An uncommon presentation of antiphospholipid syndrome

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Definitions

Moyamoya disease = cerebrovascular disease characterized by intracranial large vessels occlusion with conspicuous collateral circulation.

Moyamoya syndrome (MMS) = a moymoya like condition associated with predisposing causes such as collagenopathies, autoimmune and hematologic diseases.

Laboratory studies.

a.P.T.T. Ratio	1.33 *		(0.80 - 1.25)
LAC Ricerca Lupus Anticoagulant	Conclusioni:		Positiva.
aCL Ab.anti-cardiolipina IgG	816.0	GPL-U/ml	< 10 negativo 10 - 40 basso positivo > 40 positivo
B2GPI Ab.anti-beta2-glicop.I IgG	68.0	U/ml	< 7 negativo 7 - 10 dubbio > 10 positivo

We diagnosed antiphospholipid syndrome (APS)

Imaging.

- ✓ Brain MRI scan: right thalamic lacunar stroke **(figure 1)**.
- ✓ CT angiography (CTA)**:** left middle cerebral artery (MCA) occlusion (**figure 2A**)
- ✓ CT perfusion (CTP): left hemisphere hypoperfusion in MTT map (**figure 2B**).
- ✓ Digital subtraction angiography (DSA)**:** left MCA occlusion and prominent arterial collateralization from anterior and posterior circulation and from meningeal vessels (**figure 3**)

Moyamoya syndrome was diagnosed.

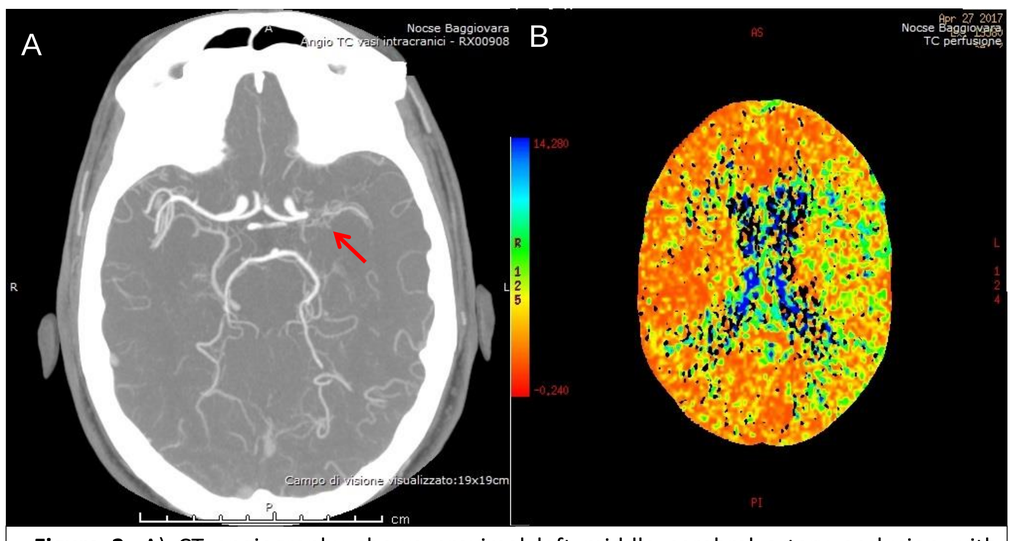

Figure 2. A) CT angiography shows proximal left middle cerebral artery occlusion with abnormal collateralization (arrow). B) CT perfusion shows left middle cerebral artery territory hypoperfusion in MTT map (increased mean transit time).

Treatment.

- MMS in adults is associated with hemorrhagic risk.
- APS syndrome needs to be treated with antithrombotic therapy.

No straightforward indications exist in these cases, we only found case reports[1–3] mentioning use of antiplatelet and anticoagulant agents.

Considering that our patient already had an ischemic stroke and that he had no signs of previous bleeding on MRI scan, **we decided to start him on anticoagulants and offer him neuroradiologic and rheumatologic follow up care.**

Clinical features.

A 51 years old man presenting with:

- ✓ left hemibody sensory loss, abruptly appeared around 1.5 years before and subsequent dystonic posture of the left hand.
- ✓ unspecified psychotic disorder consisting in a hypochondriac delirium.

Neuropsychological testing showed decreased verbal fluency and executive function deficit.

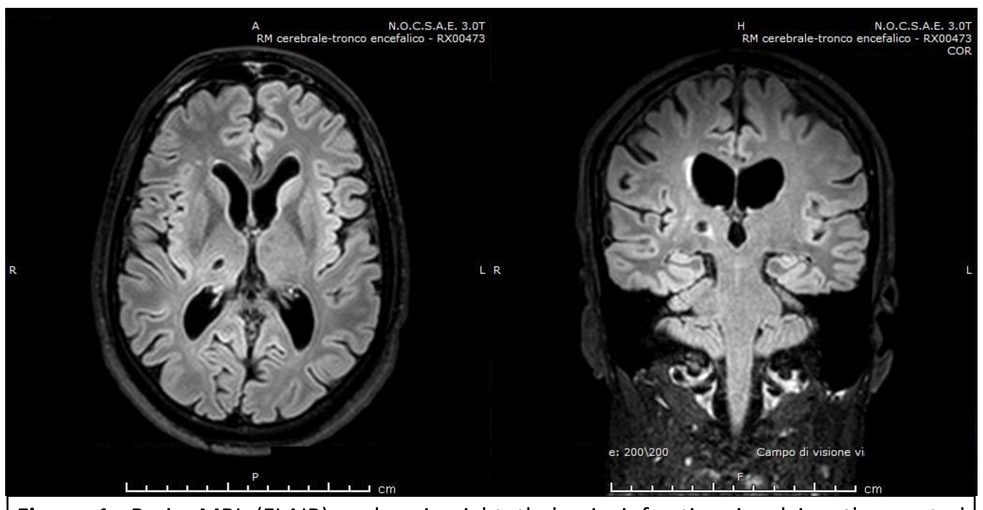

Figure 1. Brain MRI (FLAIR): chronic right thalamic infarction involving the ventral posterior nuclei

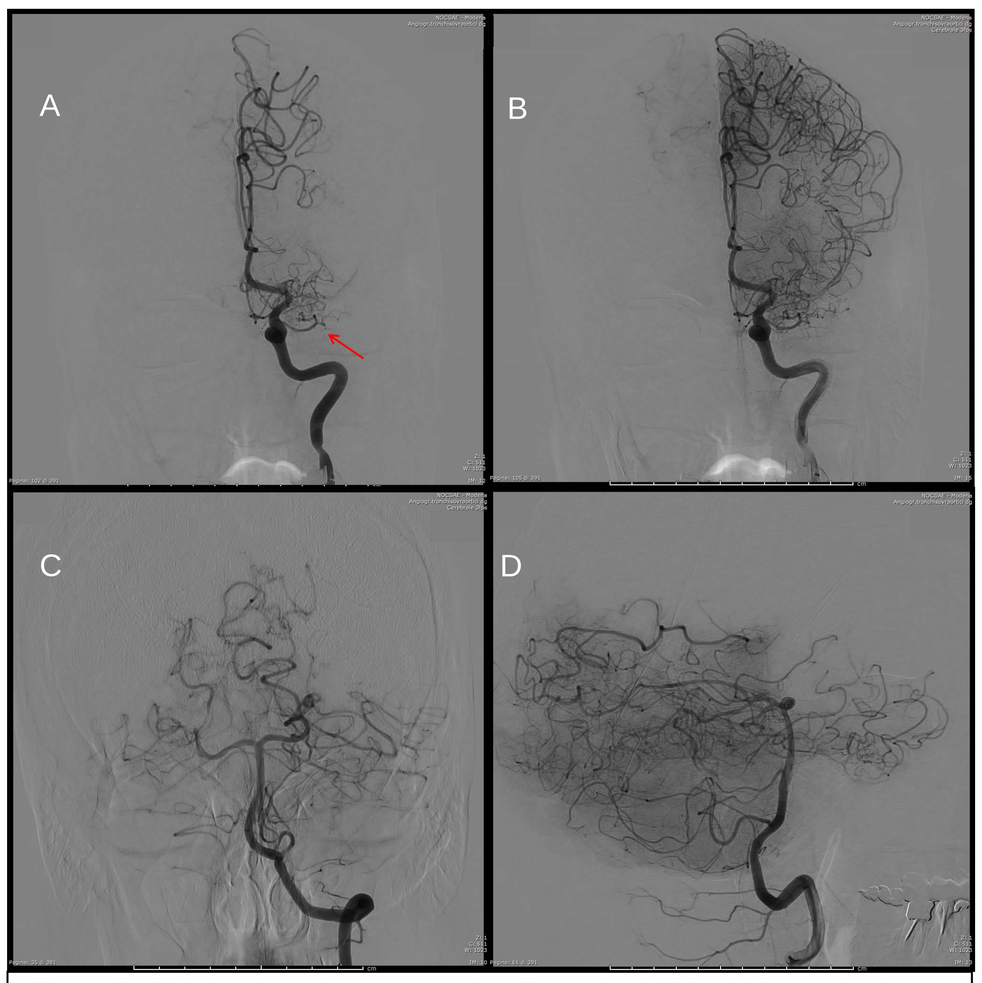

Figure 3. Digital subtraction angiography. A) proximal M1 middle cerebral artery occlusion with an extensive network of collateral vessels creating the characteristic puff of smoke appearance (arrow). B) late acquisition shows extensive collateral blood flow from the meningeal vessels. C) and D) collateral vessels originating from posterior circulation.

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