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
DOI: 10.1136/annrheumdis-2019-eular.4204

SAT0243 EFFECTS OF TOFACITINIB SUPPRESSED PULMONARY VASCULAR REMODELING OF ALLERGIC VASCULITIS IN A MURINE MODEL

Nobuhito Sasaki, Kohei Yamauchi. Iwate Medical University School of Medicine, Morioka, Japan, Division of Pulmonary medicine, Allergy and Rheumatology, Department of Internal Medicine, Morioka, Japan

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
DOI: 10.1136/annrheumdis-2019-eular.4151

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

Aman Sharma¹, Gsrnk Naidu¹, Ramesh Jois^{2,3}, Chengappa Kavachandana⁴, Rajkiran Dudam⁵, Sanjay Jain¹, Vir Negi⁴, Pui.Y Lee⁶. ¹Post Graduate Institute of Medical Education and Research, Chandigarh, Internal Medicine, Chandigarh, India; ²Vikram Hospital, Bengaluru, India; ³Vikram Hospital, Bengaluru, India; ⁴Jawaharlal Institute of Postgraduate Medical Education and Research, Puducherry, India; ⁵Hyderabad Rheumatology Center, Hyderabad, India; ⁶Boston Children's hospital, Boston, United States of America

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

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