

# Trochlear Nerve Palsy Associated With Superficial Siderosis of the Central Nervous System

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**Abstract:** A 56-year-old man with superficial siderosis of the central nervous system (SSCN) presented with complaints of trochlear palsy, visual field defects, gait ataxia, and hearing loss. He had no history of trauma and there were no signs of tumors or aneurysms. T2-weighted magnetic resonance imaging demonstrated characteristic hypointensity in the meninges. We believe that SSCN should be added to the differential diagnosis of trochlear nerve palsy. **Jpn J Ophthalmol 1997; 41:19–22** © 1997 Japanese Ophthalmological Society

**Key Words:** Diplopia, MRI, superficial siderosis, trochlear palsy, visual field defect.

## Introduction

Superficial siderosis of the central nervous system (SSCN) was first recognized at autopsy in the early 1940s;<sup>1</sup> there are only about 50 reports in the English literature.<sup>2</sup> The usual etiology is multiple small episodes of subarachnoid hemorrhage, from any source,<sup>3,4</sup> with the intra- and extracellular deposition of hemosiderin in the leptomeninges, subpial tissue, spinal cord, and cranial nerves.<sup>2,3,5–12</sup> Diagnosis of SSCN could formerly be made only at autopsy or during a neurosurgical procedure.<sup>1,3–5,7–9,12</sup> Now, however, it can be made noninvasively with magnetic resonance imaging (MRI). T2-weighted images consistently show marginal hypointensity involving several parts of the central nervous system.<sup>2,6,10,11,13–17</sup> Chief clinical manifestations of SSCN include hearing loss, cerebellar dysfunction, and dementia;<sup>1–3,5–19</sup> ocular involvement is very rare.<sup>12</sup> We describe an SSCN patient whose symptoms were trochlear palsy, visual field defects, gait ataxia, and hearing loss. We believe that this is the first case of trochlear nerve palsy associated with SSCN.

## Case Report

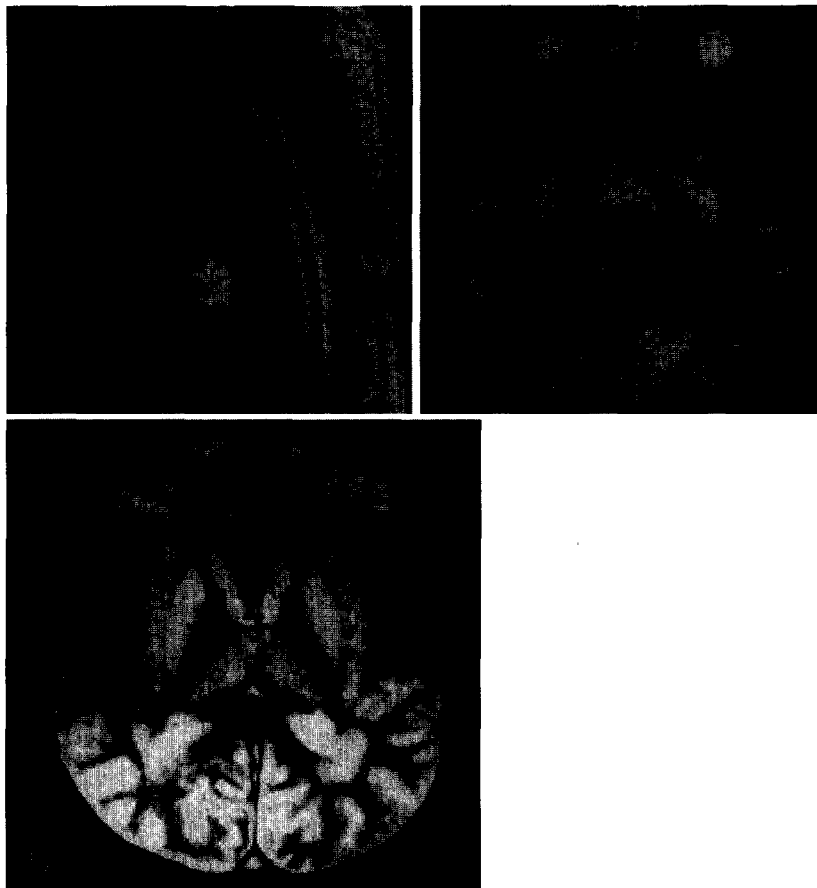
A 56-year-old man with a 10-year history of progressive ataxia and an 8-year history of diabetes mellitus, visited the local hospital otorhinolaryngology department complaining of tinnitus, in April 1992, and was diagnosed as having sensorineural deafness. Computed tomography (CT) of the brain revealed cerebellar atrophy. He was referred to the neurology department of Hokkaido University Hospital in July. A T2-weighted MRI revealed hypointensity on the surface of the spinal cord, brain stem, and Sylvian fissure, diagnosed as SSCN (Figure 1). His cerebrospinal fluid was xanthochromic, but MRI and angiography of the cerebral vessels failed to identify the bleeding source.

He complained of diplopia and was referred to our clinic in September. On initial examination, his best-corrected visual acuity was 1.0, bilaterally; the anterior segments, fundus, and intraocular pressure were normal. Eye position was 3° L/R hypertropia; tilting the head to the right abolished the hypertropia and diplopia disappeared; tilting to the left increased both the upward deviation of the left eye and diplopia. Hess-Lees red-green testing revealed left trochlear nerve palsy.

In August 1993, he suddenly noticed right homonymous hemianopsia (Figure 2a). Visual activity was

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**Figure 1.** T2-weighted MRI images: brain and spinal cord hypointensity on surface of spinal cord (**A**), brain stem (**B**), and the Sylvian fissure (**C**: arrows). High-intensity lesions, bilateral occipital lobes (**B**: arrows) (October 1993).

unchanged. The visual field soon cleared slightly, but the right superior homonymous quadrantanopsia remained. At this time, MRI revealed cerebral infarction in the left occipital lobe (Figure 2b).

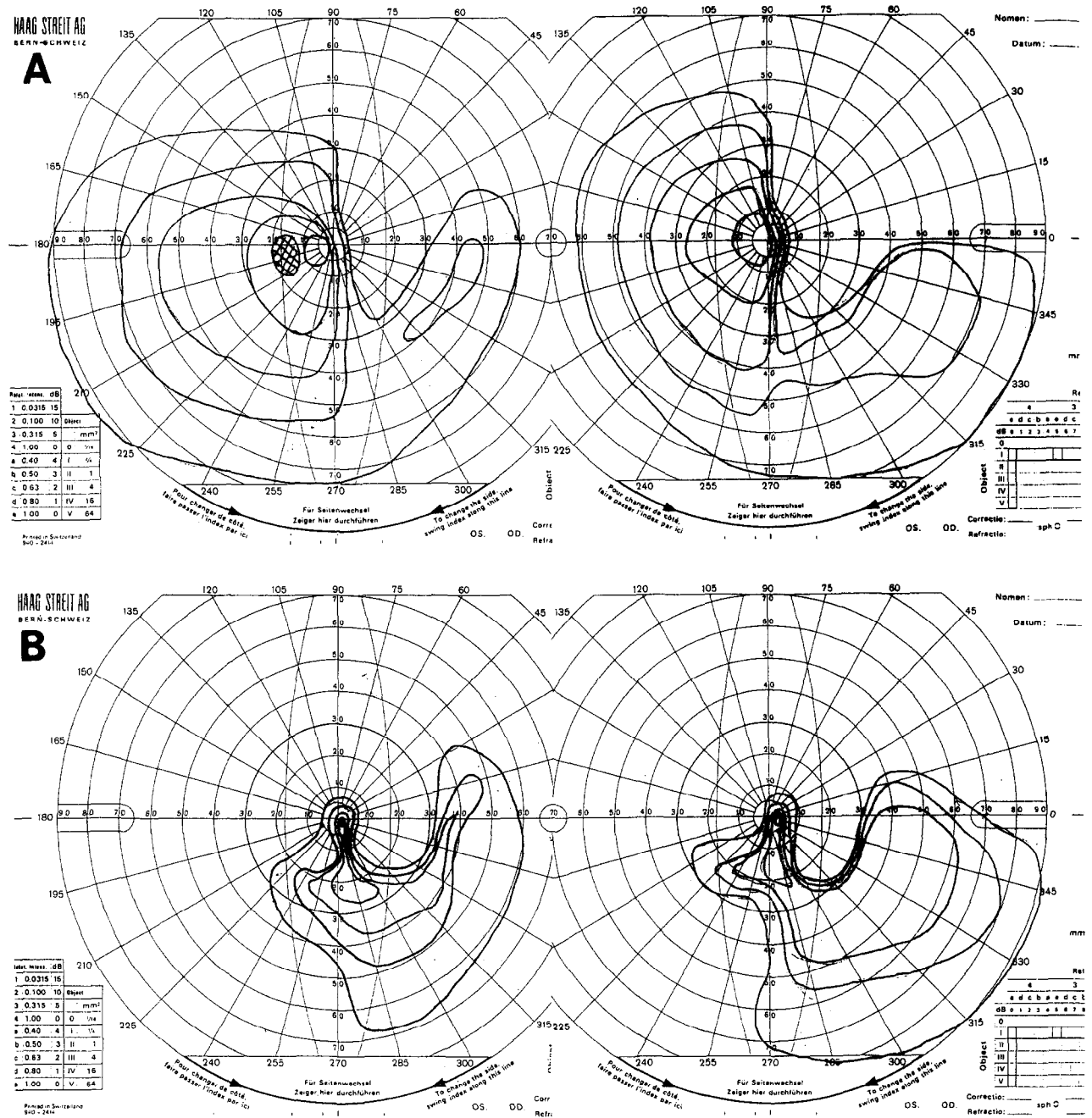
In March 1995, he noticed left visual field defects. Visual field testing revealed development of left homonymous hemianopsia (Figure 2b). Visual activity was unchanged; MRI revealed hemorrhagic cerebral infarction in the right occipital lobe (Figure 3). Treatment included an iron-chelating agent, trientine hydrochloride, 1.5 g/day; River et al<sup>11</sup> reported that this treatment produced a beneficial effect in stabilizing an SSCN patient's condition and reversing some of the symptoms in a relatively short period of time. However, after 6 months of therapy, our patient's ocular involvement remained unchanged.

### Discussion

The clinical findings of SSCN are not specific. In this case, the patient manifested diplopia due to trochlear palsy, visual field defects, sensorineural hearing loss, and cerebellar dysfunction. A T2-weighted

MRI showed the marked hypointensity on the surface of the spinal cord, brain stem, and Sylvian fissure that are characteristic of SSCN.

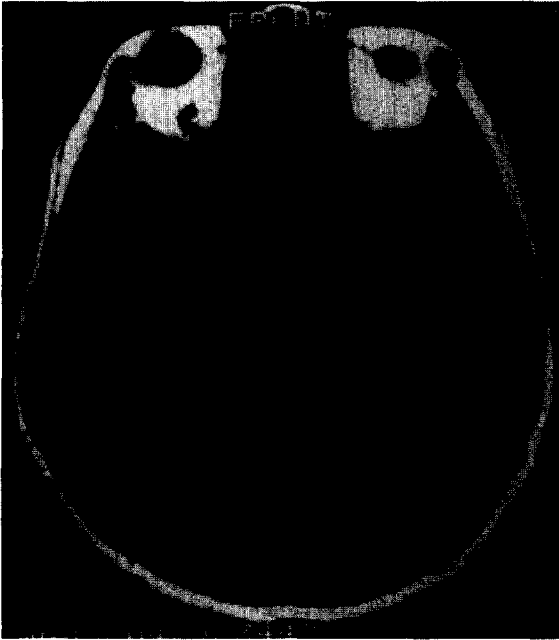
The etiology usually found is repeated episodes of subarachnoid hemorrhage.<sup>3,4</sup> Trauma, tumors, and vascular malformations may sometimes be the source of bleeding.<sup>1,5-9,11,12,14,18</sup> Stevens et al<sup>2</sup> reported that the sources of bleeding were unknown in 14 of 40 SSCN cases. In our patient, the cerebrospinal fluid was xanthochromic, but MRI and angiography of the cerebral vessels failed to identify the bleeding source in the first examination in September 1992. Bleeding of the right occipital lobe was detected in March 1995 (Figure 3). Visual field defects associated with SSCN are very rare;<sup>12</sup> in the present study, they may be caused by cerebral infarction of the occipital lobe. There have been several reports of cranial nerve defects (II, V, VII, VIII) in SSCN,<sup>11,16</sup> but we believe our current report is the first describing an association between SSCN and trochlear nerve palsy, possibly resulting from hemosiderin deposition around the proximal trochlear nerve.



**Figure 2.** (A) Visual field testing: right homonymous quadrantanopia (August 1993). (B) Visual field testing: addition of left homonymous hemianopia bilaterally (March 1995).

The etiology of trochlear nerve palsy is unknown in 35% of patients; 54% of these recover spontaneously.<sup>19</sup> Known causes include trauma, inflammation, neoplasms, and vascular disorders.<sup>19</sup> Vascular causes include diabetes, aneurysm, and arteriovenous malformation. Our patient had no episodes of trauma or inflammation; MRI revealed no tumors; angiography found no vascular disorders such as an-

eurysms or arteriovenous malformations. The influence of the patient's diabetes mellitus appeared to be negligible because it was well controlled by careful diet. We thus concluded that the trochlear nerve palsy was related to the SSCN demonstrated by the MRI findings. Although SSCN is rare, it should be included in the differential diagnosis of trochlear nerve palsy. MRI should be used for unexplained



**Figure 3.** T1-weighted MRI image: hyperintensity on surface of right occipital lobe (March 1995).

trochlear nerve palsy seen with other neurologic disorders, such as cerebellar ataxia and hearing loss.

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