DECREASED ACTIVITY OF ADAMTS13 IN PATIENTS WITH ANCA-ASSOCIATED VASCULITIS

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INTRODUCTION AND AIMS

Deficiency of ADAMTS13 plasma activity, the von-Willebrand factor processing metalloprotease may promote leukocyte and platelet adhesion to endothelial cells, thereby increasing endothelial injury. In ANCA associated vasculitis (AAV), ANCA IgG activates granulocytes, which, by releasing lytic granule constituents, damage vascular endothelium. Based on these comparable pathophysiological pathways, we aimed to study whether ADAMTS13 activity has any alterations in patients with AAV.

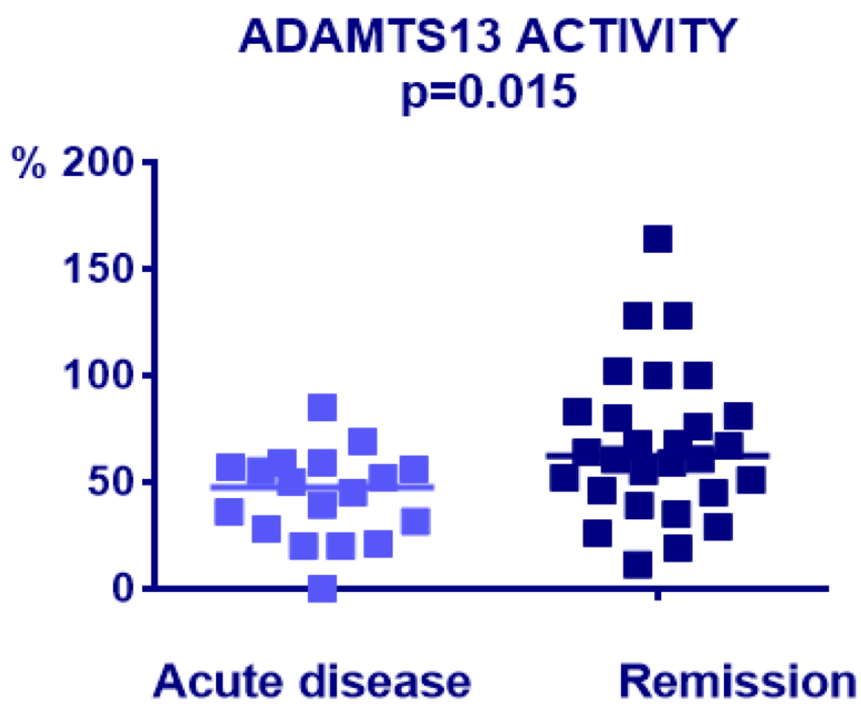
METHODS

ADAMTS13 activity (measured by the fluorescence resonance energy transfer assay) was determined in 18 subjects with active AAV, and 28 measurements were performed in patients with remission. Eleven patients from the cohort had sequential plasma samples of both active and remission stages.

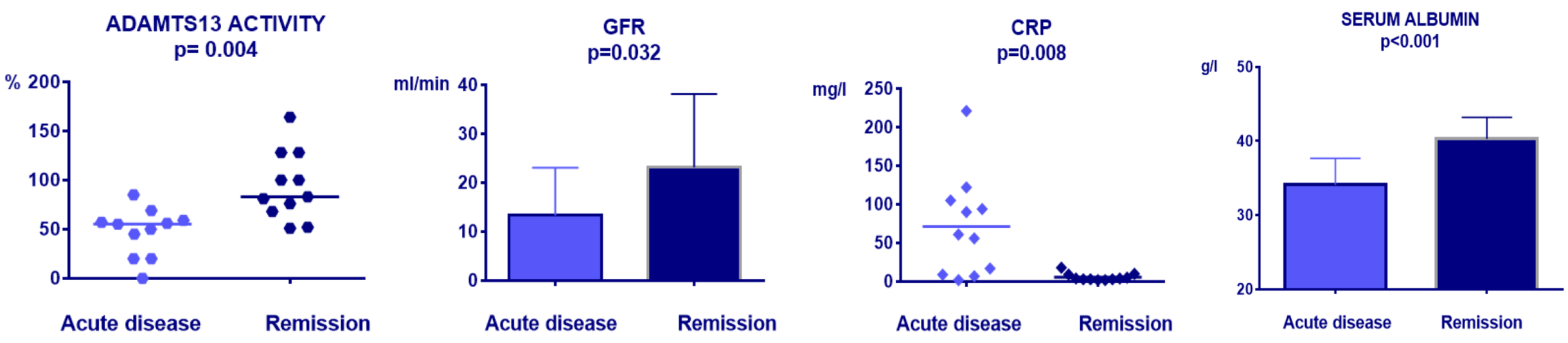
	Patients with active AAV	Patients in remission	Level of significance
n	18	28	
Male/female	4/14	10/18	p=0.513
Age (year)	64±12	59±11	p=0.114
c-/p-ANCA (n)	6/12		
a-PR3/a-MPO (U/l)	100 (28) / 49 (74)		
BVAS	20.4±5	0	**p<0.001**
Creatinine (umol/l)	358 (323)	210 (441)	p=0.079
GFR (ml/min)	11.5 (12)	27 (36)	p=0.068
Albumin (g/l)	32.4±4	39.9±3	**p<0.001**
CRP (mg/l)	53 (86)	4 (7)	**p<0.001**
Hgb (g%)	9.0±1.4	12.2±1.4	**p<0.001**
Uprotein (mg/day)	1514 (2645)	242 (787)	**p=0.003**
Dialysis (Yes/No)	4/14	8/20	**P=0.739**
LDH (U/l)	425±87	482±143	p=0.151
Platelet count (G/l)	334±119	248±62	**p=0.010**

Data are given as AVE±SD or median with IQR

ADAMTS13 ACTIVITY IN PATIENTS WITH ACTIVE AAV AND IN REMISSION



RESULTS IN 11 PATIENTS WITH SEQUENTIAL SAMPLES FROM ACTIVE AND REMISSION STAGES OF AAV



CONCLUSIONS

In patients with AAV but without evidence for microangiopathic hemolytic anemia, the activity of ADAMTS13 is decreased. Our results suggest that ADAMTS13 may serve as a systemic biomarker in AAV. Its decreased activity may develop as the consequence of endothelial dysfunction due to AAV, but it is also possible that ADAMTS13 deficiency itself contributes to the endothelial damage, which deserves further investigations.

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