relapsing-GCA were analyzed, these differences were maintained, except for the mean time from GCA diagnosis and the prevalence of ischemic optic neuropathy. Data on remissions were not available in this subgroup of GiACTA patients.

	GiACTA overall (n= 251)	GiACTA (only relapsing-GCA; n=132)	Clinical Practice (n= 22)	GiACTA (overall) vs Clinical Practice p	GiACTA (relapsing) vs Clinical Practice p
Women / men	188/63	99/33	17/5	0.99	0.97
Age, mean (SD)	69 (8.2)	69.1 (8)	69 (8)	1	0.
Inclusion criteria	ACR 1990 modified	ACR 1990 modified	ACR 1990	-	-
Newly diagnosed GCA/ recurrent GCA	119/132	0/132	0/22	**< 0.0001**	-
Time (months) from GCA diagnosis, mean (SD)	9.1 (16.8)	16.9 (20.3)	32,1 (39.6)	**0.01**	0.09
Signs/symptoms of GCA at TCZ onset#	98 (39)	59 (44.7)	9 (41.0)	0.96	0.92
PMR, n (%)	49 (19.5)	40 (30.3)	16 (72.7)	**< 0.0001**	**0.0003**
Unilateral blindness, n (%)	4 (1.6)	4 (3)	1 (4.5)	0.87	0.78
Bilateral blindness, n (%)	1 (0.4)	1 (0.8)	1 (4.5)	0.38	0.66
Ischemic optic neuropathy, n (%)	2 (0.8)	2 (1.5)	2 (9.1)	**0.003**	0.18
Amaurosis fugax, n (%)	2 (0.8)	1 (0.8)	0 (0)	0.38	0.31
Blurred vision, n (%)	14 (5.6)	10 (7.6)	0 (0)	0.53	0.68
ESR, mean (SD)	24 (19.4); n= 246	26.8 (19.6)	51.7 (35.4)	**0.002**	**0.004**
CRP, mean (SD)	7.5 (13.4); n= 250	8.4 (15.4)	4.1 (5.9)	**0.03**	**0.02**
Positive TAB, n (%)	156/172 (90.7)	82 (62.1)	16 (72.7)	**0.03**	0.47
Imaging techniques, n (%)	138 (55)	70 (53)	16 (72.7)	0.17	0.14
Positive MRA, n (%)	8 (3.2)	4 (3)	1 (4.5)	0.77	
Positive CT scan, n (%)	13 (5.2)	7 (5.3)	1 (4.5)	0.71	0.78
Positive PET/CT scan, n (%)	97 (38.7)	42 (31.8)	14 (63.6)	**0.004**	**0.008**
Patients on corticosteroids at study onset, n (%)	251 (100)	132 (100)	21 (95.4)	0.12	0.31
Dosage of prednisone at TCZ onset, mean (SD)	Recent ACG : 40 (13.1) Relapsing ACG: 30.2 (12)	30.2 (12)	26.2 (19.5)	-	0.36
Patients who had received traditional immunosuppressant agents, n (%)	27 (10.8)	23 (17)	19 (86.4)	**< 0.0001**	**< 0.0001**
Patients who had received biologic therapy, n (%)	-	-	2 (9.1)	-	-
TCZ route	SC	SC	IV	-	-
Sustained remission, n (%) §	82 (54.6)	-	6 (27.3)	**0.029**	-
Severe infection, n (%)§	9/150 (6)	-	3 (13.6)	0.39	-

includes localized headache, TA, or scalp tenderness, jaw claudication, new or worsened extremity claudication.
§ In RCT patients with active TCZ therapy were only considered; *p<0.05

Conclusions: Patients receiving TCZ in the clinical practice study have several baseline clinical and laboratory differences with regard to those included in the GiACTA trial and, therefore, data of this trial should be taken cautiously when applied in a real-world scenario.

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OP0136 MICRORNA-223-3P EXPRESSION IN AFFECTED SKIN OF ADULT IGA VASCULITIS CORRELATES WITH THE SEVERITY OF SKIN INVOLVEMENT

A. Hocevar[1], J. Pizem[2], M. Tomsic[1], D. Glavac[2]. *[1]Department of Rheumatology, University Medical Centre Ljubljana; [2]Institute of Pathology, Faculty of Medicine, University of Ljubljana, Ljubljana, Slovenia*

Background: IgA vasculitis (IgAV) represents a common systemic vasculitis in paediatric and adult population. Our current knowledge of disease pathogenesis is still very limited and there is no information on miRNAs profile in IgAV.
Objectives: The aim of our study was to determine the expression of three miRNAs (miR-148–3p, miR-155–5p and miR-223–3p) in the affected skin of adult IgAV patients.
Methods: The study included 65 skin samples from consecutive, untreated IgAV patients (61% male, median age 67.6 years, range 29–91), diagnosed between October 2014 and September 2016, and 10 control skin samples. Total RNA was isolated from tissue section of formalin-fixed, paraffin-embedded samples of biopsied IgAV vasculitic skin lesions and normal skin samples. Expression of miR-148–3p, miR-155–5p and miR-223–3p was measured using qRT-PCR. Skin miRNAs expression was then correlated to clinical characteristics of adult IgAV patients. To present relative miRNA expression the ΔΔCT method was used.
Results: We found significantly higher expression levels of miR-223–3p in the affected skin compared to controls (14-fold; $p<0.001$). The expression of the 148b-3p and miR-155–5p was near normal levels (1.05-fold and 1.13-fold increase, respectively). The differences in the expression of miR-223–3p depending on clinical parameters of IgAV are presented in Table 1. Patients with necrotic skin lesions had significantly higher miR-223 tissue expression than those with non-necrotic purpura (p=0.020). Gastrointestinal tract (GIT) involvement inversely correlated with the level of skin miR-223 expression (p=0.024). No significant relationship between renal involvement and skin miR-223 was found.
Conclusions: miR-223 expression was increased in the affected skin of IgAV in comparison to normal skin. Levels of miR-223 expression correlated with severity of skin involvement and inversely with GIT involvement.

Table 1. miR-223–3p expression in IgAV

Characteristics		Number of cases	ΔΔCT miR223–3p median	IQR1	IQR2	P value
General symptoms	YES	10	3.11	1.86	5.55	0.683
	NO	55	3.72	2.53	5.38	
Arthritis	YES	6	3.04	2.67	4.60	0.482
	NO	59	3.72	2.48	4.60	
Generalized purpura	YES	37	3.72	2.46	5.87	0.615
	NO	28	3.63	2.56	5.10	
Skin necroses	YES	32	4.68	2.94	5.84	0.020
	NO	33	3.19	1.97	4.76	
GIT involvement	YES	16	2.78	1.69	3.90	0.024
	NO	49	4.29	2.69	5.62	
Severe GIT involvement	YES	5	2.67	1.88	3.07	0.078
	NO	60	4.22	2.56	5.57	
Renal involvement	YES	28	4.29	2.69	5.72	0.260
	NO	37	3.20	1.82	5.41	
Severe renal involvement	YES	9	4.50	2.44	5.13	0.955
	NO	56	3.60	2.49	5.50	
Elevated serum IgA level	YES	30	4.63	2.71	5.93	0.041
	NO	35	3.19	1.75	4.80	

Legend: generalized purpura - purpura above the waist; GIT - gastrointestinal tract; severe GIT involvement - bloody diarrhoea or ileus or surgical intervention; severe renal involvement - acute kidney injury or nephrotic syndrome.

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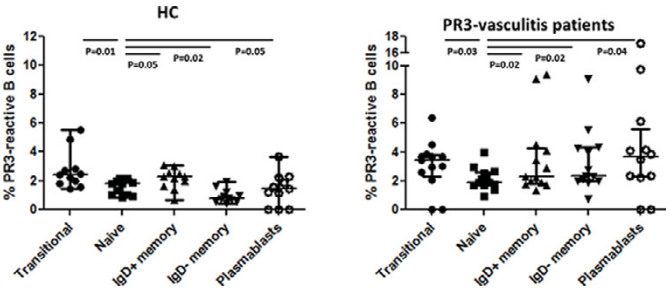
OP0137 AUTO-REACTIVE B CELLS ESCAPE PERIPHERAL TOLERANCE CHECKPOINTS IN PATIENTS WITH PR3-ANCA ASSOCIATED VASCULITIS

D. Cornec[1,2], A. Hummel[1], T. Peikert[1], J.-O. Pers[2], U. Specks[1]. *[1]Mayo Clinic, Rochester MN, United States; [2]CHRU and University, Brest, France*

Background: While extensive studies have been performed to characterize ANCA, little is known about the auto-reactive B cells that produce these autoantibodies. Indirect evidence previously suggested the presence of circulating PR3-specific B cells in patients with PR3-ANCA-associated vasculitis (AAV).
Objectives: To develop a method to detect circulating PR3-specific B cells in patients with PR3-AAV, to study their proportion among the different B-cell subsets and to assess their relationship with disease activity.
Methods: An enzymatically inactive, conformationally mature, recombinant PR3 (rPR3) was tagged using FITC or biotin. To study the ability of this rPR3 to bind specifically to cells expressing PR3-specific immunoglobulins on their surface, we used two hybridoma cell lines, MCPR3–2 (producing an anti-human PR3 monoclonal antibody) and MCPR3–13 (producing an anti-mouse PR3 monoclonal antibody, with no cross-reactivity with human PR3). We measured the proportion of PR3-FITC positive B cells among PBMCs in 13 patients with PR3-AAV and 14 healthy controls (HCs) by flow cytometry. We then developed a multi-color flow cytometry including CD19, IgD, CD27, CD38, CD24 and biotinylated rPR3 to measure the proportion of PR3-specific B cells among different B-cell subsets in an independent group of 13 patients with PR3-AAV and 11 HCs.
Results: rPR3 efficiently bound MCPR3–2 hybridoma cells but not MCPR3–13. Specificity of the staining was confirmed by competition experiments: pre-incubation of MCPR3–2 cells with untagged human rPR3 totally abrogated rPR3-FITC staining, whereas pre-incubation with mouse rPR3 had no effect. Dose-ranging experiments defined the optimal concentration of rPR3 to stain cells expressing anti-PR3 immunoglobulin. The mean (SEM) proportion of rPR3-FITC-stained B cells was higher in patients with PR3-AAV compared to HCs: 2.10% (2.33) vs 0.45% (0.19) respectively, $p<0.001$. Patients with active disease had numerically higher proportions of PR3-specific B cells than patients in remission: 3.66% (3.28) vs 1.10% (0.52), p=0.09. In HCs, the proportion of PR3-specific B cells was highest among the transitional B-cell subset, and decreased along with the maturation of B cells (figure). Conversely, in patients, the proportion of PR3-specific B cells progressively increased with the maturation of B cells (median 1.9% of naïve B cells, 2.30% of IgD+ memory B cells, 2.37% of IgD- memory B cells, and 3.68% of plasmablasts, $p<0.05$ for all comparisons with the naïve subset).



Conclusions: This study describes an original method to detect and study