

Superficial siderosis of the central nervous system: a rare cause of dementia with therapeutic consequences

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Abstract

A 75-year-old patient was evaluated for dementia. His past medical history included an ischaemic cardiomyopathy treated with aspirin daily. His neurological examination showed mild ataxia syndrome and central deafness. The neuropsychological examination did not suggest Alzheimer's disease. No specific aetiology was found from biological investigations, but MRI scans revealed a superficial siderosis, which was further confirmed with CSF exams. This case highlights the interest of MRI with echo-gradient-T2 weighted sequences in patients investigated for memory disorders. Once the diagnosis is known, specific preventive measures have to be taken: searching for a treatable source of bleeding and the interruption of antiplatelet aggregation or anticoagulant treatments.

Keywords: superficial siderosis, CNS, dementia, memory disorders

Superficial siderosis (SS) of the central nervous system (CNS) is a rare disorder induced by chronic subarachnoidal bleeding [1]. A triad of symptoms including deafness, cerebellar ataxia and pyramidal syndrome is classic, whereas dementia is inconstant and probably underdiagnosed [2, 3].

Case report

A 75-year-old man was referred for evaluation of progressive memory impairment. He had neither alcoholic nor tobacco intoxication. He reported two head traumas in the 90's and suffered from hypertension, type II diabetes and angina pectoris. His medication included anti-hypertensive agents, a statin, and aspirin (100 mg/day). Mini Mental State Examination (MMSE) was 16, with impaired language, praxis and executive functions. His memory impairment differed from the hippocampal profile usually seen in Alzheimer's disease (AD) (positive effect of cueing

and absence of intrusion errors in memory tests). Physical examination revealed both a static and a kinetic cerebellar syndrome and central deafness, with no pyramidal signs.

Biological standard tests for dementia evaluation were normal. Linear hypersignal T1, Flair and gradient-echo weighted sequences (T2*) and hyposignal in T2 were seen on the MRI (Figure 1) around the brainstem and cerebellar surface, sylvian sulcus and hippocampi. There was no evidence of vascular disease (no white matter hyperintensities, no microhaemorrhages) and no hippocampal atrophy. Cerebrospinal fluid was xanthochromic, with increased protein (1.91 g/l), and the absence of red blood cells.

The association of dementia, deafness, ataxia and typical MRI images is characteristic of SS. The circle of Willis was normal in angio-MRI. No arteriography was performed because of the patient's condition. SS was attributed to aspirin medication which was then stopped. Two years later, the patient remained stable.

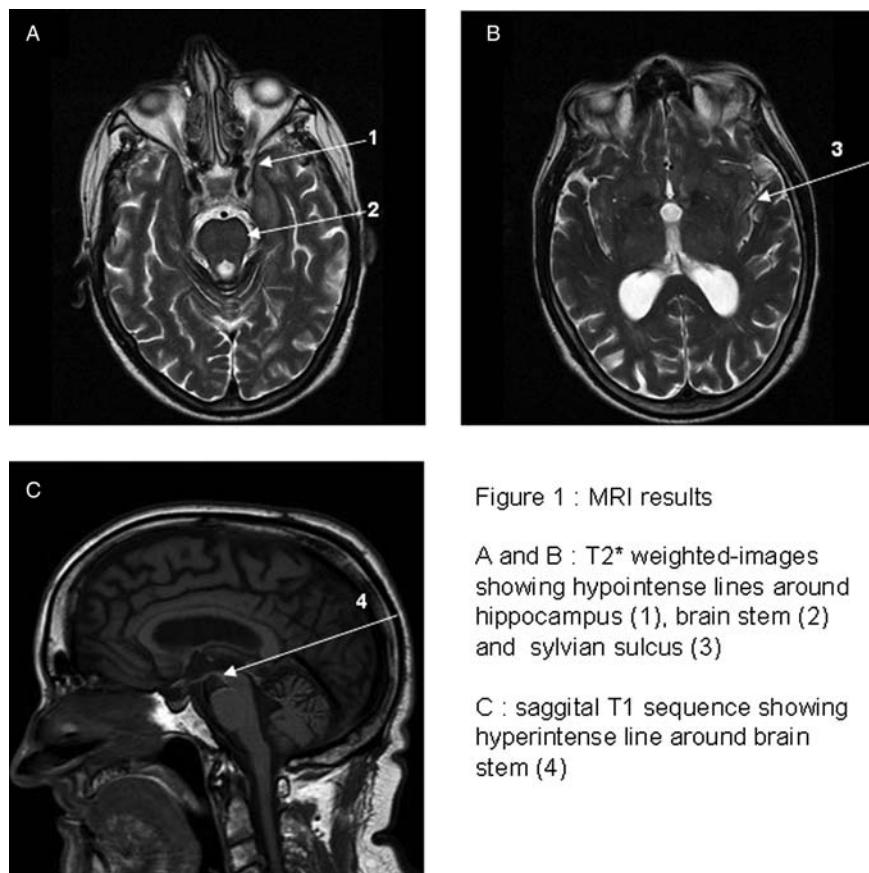


Figure 1 : MRI results

A and B : T2* weighted-images showing hypointense lines around hippocampus (1), brain stem (2) and sylvian sulcus (3)

C : sagittal T1 sequence showing hyperintense line around brain stem (4)

Figure 1. MRI results. (A and B): T2* weighted-images showing hypointense lines around hippocampus (1) brain stem (2) and sylvian sulcus (3). (C) Sagittal T1 sequence showing hyperintense line around brain stem (4).

Discussion

SS should be suspected in patients with dementia associated to deafness, cerebellar symptoms or pyramidal signs [2, 3]. Dementia is noted in 24% of patients presenting SS. Van Harskamp *et al.* reported six patients with SS and similar cognitive pattern: dysexecutive syndrome, behavioural dysfunctions and visual memory impairment [4]. A history of subarachnoidal haemorrhage, neurosurgery or head trauma is evocative but inconstant [5].

In our case, the diagnosis was finally realized by MRI, several years after the onset of the memory disorders. The cognitive deficits were not specific to any cause and not in favour of AD.

MRI is crucial for the diagnosis: a thin hypointense line is usually seen in T2* sequences around the brainstem, the cerebellum and the cerebral cortex. Cerebellar atrophy is also common [3, 6, 7]. LCS examination may reveal xanthochromia, high protein level, presence of red blood cells, but may also be normal (in 25% of patients) [5].

Because clinical and biological signs are non-specific and can be absent, standard evaluation of dementia, without any obvious aetiology, should include an encephalic MRI with T2* sequences.

Management of SS patients mainly relies on the identification of a chronic bleeding source and its treatment can stabilize or slow down its evolution [3]. If angio-MRI is normal, cerebral arteriography should be considered depending on patient's condition. The most common causes [3] are CNS tumour (21%), head trauma (13%), vascular malformation (9%) while amyloid angiopathy is rare (3%). Thirty-five per cent of SS are idiopathic. The patient's medication has to be thoroughly reviewed: stopping of antiplatelet aggregant or anticoagulant drugs is strongly advocated. If the patient takes such medication, expected benefits should be discussed regarding to the risk of cognitive impairment. Iron chelator has been proposed with inconsistent results and steroids sometimes improve the patient's condition [8, 9].

Conclusion

SS is a rare cause of dementia which can be easily diagnosed, leading to preventive recommendations. When standard imaging does not reveal a cause for dementia, MRI with T2*weighted sequences should be considered to avoid missed diagnosis.

Key points

- SS is a rare cause of dementia and memory disorders which can be easily diagnosed with MRI.
- MRI sequences include T1, T2 and gradient-echo weighted sequences (T2*).
- Depending on the context, arterial imaging should be used to identify and treat the source of chronic bleeding.
- The therapeutic consequence of this diagnosis is the cessation of antiplatelet aggregate or anticoagulant drugs.

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