

ALTERAZIONI DELLE PIASTRINE E CONDIZIONI GENETICHE

## THE BURDEN OF CEREBRAL SMALL VESSEL DISEASE IN ITTP PATIENTS FROM ACUTE PHASE TO REMISSION.

A. Truma<sup>1,2</sup>, F. M. Lo Russo<sup>1</sup>, G. Conte<sup>1,2</sup>, I. Mancini<sup>1</sup>, M. Abbattista<sup>1</sup>, J. Giannotta<sup>1</sup>, A. Artoni<sup>1</sup>, B. Ferrari<sup>1</sup>, P. Agosti<sup>1,2</sup>, F. M. Triulzi<sup>1,2</sup>, F. Peyvandi<sup>1,2</sup>.

1. Fondazione IRCCS Ca' Granda Ospedale Maggiore Policlinico, Angelo Bianchi Bonomi Hemophilia and Thrombosis Center, Milan; <sup>2</sup> Università degli Studi di Milano, Department of Pathophysiology and Transplantation, Milan.

**Background:** Immune-mediated thrombotic thrombocytopenic purpura (iTTP) is a life-threatening microangiopathy often characterized by acute neurological involvement during the acute phase followed by neurological complications during remission, due to microvascular damage. Cerebral small vessel disease (cSVD) is a condition affecting the brain small vessels leading to structural damage as assessed by magnetic resonance imaging (MRI). Age and hypertension are key risk factors for cSVD, which is often associated with stroke and dementia. To date, the prevalence of cSVD in iTTP patients remains unexplored.

**Aims:** To assess the prevalence and progression of cSVD in iTTP patients.

**Methods:** We performed a prospective study including consecutive patients admitted to our hospital for acute iTTP. Patients underwent a brain MRI during the acute phase and after one year of clinical remission. The presence of cSVD

was defined as a cSVD score  $\geq 1$  up to 4, calculated according to the sum of individual imaging markers (i.e., lacunes, cerebral microbleeds, white matter hyperintensities, and enlarged perivascular spaces).

**Results:** Twenty patients were enrolled with a median age of 52 years (Table 1). MRI was performed a median of 8 days after admission (IQR 5 - 8). During the acute phase, cSVD had a prevalence of 55% with a median score of 1 (IQR 0 - 1).

Hypertension was present in 6 patients (30%), median score 1.5 (IQR 1-3). An acute ischemic stroke was documented in 8 patients (40%), median score 1.5 (IQR 1-3). At 1-year follow-up, 12 patients underwent MRI: 9 remained stable, while 3 showed an increase of at least one point in the cSVD score.

**Conclusion:** cSVD is common in iTTP patients and may worsen over time during disease remission. Early detection and management of modifiable risk factors might be important to mitigate long-term complications.

**Email:** [ada.truma@gmail.com](mailto:ada.truma@gmail.com)

**Main characteristics of iTTP patients**

Variables	Acute (n=20)	After 1 year follow-up (n=12)
Female, n (%)	13 (65)	7 (58)
Age at episode (years), median (IQR)	52 (44- 63)	51 (42- 59)
First episode, n (%)	17 (85)	8 (80)
Neurological signs/symptoms at presentation, n (%)	16 (80)	11 (92)
Hypertension, n (%)	6 (30)	2 (22)
Diabetes, n (%)	0 (0)	0 (0)
BMI, kg/m <sup>2</sup> , n (IQR)	27 (25 - 29)	26 (23- 29)
Smoking, n (%)	11 (55)	4 (44)
Hyperlipidemia, n (%)	5 (25)	3 (33)
Positive DWI, n (%)	8 (40)	0 (0)
Total cSVD <sup>1</sup> score, n (%)		
0	9 (45)	5 (42)
1	7 (35)	5 (42)
2	2 (10)	2 (17)
3	2 (10)	0 (0)
4	0 (0)	0 (0)
cSVD burden, n (%)		
score > = 1	11 (55)	7 (58)
Moderate to high disease (score 2-4)	4 (20)	2 (17)

MRI magnetic resonance imaging, cSVD cerebral small vessel disease, DWI diffusion weighted imaging

<sup>1</sup>The total cSVD burden score is the sum of neuroimaging markers: presence of white matter hyperintensities on MRI (deep tissue Fazekas score 2 or 3 and/or periventricular Fazekas score 3) = 1 point; presence of one or more lacunes = 1 point; presence of one or more cerebral microbleeds detected by SWI (Susceptibility Weighted Imaging) = 1 point; presence of enlarged perivascular spaces in basal ganglia (≥10 enlarged spaces) = 1 point