commonly with fever (29% vs 17.4%, p=0.002), headache (31% vs 18%, p=0.002), pain abdomen (11% vs 5.6%, p=0.031), systolic hypertension (66.4% vs 48.4%, p<0.001), cardiomyopathy (15.1% vs 5.4%, p<0.001) and raised creatinine (16% vs 4.7%, p<0.001) while claudication as presenting symptom was less common in jTA (39%) as compared to aTA (55%), p=0.003. Pulse abnormality tended to be commoner in aTA. Angiographically, type-I disease (5.1% vs 22.6%, p<0.001) and coronary involvement (8.3% vs 20.6%, p=0.016) was less common while type-IV disease occurred more frequently (25% vs 14.3%, p=0.004) in jTA than in aTA. Logistic regression showed similar results after adjustment for gender. Median ITAS ^{2010} score was higher in jTA [7 (2–14)] than aTA [5 (2–11)].
Follow up was available for 77 and 287 patients with jTA and aTA respectively. Median follow up duration was $32^{10–61}$ months for jTA and $27^{10–59}$ months for aTA. **CR** was attained more frequently in jTA (n=67; 87%) than aTA (n=190; 66.2%), p=0.001. Another, 7 (9%) and 55 (19.2%) of patients with jTA and aTA respectively achieved partial response with immunosuppression. Among patients with initial CR, relapse of active disease during further follow up was observed more frequently in jTA [n=20, (29.9%)] as compared to aTA [n=50,(26.6%)], p=0.029. Altogether, persistently stable disease course was more common in jTA (62.3%) than aTA (47.5%), p=0.029.
Conclusions: In our large cohort of TA treated with uniform immunosuppression protocol, systemic features, hypertension, cardiomyopathy, renal dysfunction and type IV disease are more commonly observed in jTA while claudication, pulse abnormality, coronary involvement and type I disease are more frequent in aTA. Patients with jTA respond better to immunosuppression but relapse more frequently than aTA. Persistent stable disease course is commoner in jTA patients.

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Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2018-eular.5158

THU0462 LONG TERM FOLLOW-UP RESULTS OF TAKAYASU ARTERITIS COHORT: A TERTIARY-SINGLE CENTRE STUDY

S. Kaymaz Tahra, F. Alibaz-Oner, H. Direskeneli. *Department of Rheumatology, Marmara University, ıstanbul, Turkey*

Objectives: To assess the clinical characteristics and long term follow-up outcomes of patients with Takayasu's arteritis (TAK) in a tertiary referral centre.
Methods: In this retrospective study, 107 (F/M: 96/11) patients fullfilling ACR 1990 criteria for Takayasu Arteritis and referred to our centre between 2004 and 2017 were investigated. All clinical and demographic data during first diagnosis and longitudinal follow-up were abstracted from medical records. Relapse was defined according to the physician's global assessment (PGA).
Results: The median age was $30^{14–67}$ years at symptom onset and $33^{14–68}$ years at diagnosis. Median follow-up duration was 72 (6–264) months. According to Hata Angiographic Classification, Type 5 (51.8%) and Type 1 (38.8%) were the most common patterns with the most frequently affected vessel subclavian artery (82.2%). At diagnosis 0.5–1 mg/kg/day corticosteroid treatment was started in 94.6% patients and a steroid-sparing immunsuppressive (IS) agent in 96.3% of the patients. An initial pulse steroid (1 g/day) therapy was chosen for 8 patients. Before diagnosis 24% patients had a history of a revascularisation procedure. After IS treatments, 24% of the patients were undergone a new revascularisation procedure. During follow-up, biologic agents were chosen for 13.8% of the

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