Table. Proportion of TAB-positive GCA stratified by the main covariates.

Study and pt. characteristics	No. of studies (pt.)	Positive TAB (%) (95% CI)	I ² (%)
Mean age at diagnosis	<72.7 yrs 10 (961)	81.2 (71.1–88.4)	91
≥72.7 yrs	11 (1067)	74.8 (65.1–82.6)	91
Females	<71% of pt. 12 (1156)	78.3 (67.0–86.5)	94
≥71% of pt.	11 (1319)	81.6 (73.9–87.4)	88
Polymyalgia rheumatica	<42% of pt. 7 (629)	70.6 (59.4–79.9)	87
≥42% of pt.	7 (1024)	79.2 (66.2–88.0)	94
Ophthalmological signs	<21% of pt. 6 (893)	81.8 (66.2–91.2)	95
≥21% of pt.	7 (606)	72.2 (60.4–81.6)	87
Large-vessel involvement	<25% of pt. 2 (373)	63.0 (50.6–73.8)	78
≥25% of pt.	2 (203)	61.1 (54.2–67.5)	0
Study purpose	Diagnostic 3 (153)	88.8 (68.5–96.7)	75
Other	29 (2939)	76.0 (70.4–80.9)	90
Study type	Retrospective 24 (2688)	75.5 (69.3–80.8)	91
Prospective	8 (404)	83.1 (70.6–90.9)	86
Year of publication	<2012 16 (1806)	84.4 (77.1–89.6)	92
≥2012	16 (1286)	68.4 (63.4–72.9)	69

Pt : patients

Disclosure of Interests: Emma Rubenstein: None declared, Carla Maldini: None declared, Solange GONZALEZ-CHIAPPE: None declared, Sylvie Chevreton: None declared, Alfred Mahr Consultant for: Chugai Pharma France, Speakers bureau: Roche SAS Chugai Pharma France
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SAT0243 EFFECTS OF TOFACITINIB SUPPRESSED PULMONARY VASCULAR REMODELING OF ALLERGIC VASCULITIS IN A MURINE MODEL

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Background: We reported allergic granulomatous vasculitis with eosinophil infiltration in an asthma model of C57BL/6 sensitized with ovalbumin (OVA). TGF- β and IL-6 are thought to play an important role in fibroblasts proliferation and is critical to vascular remodeling in vasculitis. Tofacitinib inhibits vascular endothelial cells proliferation and canalization.

Objectives: To elucidate the role of tofacitinib in vascular remodeling of allergic granulomatous vasculitis, we examined the effects of tofacitinib on the vasculitis of the murine model.

Methods: C57BL/6 mice (6-8 weeks) were sensitized with ovalbumin (OVA) and alum. The positive controls (n=9) were exposed to aerosolized OVA daily for 7 days. The other group of mice (tofacitinib treated mice (n=9)) were administered with tofacitinib (100mg/kg intraperitoneal administration) in parallel with daily exposure to aerosolized OVA for 7 days. On 7th day, bronchoalveolar lavage (BALF) was performed and the lungs were excised for pathological analysis. Cytokines in BALF were measured.

Results: The total cell number and the number of Eosinophils in BALF on the 7th day were decreased significantly in the tofacitinib-treated mice compared with those of the control-positive mice. The blood eosinophil counts in the positive control increased after OVA inhalation. The blood eosinophil counts in the tofacitinib treated mice were lower on the than those in the positive control.

Figure 1. a. Positive control: Totally occluded pulmonary artery by intraluminal myofibroblasts in the OVA-sensitized mice with exposure to OVA in 7th day. (HE staining) b. Tofacitinib: Intraluminal myofibroblast accumulation was not observed in the OVA-sensitized mice with exposure to OVA and treated with tofacitinib in 7th day. (HE staining).

The concentrations of IL-4, IL-5, IL-6 and TGF- β in BAL fluids reduced significantly in the tofacitinib treated group. The pathological scores reduced significantly in the tofacitinib treated group compared to the

positive control group. Intra luminal infiltration and proliferation of Ki67 positive myofibroblasts, IL-6 positive cell and α -SMA positive cells in pulmonary arteries were reduced dramatically in the tofacitinib treated group compared to the positive control group.

Conclusion: Tofacitinib suppressed pulmonary vascular remodeling in a murine model of allergic vasculitis with eosinophil infiltration. Tofacitinib is a hopeful therapeutic drug for Eosinophilic granulomatosis with polyangiitis.

Disclosure of Interests: None declared

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SAT0244 EXPANDING SPECTRUM OF ADENOSINE DEAMINASE 2 (DADA2) MANIFESTATIONS: EXPERIENCE 13 PATIENTS FROM INDIA

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Background: Deficiency of adenosine deaminase 2 (DADA2) is a recently described hereditary autoinflammatory disease with limited published literature [1-5]. There are no previous case series published from Asia.

Objectives: To describe the clinical features and treatment responses of patients diagnosed with DADA2 from India.

Methods: Patients diagnosed with DADA2 at 4 different centres in India were included. Details of clinical features, laboratory investigations, ADA2 activity, genetic analysis of CECR1 gene, therapy received and outcomes were noted.

Results: A total of 13 patients (12 Indians and one Syrian) were diagnosed with DADA2 between April 2017 and January 2019. The median age was 22 years (range: 7-39 years) and 8 (61.5%) patients were males. The diagnosis was made at a median duration of 30 months (range: 1-360 months) from the onset of first symptom. Figure 1 shows the various clinical manifestations. One patient had unknown manifestation of bad obstetric history mimicking APLA syndrome. ESR (median: 49.5 mm/hr; range: 15-130 mm/hr) was raised in all except one patient who was in remission at the time of diagnosis. CRP value was available in 11 patients and was elevated in all (median: 51.4 mg/L; range: 17.6-140 mg/L). Immunoglobulin levels assessed in 4 patients were normal. Neuroimaging done in nine patients showed infarcts in 5, bleed in 3, multiple aneurysms and features of PRES in one patient each. Abdominal microaneurysms were noted in 5 patients. Arterial occlusion of peripheral limb vessels was noted in 2 patients. ADA2 activity was assessed in 9 patients and all had nearly undetectable activity. CECR1 gene mutation analysis was available in 9 patients: 8 patients were homozygous for p. (Gly47Arg) missense mutation at exon 2 of CECR1 gene while one patient displayed compound heterozygosity for two novel mutations, P. (Leu188Val) in exon 4 and C.753+2 T>A in intron 4 of CECR1 gene. Eight patients were started on anti-TNF drugs (adalimumab in 7 and etanercept in 1). One patient required higher doses of adalimumab (40mg followed by 120mg at week 1 and 80mg at week 2) for severe gastrointestinal involvement but later died due to disseminated herpes infection. Rest of the seven patients started on anti-TNF drugs achieved disease control. Our patients had CNS, eye and GI manifestations lower than those reported by Zhou et al¹ but higher than those reported by Nanthapal et al² and Navon Elkan et al⁴

Clinical features of 13 DADA2 patients

Figure 1. Clinical manifestations noted in our cohort of 13 patients with DADA2.

Conclusion: The spectrum of clinical presentations in Indian patients is different from those reported in previous cohorts. Anti TNF therapy was effective in majority.

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SAT0245

CHOROIDAL EVALUATION IN PATIENTS WITH CHILDHOOD POLYARTERITIS NODOSA (PAN) AND ADENOSINE DEAMINASE-2 DEFICIENCY (DADA-2)

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Background: Polyarteritis nodosa (PAN) is a systemic necrotizing vasculitis affecting small or medium arteries with a negative ANCA serology and no evidence of glomerulonephritis.

Objectives: The aim of this study was to evaluate the choroid with optical coherence tomography (OCT) in children with polyarteritis nodosa (PAN) and adenosine deaminase-2 deficiency (DADA-2).

Methods: The study included all PAN and DADA-2 patients (n=15), examined between June 2017 and September 2018, and an age and gender-matched control group (n=15). After ocular examination, choroidal images taken with sd-OCT (Heidelberg Spectralis) were evaluated with regard to choroidal thickness (ChT) at five points (750 and 1500 microns from the center of the fovea both in the temporal, nasal quadrant and under the fovea), total subfoveal choroidal area (TCA), luminal area (LA), stromal area (SA) and choroidal vascularity index (CVI).

Results: None of the patients had active ocular complaints or findings. The mean (±SD) age was 8.4 ± 3.69 years. ChT at 3 points, TCA, LA, and SA were found to be higher in patients with PAN and DADA-2. The CVI values were similar in both groups. No correlations were found between the OCT findings, activity score index (PVAS) and the biochemical parameters (Erythrocyte sedimentation rate, leukocyte, C-Reactive Protein).

Conclusion: The results of this study showed that the choroid was thicker in patients with PAN and DADA-2 than in the control group, suggesting that PAN and DADA-2 may affect the choroid. Ophthalmologic evaluation is important in PAN and DADA-2 patients, even in the absence of relevant complaints.

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SAT0246

USING BONE MINERAL DENSITY VERSUS THE RATIO OF BODY MASS INDEX TO BONE MINERAL DENSITY TO PREDICT FRACTURE RISK IN POLYMYALGIA RHEUMATICA

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Background: Polymyalgia Rheumatica (PMR) is the commonest inflammatory rheumatic condition that affects the elderly population and treatment with long-term corticosteroids is common¹. Whilst steroid treatment is beneficial in managing symptoms, it has many side effects including increasing the risk of osteoporosis and hence fractures². There has been recent research that suggests using the ratio of BMD to Body Mass Index (BMI) is a better marker of predicting fracture risk in obese patients than BMD alone³.

Objectives: Our research set out to find out whether BMD alone or the ratio of BMI to BMD is a better predictor of fracture risk in patients with PMR.

Methods: Data were used from a cohort of PMR patients referred for DEXA scan to a District General Hospital between June 2004 and October 2010. The following were recorded: age, sex, whether a fracture was sustained, whether they had had steroid therapy at any point, BMI, BMD at L1-L4, BMD at femoral neck (left and right) and BMD at hip (left and right). Logistic regression models were fitted using fracture as the dependent variable. The independent variables for the first set of logistic regression models were BMD at each level and for the second set BMI: BMD ratio at the same levels. Data were adjusted for sex and age at scan. Logistic models were compared using area under the ROC curves (AUC).

Results: 714 patients were used in the study, of whom 532 (75%) were female. Mean age was 70.5 (SD 8.84) with age range 45.8 to 96.5 years. 703 (98%) were recorded to have had steroid therapy at any point. Mean BMI was 28.2 kg/cm² (SD 5.24). 156 (22%) had sustained a fracture. Odds ratios and AUC values for each level were as shown in the table. The fit of the models using the BMD alone was superior to the fit of the models using the ratio as the AUC values were greater for BMD alone.

Table 1. – Odds ratios (age- and sex-adjusted) and AUC values

Level	Odds Ratio and CI (BMD)	AUC (BMD)	Odds Ratio and CI (BMI:BMD)	AUC (BMI: BMD)
L1	0.192 (0.0656, 0.560)	0.6789	1.04 (1.01, 1.07)	0.6621
L2	0.138 (0.0529, 0.358)	0.6977	1.06 (1.03, 1.09)	0.6777
L3	0.192 (0.0795, 0.463)	0.6881	1.06 (1.03, 1.09)	0.6730
L4	0.243 (0.108, 0.544)	0.6837	1.05 (1.02, 1.09)	0.6731
L1 to L4	0.150 (0.0560, 0.400)	0.6837	1.06 (1.03, 1.09)	0.6737
L FEMORAL NECK	0.103 (0.0219, 0.492)	0.6727	1.04 (1.01, 1.06)	0.6576
R FEMORAL NECK	0.0785 (0.0143, 0.430)	0.6837	1.05 (1.02, 1.08)	0.6750
L TOTAL	0.0980 (0.0233, 0.412)	0.6824	1.06 (1.03, 1.10)	0.6777
R TOTAL	0.0623 (0.0136, 0.285)	0.6917	1.07 (1.04, 1.11)	0.6821

Conclusion: This study identifies that the BMI:BMD ratio does not provide better indication of fracture risk than BMD alone in our cohort of patients with PMR. We have previously shown that the same is true for patients with rheumatoid arthritis. A limitation of this study is not stratifying by steroid use.

Further work will be done to study the role of the ratio in predicting fracture risk in patients with other conditions.

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