Recurrent ischemic stroke in patients with thrombotic thrombocytopenic purpura (TTP): description of two cases.

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Background

TTP is a rare and potentially fatal disease associated with ADAMTS13 deficiency. It affects mainly adults and it's characterized by thrombocytopenia, microangiopathic haemolytic anaemia, fever, neurological and renal abnormalities. Before the availability of effective therapies such as plasma exchange (PEX), most of the patients died due to systemic microvascular thrombosis resulting in cerebral and myocardial infarctions or renal failure[1].

Patient 1

C.B., female, 50-year-old

- 30-year-old: diagnosis of TTP after the delivery of her first child ➡ *ischemic stroke in right frontal and occipital lobe* (treated with PEX)
- 36-year-old: second pregnancy ➡ *ischemic stroke in left internal capsule and thalamus* (treated with PEX)
- 48-year-old: ➡ *ischemic stroke in right thalamus*
- 50-year-old: ➡ *ischemic stroke in left MCA area*

Investigations to rule out other causes of stroke: negative

Measurement of ADAMTS13 activity: severe deficiency

The patient developed a **multi-infarct dementia** and a significant disability due to global aphasia and right hemiplegia.

Patient 2

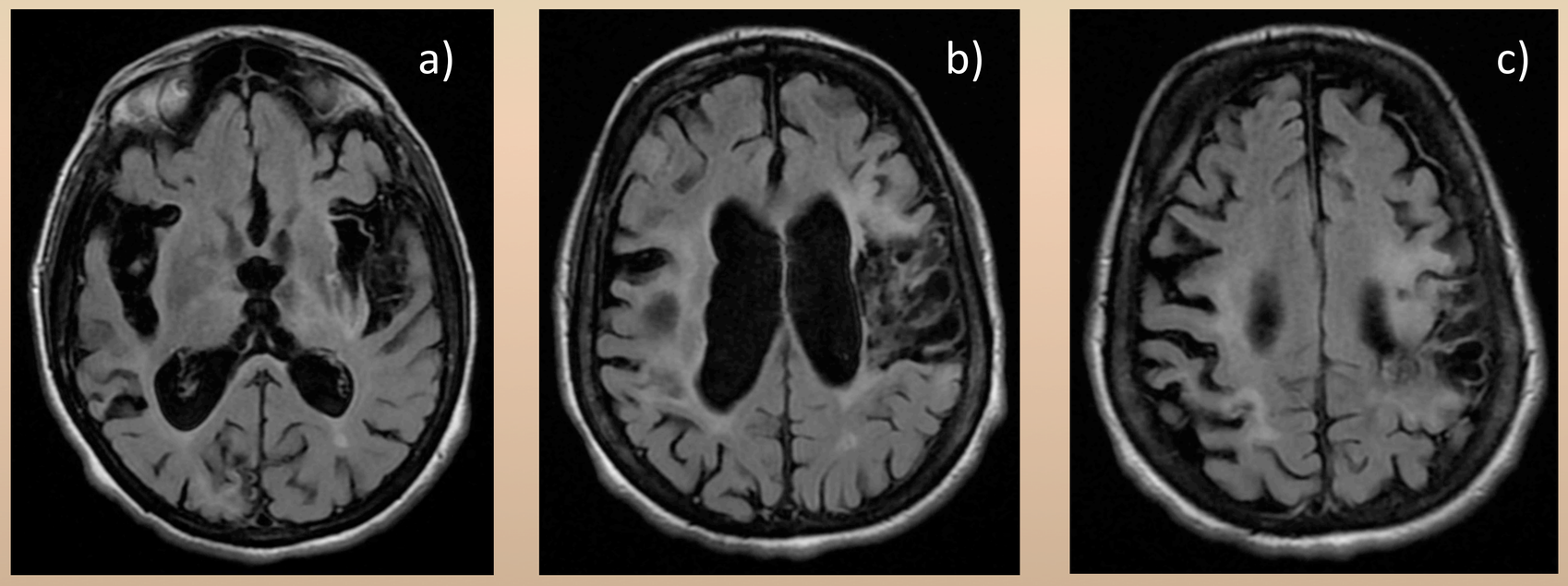
A.B., male, 62-year-old

- 22-year-old: diagnosis of TTP made after repeated episodes of hematemesis and thrombocytopenia
- 54-year-old: *ischemic stroke in the right occipital lobe*
- 55-year-old: *ischemic stroke in a peri-rolandic area* (treated with PEX and corticosteroids and then rituximab)

Vascular risk factors and comorbidities: hypertension, smoking, HCV infection

Measurement of ADAMTS13 activity: not available

No relapses of TTP



Patient 1
Figure a-b-c: Brain MRI, T2 FLAIR sequences

Discussion

TTP has been described as a possible cause of ischemic stroke mainly the microvascular type. The recommended treatment is PEX. The use of thrombolytic therapy is controversial and poorly described[2]. Early diagnosis of TTP is crucial for clinical outcome because a prompt treatment significantly reduces morbidity and mortality. Measurement of ADAMTS13 is not necessary for diagnosis, although a serious deficiency correlates with an increased risk of relapse[2].

References:

[1] George JN. Clinical practice. Thrombotic thrombocytopenic purpura. The New England Journal of Medicine. 2006 May 4; 354 (18): 1927-35.

[2] Boattini M., Procaccianti G. Stroke due to typical thrombotic thrombocytopenic purpura treated successfully with intravenous thrombolysis and therapeutic plasma exchange. BMJ Case Reports. 2013 Jan 28; 2013

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