Acta Neurol. Belg., 2011, **111**, 160-161

Superficial siderosis associated with multiple dural tears opening into a spinal epidural fluid collection

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Key words: Superficial siderosis; intraspinal cavity; multiple dural leaks; high-resolution MRI; dynamic CT-myelography.

Introduction

Superficial siderosis is characterized by the accumulation of hemosiderin all around the central nervous system. The cause is a chronic bleeding in the subarachnoid space, with preferential localisation around the cerebellum and the eighth cranial nerves that explains the typical clinical manifestations: cerebellar ataxia and sensorineural hearing deficits (1). The best diagnosis method is cerebral MRI which demonstrates the presence of a pathognomonic T2 or T2*-hyposignal rim at the brain surface. Dural pathology represents the most frequent aetiology, followed by tumours and vascular malformations (2). However, in one third of the cases, aetiology remains elusive. Currently, no treatment is able to reduce the toxicity of hemosiderin. The only treatment is the eradication of the source of bleeding, to prevent detrimental evolution towards debilitating end-stage disease.

Case history

We report on the case of a 63-year-old male with a fifteen-year history of hypoacousia. In addition to long-standing sensorineural hearing loss, he had been suffering from dysuria and worsening gait difficulties since ten years. He had no known history of heavy traumatism. The clinical examination demonstrated a static and dynamic cerebellar ataxia. The MRI of the brain revealed a T2-hyposignal rim predominant around the brainstem and cerebellum compatible with hemosiderin deposits (Fig. 1). The spinal MRI revealed the same T2-hyposignal rim along the spinal cord. Palliative iron chelating therapy with deferoxamine mesilate was not tolerated by the patient.

There was no MRI evidence for a tumour or a vascular malformation. The only other abnormality was an anterior spinal epidural fluid collection extending from C2 to T12 levels. Selective angiography of the brain and spinal cord were normal. In search of a dural pathology, a CT-myelography demonstrated an opening between the intradural space and the fluid-filled cavity, but the level of the leak was unclear. Dynamic myelography was considered but not realised due to the high radiation dose. Therefore we performed a high-resolution full spine 3T MRI 3D T2 weighted (thickness 1.8 mm), which led to the strong suspicion of six dural tears between T1 and T11 (Fig. 1).

Discussion

Thanks to MRI, superficial siderosis is more frequently diagnosed and its aetiology more commonly determined. During the last decade, dural pathology became the most often identified source of chronic bleeding, superseding tumours and vascular malformations (3). Meningocele and post-hemispherectomy cavities have been frequently associated with chronic bleeding. Superficial siderosis associated with spinal epidural fluid collection has been recently reported in 18 cases (4-13). Such an abnormality was not considered in the largest compiled series of 270 patients (2). It has been hypothesised that a small epidural vessel could bleed chronically but the relationship between the dural tear and vessel fragility remains to be proven. Recently, dynamic

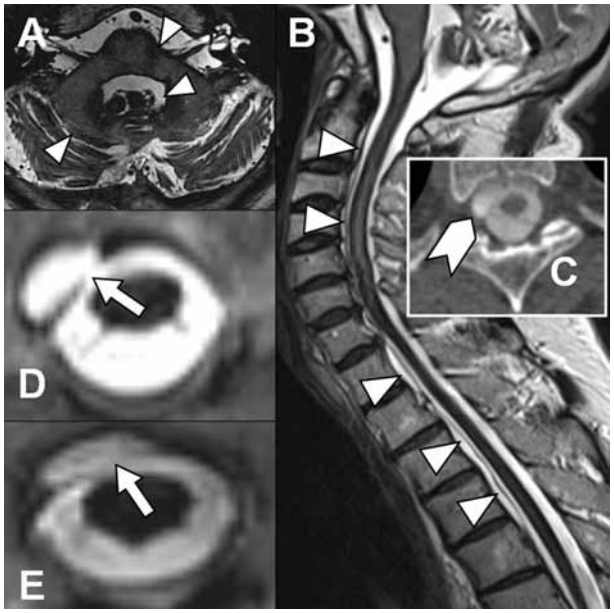

FIG. 1. — A. MRI of the posterior fossa, demonstrating the T2-hypointense rim around the surface of the brainstem, eighth cranial nerves and cerebellum; B. 3T spinal MRI, showing the anterior spinal epidural fluid collection (arrowheads) spreading from C2 to T12 (not shown); C. CT-myelography, with leaking of the contrast fluid from intradural space into the cavity; D, E. 3T spinal MRI: dural tears at the D6, D8-D9 levels, respectively.

CT-myelography has been proposed to identify the level of the dural leak when a spinal epidural fluid collection is present (14). However, this technique is time-consuming and delivers high radiation dose; 3T MRI of the spine with millimetric slices might be proposed for screening and dynamic myelography realized only if tears are suspected.

When a unique dural tear is identified, surgical intervention is able to normalize the cerebrospinal fluid levels in ferritin, transferrin, and /or tau protein (7, 8, 10-12). However, the follow-up periods were too short to support a clinical benefit in the long term. When multiple dural tears are identified, the challenge would be twofold: to identify the dural tear that is responsible for the chronic bleeding and to perform an effective occlusion of the culprit dural tear.

To our knowledge, this is the first report of superficial siderosis associated with a large spinal epidural fluid collection and multiple dural leaks identified by high-resolution 3T MRI.

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