Hearing Loss and Bilateral Recurrent Peripheral Facial Nerve Palsy in Superficial Siderosis

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Citation

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Abstract

Superficial siderosis of the central nervous system (SSCN) is a very rare disorder. The clinical syndrome of SSNC consists of sensorineural hearing loss, cerebellar ataxia and myelopathy. A 58 year-old man who has a bilateral sensorineural hearing loss, recurrent peripheral facial nerve palsy and gait instability for 10 years with a history of recurrent head trauma is presented. The neurological examination revealed bilateral sensorineural hearing loss, cerebellar ataxia, neurogenic bladder, anosmia and mild spasticity of the lower extremities.
In patients with progressive bilateral cochleo-vestibular deficit and recurrent peripheral facial nerve palsy of unknown etiology, MRI is the examination of choice to confirm SS.

INTRODUCTION

Recurrent or persistent haemorrhage into the subarachnoid space causes superficial siderosis of the central nervous system (CNS). Superficial siderosis (SS) of the central nervous system (CNS) is considered a rare disorder resulting from deposition of the iron-containing pigment haemosiderin in the leptomeninges and subpial tissue. In the past SS could be diagnosed only by surgical inspection of the brain or at post-mortem examination. Since the advent of magnetic resonance imaging (MRI), a reliable tool for in vivo detection of SS appears to exist with signal loss at the CNS surfaces on T2-weighted scans being pathognomonic. The clinical manifestations include an insidiously progressive cerebellar ataxia, dysarthria and sensorineural hearing loss. Later in the course of the disease, a spastic myelopathy and dementia may develop ([1]). The usual sources of the subarachnoid bleeding are dural abnormalities, vascular lesions, the history of neuro-surgical procedures, and tumours. However, the source of the bleeding remains obscure in approximately 40% of the cases ([1,2,3]).

In this case report, the presentation, diagnosis, pathogenesis, treatment and prognosis of SS are reviewed.

CASE REPORT

A 58-year-old man suffered from recurrent peripheral facial nerve palsy and bilateral hearing loss. He complained of a progressive hearing loss bilaterally as well as of recurrent episodes of positional vertigo and peripheral facial nerve palsy which occurred during several days for 10 years. He was experienced multiple head trauma in his medical history.

Physical examination was remarkable for end-gaze horizontal nystagmus to the left, bilateral anosmia, slurred speech, bilateral symmetrical pyramidal signs, and bilateral sensorineural hearing loss. Cognition was normal. Neurogenic detrusor overactivity found in the urodynamic examination. The patient was unable to perform tandem walking and had exhibited a very unsteady gait. There was also slight dysmetria and dysdiadochokinesia in the upper extremities. Heel-shin test was severely disturbed. Romberg's sign was positive.

The electroencephalogram (EEG) was normal. MRI examination showed linear hypointensities in axial T2- and T2-weighted images, mostly prominent in the posterior fossa, particularly pronounced in the cerebellar folia at the level of the hemispheres, cerebellar vermis (Fig 1) and cervical spinal cord (Fig 2). Cerebral angiography excluded an associated venous angioma or an arteriovenous malformation. All spinal MRI investigation was normal.

Further large systemic investigations were performed with blood laboratory testing (TPHA, Lyme, lead and mercury measurements), electrocardiogram, chest radiograph,

abdominal sonography, electroencephalogram, cerebral angiography, spinal MRI, and ascending lumbarcervical pan myelography. All of these examinations were normal. The patient rejected the lumbar puncture.

Axial T2-weighted MRI scan (Figure 1a and 1b) show heavy superficial signal loss at the level of pial surface of the pons and cerebral peduncles.

Figure 1

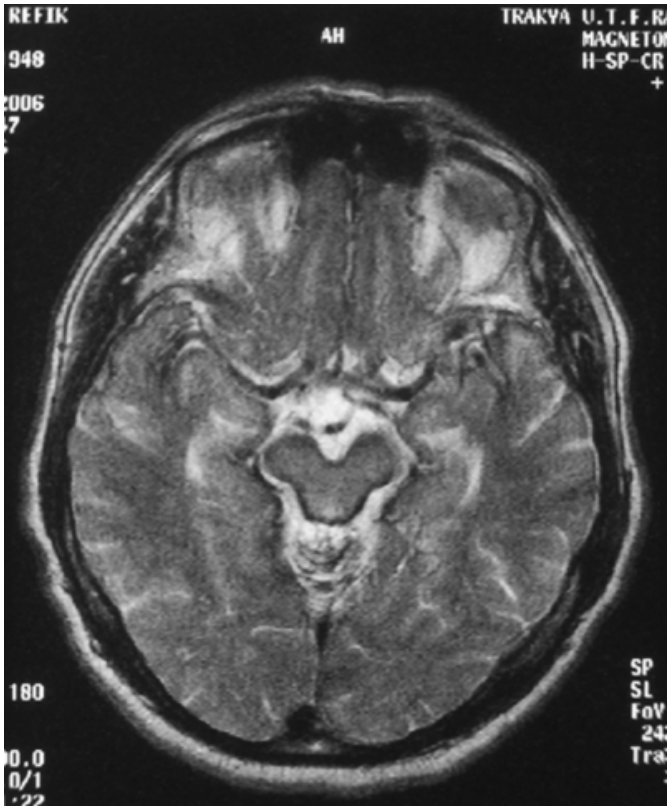


Figure 2

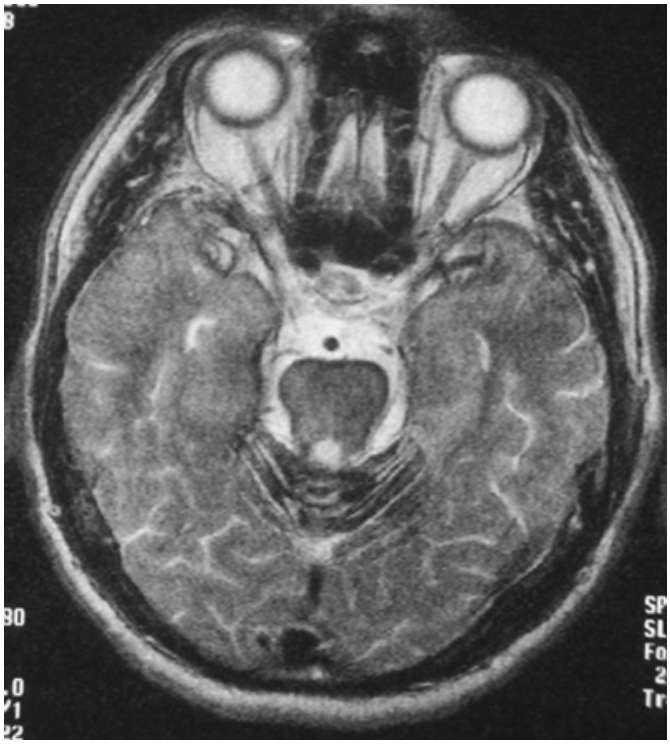
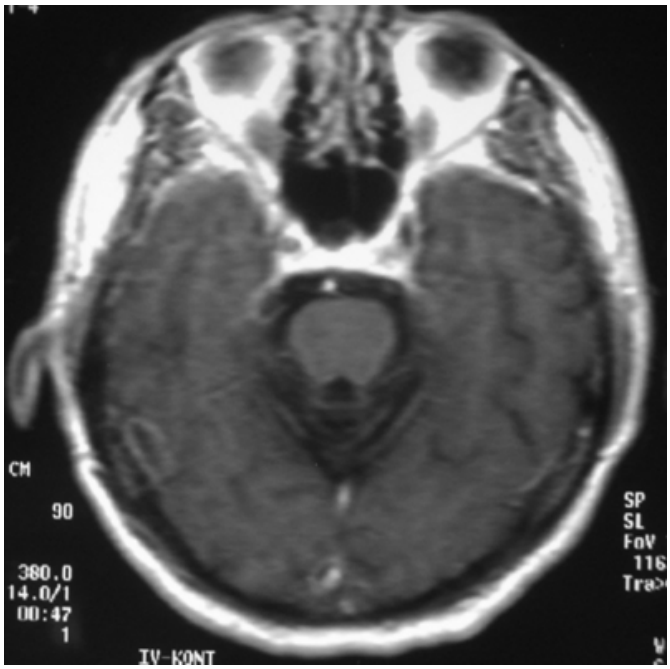
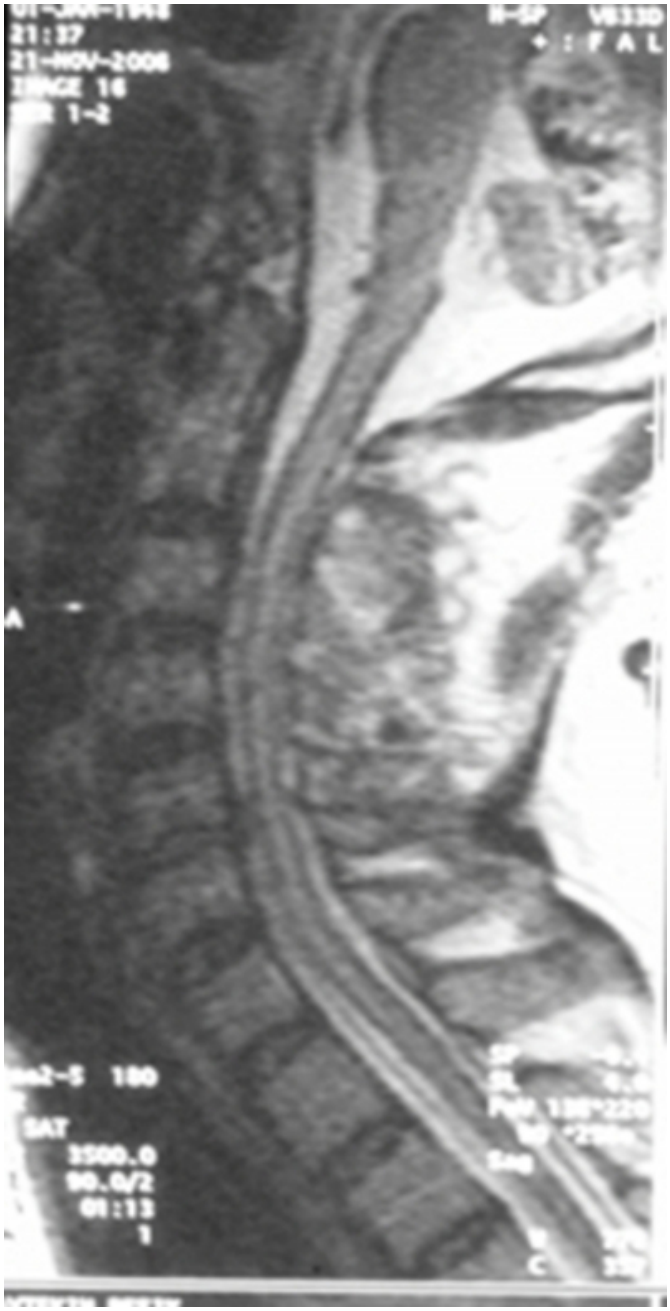


Figure 3



Parasagittal T2-weighted images of patient (Figure 2) show lineer superficial hypointensity at the cervical spinal cord.

Figure 4



Therapy with trientine, a chelating agent, was initiated (1800 mg trientine dihydrochloride ? day equivalent to 1200 mg trientine base). At the follow-up visit 6 months later, there was a slight improvement of dysarthria.

DISCUSSION

Superficial siderosis of the central nervous system is a rare condition characterized by deposition of hemosiderin in the leptomeninges, subpial tissue and spinal cord. Magnetic resonance imaging provides prompt diagnosis of this entity.

The patient reported here showed the characteristic features of this disease: in our case cerebellar ataxia developed insidiously over years, and bilateral sensorineural deafness was present. Upper motor neuron signs reflected spinal cord involvement as described before. Besides these cardinal features, however, the variety of symptoms such as anosmia, neurogenic bladder, bilateral peripheral facial nerve palsy possibly associated with superficial siderosis is wide ([1,2,3,4,5]). The progression of symptoms is generally very slow and several years, even decades, may pass before medical attention is sought.

The underlying source of superficial siderosis could be identified in approximately 60% of 68 cases studied in a literature survey. In these patients, CSF cavity lesions such as hemispherectomy, meningocele, encephalocele, chronic sub-occipital hematoma and cervical root lesions such as avulsion and epidural cysts were found in 47%, tumors in 35% (ependymoma and less commonly meningioma, oligodendroglioma, pineocytoma, paraganglioma) and vascular abnormalities in 18% (arteriovenous malformation, aneurysm, cavernous angioma). The source of the hemorrhage remained obscure in 40% of the patients ([1,11]).

We noticed a slight reduction of dysarthria and gait ataxia after a treatment period of 6 months. This is in line with the results of River et al. ([3]) who found a slowing of progression of superficial siderosis over a period of 2 years and a reduction of CSF iron, but not of CSF ferritin, suggesting that trientine is only partially effective ([8,9]). We could not investigate CSF ferritin levels. However, trientine treatment had no effect on the course of the disease in two other patients ([3,10]).Angstwurm et al present a forester aged 66-years with ataxia, myelopathy, and bilateral hearing loss due to superficial CNS siderosis associated with anti-Ri antibodies whose condition improved impressively after steroid therapy ([7]).

To conclude, it is important to suspect superficial siderosis of the central nervous system in clinical practice in order to avoid delay in diagnosis and treatment due to the insidious and protracted course of the disease, its rarity and non-specific features. Even slight MRI features consistent with an initial degree of SS should not be underestimated.

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