Superficial Siderosis in a Patient with Possible Cerebral Amyloid Angiopathy

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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.
- Audiology 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.

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.

REFERENCES


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.