Superficial Siderosis in a Patient with Possible Cerebral Amyloid Angiopathy

A.Lupo[1], G. Arabia[1], G. Barbagallo[1], L.I. Manfredini[1], R. Nisticò[2], P. Perrotta[2], M. Casaletto[2], A. Gambardella[1], A. Quattrone[1,2]

1. Institute of Neurology, University Magna Graecia, Catanzaro, Italy.
2. Neuroimaging Unit, Institute of Bioimaging and Molecular Physiology, National Research Council (CNR-IBFM), Catanzaro, Italy

INTRODUCTION

Superficial siderosis (SS) of the brain is a rare syndrome with hemosiderin deposits in the leptomeninges, subpial layer, and ependymal surface. The most common clinical presentation is slowly progressive cerebellar ataxia, often associated with hearing impairment. Despite extensive investigations, the cause of bleeding remains frequently undetermined. Here we describe a patient with SS in whom an extensive laboratory investigations favored the diagnosis of possible cerebral amyloid angiopathy (*CAA*).

CASE REPORT

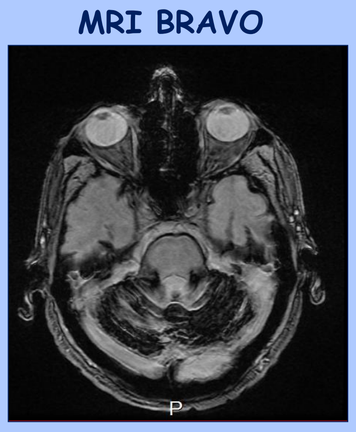
A 62-year-old man was admitted to our hospital with a 3-year history of instability of gait, urinary incontinence, slurred speech, dysphagia, memory impairment, orthostatic headache and dizziness. He complained hearing loss and mood depression with irritability during the last 5 years. These symptoms progressively worsened. There was no previous history of trauma, intradural surgery, neck or backache.

EXAMINATION

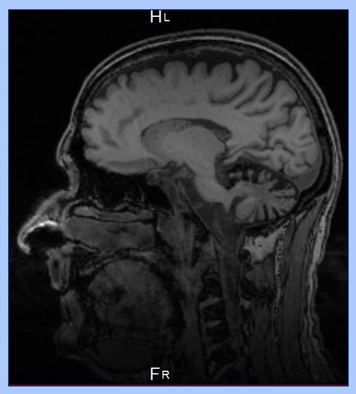
- **Neurological examination:** dysarthria, dysphagia, cerebellar ataxia and brisk tendon reflexes.
- **Audiometry test:** bilateral sensorineural hearing loss.
- **Cognitive evaluation:** mood depression.
- **Hematological investigation:** normal.
- **EMG and PEM:** normal
- **PESS**: deficit in conduction in lower limbs
- **Intracranial angiography:** normal.

❖**Brain MRI:** extensive rims of hypointensity on T2 throughout the leptomeninges, in the posterior fossa, cerebellar atrophy, without contrast enhancement.

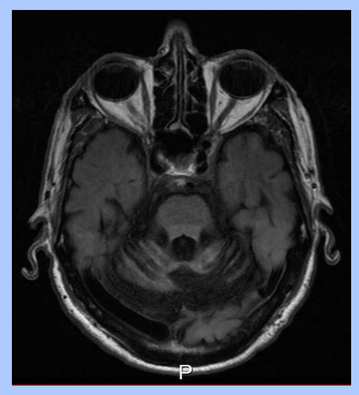
MRI BRAVO



MRI GRE-T2*



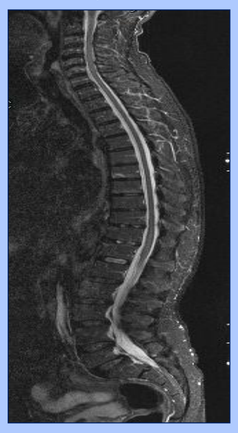
MRI FLAIR-T2



❖**CSF examination:** high levels of **total tau protein.**

❖**Spine MRI:** rims of hypointensity along the superficial surface of the whole spinal cord and medullary cone on T2-images.

MRI Spinal Cord



DISCUSSION

CAA is a small vessel disease characterized by deposition of beta-amyloid in the walls of the cortical and leptomeningeal vessels. The definite diagnosis of *CAA* requires a postmortem examination. According to the modified Boston criteria, as in our patient, the diagnosis in vivo of possible *CAA* requires the presence of focal or disseminated superficial siderosis, age≥55 years and absence of other causes of SS.

Modified Boston criteria for CAA

Possible CAA	Clinical data and MRI or CT demonstrating:	Clinical data and MRI or CT demonstrating:
	• Single lobar, cortical, or corticosubcortical hemorrhage	• Single lobar, cortical, or corticosubcortical hemorrhage or
	• Age ≥55 y	• Focal[b] or disseminated[c] superficial siderosis
	• Absence of other cause of hemorrhage	• Age ≥55 y
		• Absence of other cause of hemorrhage or superficial siderosis

CONCLUSION

There is now good evidence that the CSF levels of the tau protein are high in patients with probable *CAA* compared to controls. In this way, the present case indicates that this biomarker may have a role in the workup of patients with SS, especially to disentangle the several other causes of SS.

REFERENCES

1. Fearnley Julian M, Rudge Peter. *Superficial siderosis of the central nervous system.* Brain (1995). 118: 1051 – 1066.
2. Kumar Neeraj. *Superficial siderosis: associations and therapeutic implications.* Arch Neurol (2007). 64: 491 – 496.
3. Linn J, Halpin A, Demaerel P, Ruhland J, Giese A.D, Dichgans M, van Buchem M.A, Bruckmann H, Greenberg S.M. *Prevalence of superficial siderosis in patients with cerebral amyloid angiopathy.* Neurology (2010). 74: 1346 – 1350.

XLVI Congresso SIN Genova 2015