

**SHORT REPORT**

# Superficial cerebral and spinal haemosiderosis caused by secondary tethered cord syndrome after resection of a spinal lymphoma

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Superficial haemosiderosis results from chronic subarachnoid haemorrhage during which haemosiderin is deposited in the leptomeninges around the brain, spinal cord and cranial nerves. We describe an exceptional case of superficial haemosiderosis characterised by two special aspects. (1) The cause was a secondary tethered cord syndrome due to dural adhesions which had developed 8 years after resection of a thoracic lymphoma and (2) an explorative neurosurgical procedure with complete untethering caused normalisation of the cerebrospinal fluid and stopped disease progression.

Superficial haemosiderosis of the central nervous system generally manifests initially with a progressive cerebellar syndrome and cranial nerve palsies, especially of the eighth nerves. Later in the course of the disease, spastic myelopathy and dementia may develop.<sup>1</sup> Superficial haemosiderosis results from chronic or intermittent haemorrhages into the subarachnoid space during which haemosiderin is deposited around the leptomeninges and subpial tissue surrounding the brain, spinal cord and nerve roots.<sup>2</sup> Its various aetiologies include vascular abnormalities, tumours and trauma induced dural lesions.<sup>3–5</sup> The cause remains unclear in approximately 50% of the patients,<sup>3</sup> thus precluding specific treatment to prevent any further progression.

We describe an exceptional case of superficial haemosiderosis caused by a secondary tethered cord syndrome, which developed 8 years after resection of a thoracic lymphoma.

## CASE REPORT

A 63-year-old man presented with a 1 year history of progressive gait unsteadiness, weakness and numbness of both lower extremities. He complained of increasing bilateral hearing loss over the past 3 years. Eight years earlier he had undergone a surgical resection of a low grade malignant B-cell non-Hodgkin's lymphoma of the thoracic spine (Th6) and subsequent radiation (from Th2 to L4).

Neurological examination revealed a mixed cerebellar and sensory ataxia with severe gait impairment, paraparesis with hyperreflexia, hypesthesia at the Th5–7 levels and below Th12, loss of joint position sense of the toes, pathological vibration sense of the lower extremities and moderate bilateral sensorineural hearing loss. Neuro-ophthalmological examination showed a pathological head thrust test to the right side, indicating a high frequency deficit of the vestibulo-ocular reflex, gaze evoked nystagmus, head shaking nystagmus to the left side and saccadic smooth pursuit.

## Magnetic resonance imaging

In 1998 (before resection of the spinal lymphoma), cerebral and spinal MRI showed no signs of haemosiderosis (fig 1A). Eight

years after resection of the lymphoma, cerebral and spinal MRI revealed the typical finding of superficial haemosiderosis of the central nervous system: a hypointense rim around the brainstem, vermis and cerebellar convexities, and along the interhemispheric and sylvian fissures (fig 1B), as well as linear haemosiderin deposits on the surface of the whole spinal cord (fig 1C). A complementary three dimensional CISS (constructive interference in steady state) MRI showed an adhesion between the spinal cord and the dorsal dura at the level of Th7 (fig 1D).

## Cerebrospinal fluid examinations

CSF tests were performed 4 years and 1 month before and once after admission to our department, and again 3 months after the second surgery (see below) (table 1). The repeated lumbar punctures revealed xanthochromic CSF with an increasing number of erythrocytes and macrophages loaded with haemosiderin.

On the basis of these findings, the site of bleeding was suspected, and an explorative operation was performed.

## Surgical procedure

With the patient in the prone position, the old surgical approach was chosen but was extended caudally. After cautious dissection of the scar, in part calcified, and a laminectomy of the seventh thoracic vertebra, a small epidural cyst was opened and removed. The dura was exposed, and ultrasound showed adhesions of the spinal cord to the dura at the level exposed. After the dura was opened, widespread arachnoidal thickening with strong dural adhesion was seen (fig 1E). Once the arachnoid was perforated, CSF drained under pressure. Now a thickened, in part calcified, tissue string connecting the spinal cord to the arachnoid layer could be observed; it caused a tethering of the spinal cord at this level (fig 1F). The tissue string was supplied by multiple capillaries, which readily started to bleed when manipulated. After coagulation and removal of the tethering tissue, the spinal cord floated freely without restriction (fig 1G). A primary closure of the dura was performed.

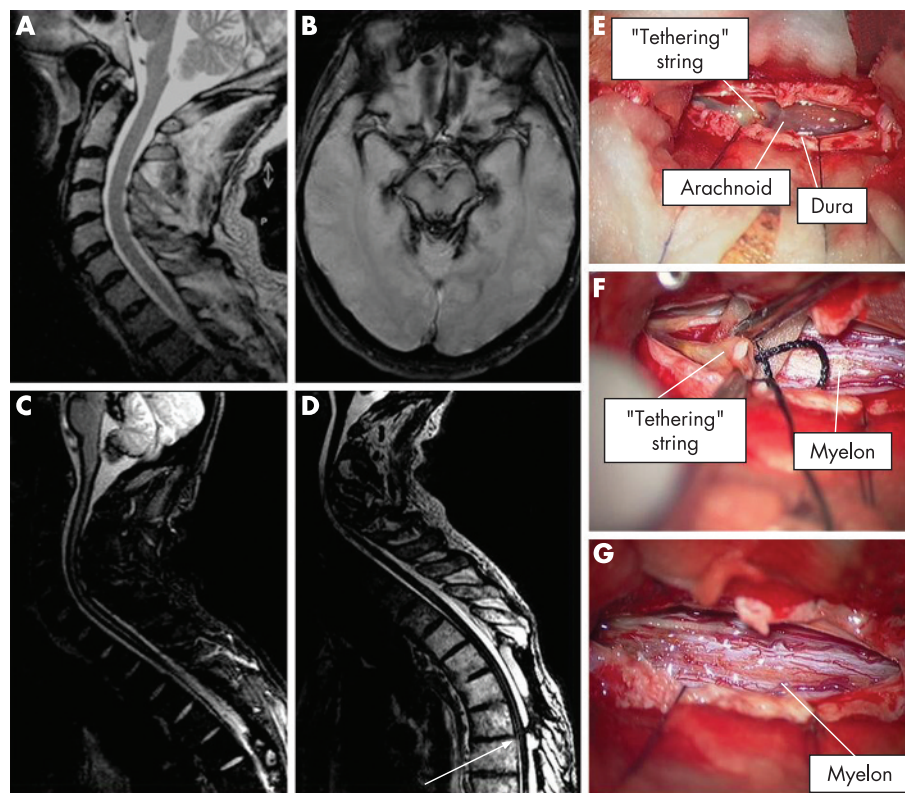
## Postoperative course

At a 3 month follow-up, the patient was still neurologically stable. CSF was normal, without xanthochromia (table 1). MRI of the brain and spine showed persistent haemosiderosis.

## DISCUSSION

A combination of different MRI techniques suggested the possibility of a secondary tethered cord syndrome. Dural tears are assumed to produce scar formations with fragile blood vessels, which bleed intermittently and thereby cause superficial haemosiderosis.<sup>6,7</sup>

Because of the progressive course of the disease, it is imperative to identify the bleeding source. Thus an explorative



**Figure 1** (A–D) Magnetic resonance imaging. (A) In 1998 (before resection of the spinal lymphoma), sagittal T2 weighted MRI showed no signs of haemosiderosis. (B) In 2006 (8 years after resection of the spinal lymphoma), transversal T2 weighted gradient echo MRI (TR/TE=1000/22 ms) showed the superficial siderosis as a hypointense rim of haemosiderin deposits along the sylvian fissures, around the midbrain and cerebellar folia. (C) Sagittal T2 weighted gradient echo (A; TR/TE=500/26 ms) MRI revealed linear haemosiderin deposits around the brainstem and spinal cord. (D) High resolution sagittal three dimensional CISS (constructive interference in steady state) MRI (TR/TE=8.8/4.4 ms) of the cervical and thoracic spinal cord showed an adhesion between the spinal cord and the dorsal dura at the Th7 level (arrow). (E–G) Intraoperative images. (E) After opening of the dura, thickened arachnoid as well as a tissue string connecting the dura and spinal cord was seen. (F) After opening of the arachnoid, the spinal cord was displayed. The connecting “tethering” tissue string was removed cautiously. (G) After removal of the string, the spinal cord was mobile and unbent.

**Table 1** Cerebrospinal fluid examinations: CSF tests were performed 4 years and 1 month before and once after admission to our department, and again 3 months after the neurosurgical procedure with complete untethering

Date	CSF	Cell number/ $\mu$ l	Erythrocytes/ $\mu$ l	Ferritin (ng/ml)	Protein (mg/dl)	Cell differentiation
Before treatment						
Aug 2002	Xanthochromic	23	3221	309	68	Abnormal (siderophages)
Jan 2006	Xanthochromic	12	2300	Not done	130	Abnormal (siderophages)
Feb 2006	Bloody	29	9899	269	112	Abnormal (siderophages)
After treatment						
July 2006	Clear	7	3	111	108	Normal

surgical approach and untethering seemed justified. The bleeding and progression were effectively stopped. This was confirmed by the repeated CSF test 3 months after surgery which was completely normal without xanthochromia. Long term outcome data of patients with superficial haemosiderosis who have undergone closure of the leakage are not yet available. The haemosiderin deposits seem to remain on the surface of the nervous system,<sup>6</sup> even after successful occlusion of the bleeding site. It remains unclear if these haemosiderin deposits continue to injure neuronal tissue.<sup>4</sup>

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