Recurrent brain edema in a patient affected by Cerebral amyloid angiopathy-related inflammation (CAA-ri)

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Background: CAA-ri is a rare and aggressive disease. It is a subtype of CAA, caused by an inflammatory response to the vascular deposits of ß-amyloid in the brain.

A 69-year-old woman was admitted to our hospital following episodes of recurrent transient paresthesias lasting 15 minutes, spreading from the perioral region to the right arm, hemithorax, and leg, followed by headache lasting 30-minutes. The neurological examination on admission was normal.

Diagnostic work -up:
- Blood tests: normal
- Brain MRI (fig.1)
- EEG: normal
- CSF analysis: high titer of anti-Aß autoantibody (91,4 ng/mg)

DIAGNOSIS OF CAA-RI

High dose corticosteroid therapy and antiepileptic drugs were started with clinical and neuroradiological improvement (Fig.2)

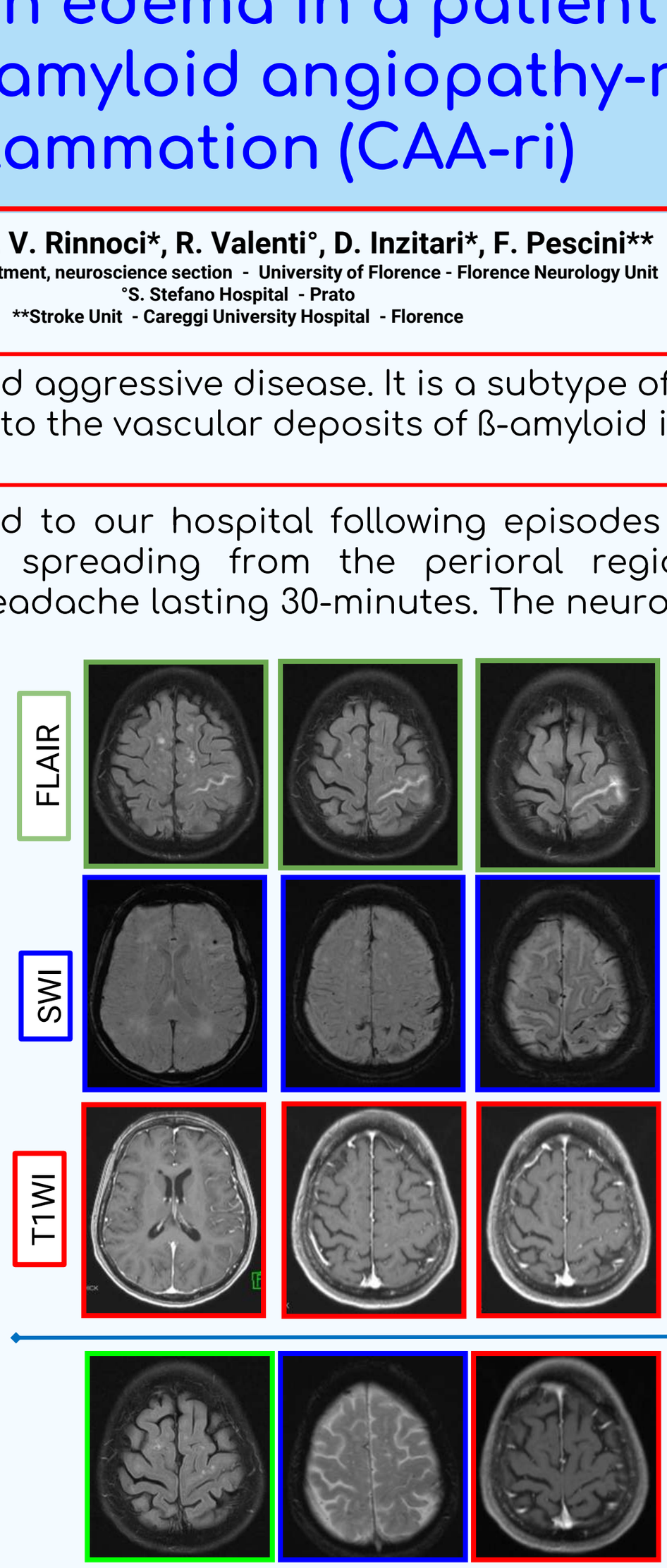

Fig.1: Brain MRI shows hyperintensity of left central sulcus (FRLAIR) with contrast-enhancement (T1WI). Cortical/subcortical microbleeds and bilateral hypointensities of parietal sulci are visible on the left side (SWI)

After one-month of therapy

Fig.2: Brain MRI shows disappearance of the hyperintense left central sulcus on FLAIR and no more contrast-enhancement on T1WI.

1 year later

A severe relapse occurred, characterized by hypoesthesia of right hemiface and arm, weakness and incoordination of right leg and balance impairment. The patient complained of difficulty concentrating and anomia. Brain MRI showed recurrence of brain edema (Fig.3)

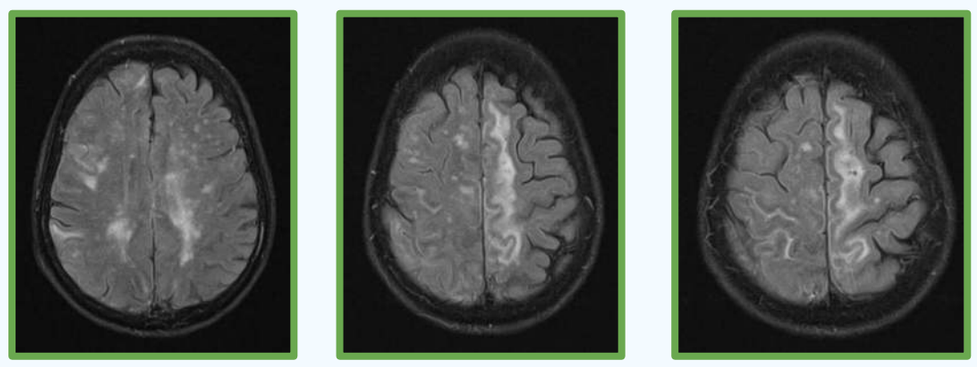

Fig.3. Brain MRI shows cortical/subcortical vasogenic edema of the left occipital lobe, medial portion of left frontal lobe, posterior portion of right temporal lobe and right fronto-parietal region associated to subarachnoid hemorrage0

High-dose corticosteroids therapy was started → Improvement of the cognitive and focal neurological deficits and resolution of vasogenic edema on MRI → Mycophenolate mofetil was prescribed (azathioprine-induced hepatotoxicity)

DISCUSSION: CAA is characterized on neuroimaging by white matter lesions, intracerebral haemorrhage, microbleeds, and cortical superficial siderosis. The most common presentation is acute hemorrhagic stroke. In a subset of patients, the amyloid deposition is accompanied by inflammation (CAA-ri), characterized by subacute cognitive decline, headaches, seizures, and stroke-like signs. Immunosuppressive treatment may improve both clinical and neuroimaging features as reported, although patients with recurrent symptoms have been described.

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