Conclusion: A two compartments model including feedback mechanism from the circulating ANCA well described concentration-ANCA levels relationship. The potency of rituximab was higher in patients with PR3-ANCA than in patients with MPO-ANCA.

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SAT0237

MYOCARDIAL FDG-PET/CT FINDINGS IN CHILDREN WITH TAKAYASU ARTERITIS

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Background: Takayasu arteritis (TAK) is a chronic large-vessel vasculitis affecting the aorta and its major branches¹⁾. In Japan, ¹⁸F-fluorodeoxyglucose positron emission tomography with computed tomography (FDG-PET/CT) was approved since April 2018. Recently, usefulness of FDG-PET/CT to diagnose TAK or assess disease activity has been reported²⁾. However, the findings in children with TAK except for vascular lesions remain unknown.

Objectives: We aimed to retrospectively analyse the FDG-PET/CT findings in children with TAK except for vascular lesions.

Methods: This is a single-center retrospective observational study. We evaluated 9 children with TAK and compared them with 10 children with other rheumatic disease (systemic juvenile idiopathic arthritis; 5, polyarticular juvenile idiopathic arthritis; 1, juvenile dermatomyositis; 1, Crohn's disease; 2, Kikuchi's disease; 1) (control group). All children underwent FDG-PET/CT before the treatment. We compared the two groups for the age at performed FDG-PET/CT, sex, duration of onset to performed FDG-PET/CT, FDG-PET/CT findings. Binary data were analyzed by Fisher's exact test.

Results: Median age of the performed FDG-PET/CT was 13.9 (range; 11.1-14.9) years old in TAK children and 5.4 (1.4-12.4) years old in control group. Both groups include 6 females. Median duration of onset to performed FDG-PET/CT was 1 (range; 0-12) months in TAK children and 0 (0-12) months in control group. FDG accumulation in bone marrow was observed in 7 TAK patients (78%) and in 7 (70%) of control group. Moreover, FDG accumulation in spleen was observed in 7 TAK patients (78%) and in 10 (100%) of control group. These findings were common in both groups. On the other hand, FDG accumulation in left ventricular myocardium was observed in 8 TAK patients (89%) but in 3 (30%) of control group (p=0.02). Six TAK children received follow-up FDG-PET/CT. Myocardium FDG accumulation was continued in 3 children but resolved in 2. TAK children with persistent accumulation in myocardium had ascending aorta dilation later.

Conclusion: FDG accumulation in left ventricular myocardium would be a characteristic finding in children with TAK, and possibly related to later ascending aorta dilation.

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SAT0238

SHORTTERM PROGNOSIS OF A PROSPECTIVELY FOLLOWED COHORT OF UNSELECTED ADULT IGA VASCULITIS PATIENTS – A SINGLE RHEUMATOLOGY CENTRE EXPERIENCE

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Background: The acute phase of adult IgA vasculitis (IgAV) is well characterized. Data of even short-term prognosis of adult IgAV is scarce, mostly retrospective and usually limited to patients with significant renal disease.

Objectives: Our aim was to examine the short-term prognosis in a cohort of unselected adult IgAV cases.

Methods: We analysed medical records of newly diagnosed, histologically proven cases of IgAV at our department of rheumatology between 1 January 2010 and 31 May 2018, that were followed until 30 November 2018.

Results: During the 101-month observational period we identified 245 IgAV cases. The clinical characteristics at presentation and the initial treatment are shown in Table 1. During the acute phase of the disease four patients died (3 due to active vasculitis, 1 due to generalized CMV infection and heart failure). During the follow-up period 23 patients died, all deaths were unrelated to IgAV. 28 patients were lost to follow-up. The clinical characteristics of the remaining 190 patients who were followed for a median 12.7 (IQR 6.9–24.5) months at follow-up are shown in Table 2.

Table 1. The clinical characteristics at presentation and initial treatment of IgAV

% male	58.8		
Median age (IQR), years	64.7 (45.6–77.3)		
Purpura, # (%)	245 (100)		
necrotic, # (%)	114 (46.5)		
generalized, # (%)	121 (49.4)		
Joint involvement, # (%)	98 (40.0)		
GI tract involvement, # (%)	74 (30.2)		
severe GI, # (%)	18 (7.3)		
Renal involvement, # (%)	110 (44.9)		
severe, # (%)	29 (11.8)	Treatment, # (%)	190 (77.6)
median BVAS-3 (IQR)	8 (3–15)	local GC, # (%)	29 (11.8)
Newly diagnosed malignancy, # (%)	3 (1.2%)	systemic GC, # (%)	172 (70.2)
		cyclophosphamide, # (%)	24 (9.8)
		IVIg, # (%)	7 (2.9)

Legend: # - number of patients; IQR – interquartile range; GI - gastrointestinal tract; severe GI tract involvement - bloody diarrhoea or ileus or surgical intervention; severe renal involvement - acute renal failure or nephrotic syndrome; BVAS-3 - Birmingham vasculitis activity score; GC – glucocorticoids; IVIg – intravenous immunoglobulin; Malignancy – concurrently diagnosed malignancy.

Table 2. Clinical characteristics at last follow-up visit

	Pts. No (%)
Relapse	30 (15.8)
one; two; more than three	21 (11.1); 5 (2.6); 4 (2.1)
skin limited	22 (11.6)
skin+renal; skin+GI; renal	3 (1.6); 1 (0.5); 3 (1.6)
Urinary abnormalities	79 (41.6)
microhematuria	68 (35.8)
proteinuria - <1g/day; ≥1g/day	20 (10.5); 7 (3.7)
Renal function worsening	22 (11.6)

Conclusion: Contrary to the experience in children, IgAV in adults does not seem to be a benign disease even in the short-term. Admittedly, it is impossible to conclude whether this is due to the nature of the adult IgAV or because adults have more comorbidities than children.

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