

# Superficial Siderosis of the CNS: MR Diagnosis and Clinical Findings

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**PURPOSE:** To report the clinical and neuroradiologic findings of superficial siderosis of the CNS, due to chronic subarachnoid bleeding of unknown origin. **MATERIALS AND METHODS:** We observed seven cases. The main clinical manifestations were progressive deafness and ataxia. Four patients had had previous cranial or cervical trauma, with root avulsion in two, many years before onset of deafness and ataxia. Neuroradiologic studies included MR (0.5 T in four and 1.5 T in three) and angiography of the brain in all cases, CT in six cases, MR of the spine in six, and myelography in four. **RESULTS:** MR demonstrated a rim of marked hypointensity in T2-weighted images, consistent with hemosiderin deposits, on the surface of cerebellum, brain stem, inferior part of cerebral hemispheres, and spinal cord. CT showed cerebellar atrophy in five cases, and a rim of mild hyperdensity around the brain stem in two. Angiographic studies were negative. Myelography showed cervical nerve root avulsion in two cases and a cervicodorsal extradural cyst in one. Cerebrospinal fluid contained RBCs in all the six examined cases. **CONCLUSION:** Although CT may occasionally suggest the diagnosis of superficial siderosis, MR demonstrates this abnormality to better advantage.

**Index terms:** Iron, brain; Hemosiderosis; Arachnoid, hemorrhage

AJNR 14:227-236, Jan/Feb 1993

Superficial siderosis (SS) of the central nervous system (CNS) is a rare condition, first described by Noetzel in 1940 (1), characterized by deposition of hemosiderin on the leptomeninges, on the surface of the brain, cerebellum, brain stem, cranial nerves, and spinal cord. Progressive bilateral hearing loss and ataxia are the main clinical manifestations (2, 3). The deposition of hemosiderin is due to repeated chronic subarachnoid or intraventricular bleeding. The majority of the neuropathologic cases described in the literature are secondary to repeated hemorrhages from tumors

(especially ependymomas) (2, 4), vascular malformations (5), subdural hematomas (5, 6), or are secondary to hemispherectomy (6, 7). However, in approximately one fourth of the cases, no evidence of the source of bleeding was found, even at autopsy (6, 8).

Before the introduction of magnetic resonance (MR) imaging, the diagnosis was difficult, usually made only at necropsy, or at operation (2). MR can now detect hemosiderosis in vivo due to the strong paramagnetic effect of iron-containing pigments (4).

We report the clinical and neuroradiologic findings of seven cases of SS of the CNS, in which no definite source of hemorrhage could be found. However, our series and a recent observation (Nudelman J, Seltzer S, Cohen WA, paper presented at the annual meeting of the American Society of Neuroradiology, Washington, June 1991) suggest the importance of old cranial or cervical traumas as a possible cause of SS of the CNS.

## Materials and Methods

The seven cases, six men, one woman, age ranging from 42 to 56 years (mean, 49 years), were collected during

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Received October 7, 1991; revision requested November 26; final revision received March 3, 1992 and accepted March 9.

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AJNR 14:227-236, Jan/Feb 1993 0195-6108/93/1401-0227

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the period 1988–1991 in four different centers. Two of these cases have been recently reported in a neurologic journal (9). The clinical presentation was progressive deafness for all the patients, lasting from 2 to 12 years prior to the diagnosis. Ataxia was a prominent symptom for six of seven patients; in the seventh patient (case 5), ataxia was mild (Table 1). Three patients (cases 3, 5, and 7) had severe cervical trauma (with documented cervical root avulsion in two) 10 to 25 years before the beginning of hearing loss. In that period, two of these patients (cases 3 and 7) also had repeated episodes of seventh nerve palsy. One patient (case 2) experienced a cranial trauma 20 years before the hearing loss, with disturbances of consciousness lasting for 8 days, but no sequelae. One patient (case 1) had been an alcoholic, but had given up alcohol 8 years before the onset of the present symptoms. Twenty-eight months after MR, this patient died from pneumonia in a cachectic condition in another institution. Autopsy was not performed. Cerebrospinal fluid (CSF) examination was available in six out of seven cases. It was performed within 1 month after MR study in five cases, and showed the presence of red blood cells (RBC) (cell count: 180 to 700 RBC/mL), xanthochromia, and high protein content. In the sixth case (case 7), CSF examination was performed at the beginning of the symptomatology, 8 years prior to MR, during myelography showing posttraumatic cervical root avulsion, and was reported as normal. An examination repeated in 1991, 2 months after MR study, demonstrated RBCs in the CSF

(see Table 1). In case 2, three subsequent lumbar punctures performed at 1-month intervals showed constant presence of blood with RBC ranging from 150 to 350/mL.

Pre- and postcontrast CT examinations of the head were available in six cases (Table 2).

MR was performed in four cases with 0.5-T magnet, and in three cases with 1.5-T magnet. In all the patients, brain studies were performed with at least sagittal T1-weighted and transverse spin-echo (SE) intermediate and T2-weighted images; often sagittal or coronal SE intermediate and T2-weighted images were also available. Number of excitations was 2–4 for T1-weighted images, and 1–2 for T2-weighted images. The matrix was  $256 \times 256$ . Gradient-echo (GRE) (two fast field echo, one FLASH) T1-weighted T2\* images were obtained in three cases (two in 0.5-T, one in 1.5-T studies). In four cases, postcontrast studies (Gd-DTPA, 2 mL/kg), were also obtained. The spine was examined in six patients with sagittal SE T1- and T2-weighted images (Table 2).

Complete cerebral angiographic studies of both internal and external carotid arteries and of the vertebral arteries were performed in all the patients; myelography was performed in four (Table 2).

## Results

The radiologic findings are summarized in Table 2.

TABLE 1: Clinical data in patients with superficial siderosis of the CNS

Patient/Sex/Age	Previous History	Hearing Loss (Duration)	Other Symptoms	RBCs/mL	CSF Examination Protein/100 mL	Time Interval MR-CSF Examination
1/M/45	Alcohol abuse	2 yr	Ataxia dysarthria	Bloody	121 mg	CSF 5 days after MR
2/M/42	Head trauma at age 12	10 yr	Dysarthria ataxia tremor	180 RBCs 300 RBCs 350 RBCs	91 mg 87 mg 89 mg	CSF 18 days, 1 and 2 months after MR
3/M/52	Cervical trauma at age 21; repeated episodes of 7th nerve palsy	3 yr	Headache ataxia dysarthria dysphagia paraparesis	700 RBCs	87 mg	CSF 10 months after brain MR, 20 days after spine MR
4/M/49	Not significant	9 yr	Ataxia paraparesis	430 RBCs	68 mg	CSF 21 days after MR
5/M/47	Cervical trauma with root avulsion at age 22	8 yr	Anosmia hypotonia ataxia	Bloody	55 mg	CSF 1 month after MR
6/F/45	Not significant	3 yr	Ataxia hemihypoesthesia 7th nerve palsy			
7/M/49	Cervical trauma with root avulsion at age 20	12 yr	Ataxia anosmia 7th nerve palsy urinary dysfunction paraparesis	Clear in 1983; Bloody, xanthochromic in 1991	43 mg 41 mg	CSF 7 years before MR CSF 2 months after MR

TABLE 2: Radiologic findings in patients with superficial siderosis of the CNS

Patient/Sex/Age	CT <sup>a</sup>	Field Strength/Type of Examination	MR										Myelography (Carotid and Vertebral)	Angiography (Carotid and Vertebral)
			Areas with Rim of Hypointensity on T2-Weighted Images			Other Findings				Spinal Cord Sac <sup>b</sup>	Dural Sac <sup>b</sup>	Other Findings		
			Cerebral Hemisphere	Interhemispheric Sulci	Chiasm	Brain Stem	Cerebellum Vermis Hemisphere	Spinal Cord	Dural Sac					
1/M/45	Atrophy of upper vermis	0.5 T/ head-spine	+	++	++	++	+++	++	++	+	-	Increased s.i. and destructive changes of superior vermis folia	Not performed	Negative
2/M/42	Atrophy of upper vermis	0.5 T/ head-spine head Gd-DTPA	+	++	+	+	++	++	+	-	-	Increased s.i. and destructive changes of superior vermis folia	Not performed	Negative
3/M/52	Negative	0.5 T/ head-spine	-	-	+	++	++	++	+	+	+	Increased s.i. of superior vermis folia	Anterior epidural cyst in cervicodorsal region	Negative
4/M/49	Atrophy of upper vermis, hyperdense rim around brain stem	0.5 T/ head-spine head Gd-DTPA spine Gd-DTPA	+	++	-	++	+++	++	++	+	++	Increased s.i. of superior vermis folia	Negative	Negative
5/M/47	Not performed	1.5 T/ head-spine head Gd-DTPA	+	++	++	++	++	++	++	++	-	Mild destructive changes of superior vermis folia	Right C8 root avulsion	Negative
6/F/45	Negative	1.5 T/ head	+/-	++	-	++	++	++	+	n.e.	n.e.	None	Not performed	Negative
7/M/42	Atrophy of upper vermis, hyperdense rim around mid-brain	1.5 T/ head-spine head Gd-DTPA	+/-	++	++	++	+++	++	++	++	++	Destructive changes of superior vermis folia	Left C8 root avulsion	Negative

Note. — +/- = doubtful; + = minimal; ++ = moderate; +++ = marked hypointensity in T2-weighted images; n.e. = not examined; s.i. = signal intensity.

<sup>a</sup> Pre- and postcontrast studies in all cases.

<sup>b</sup> The hypointensity was present only at the bottom of the dural sac.

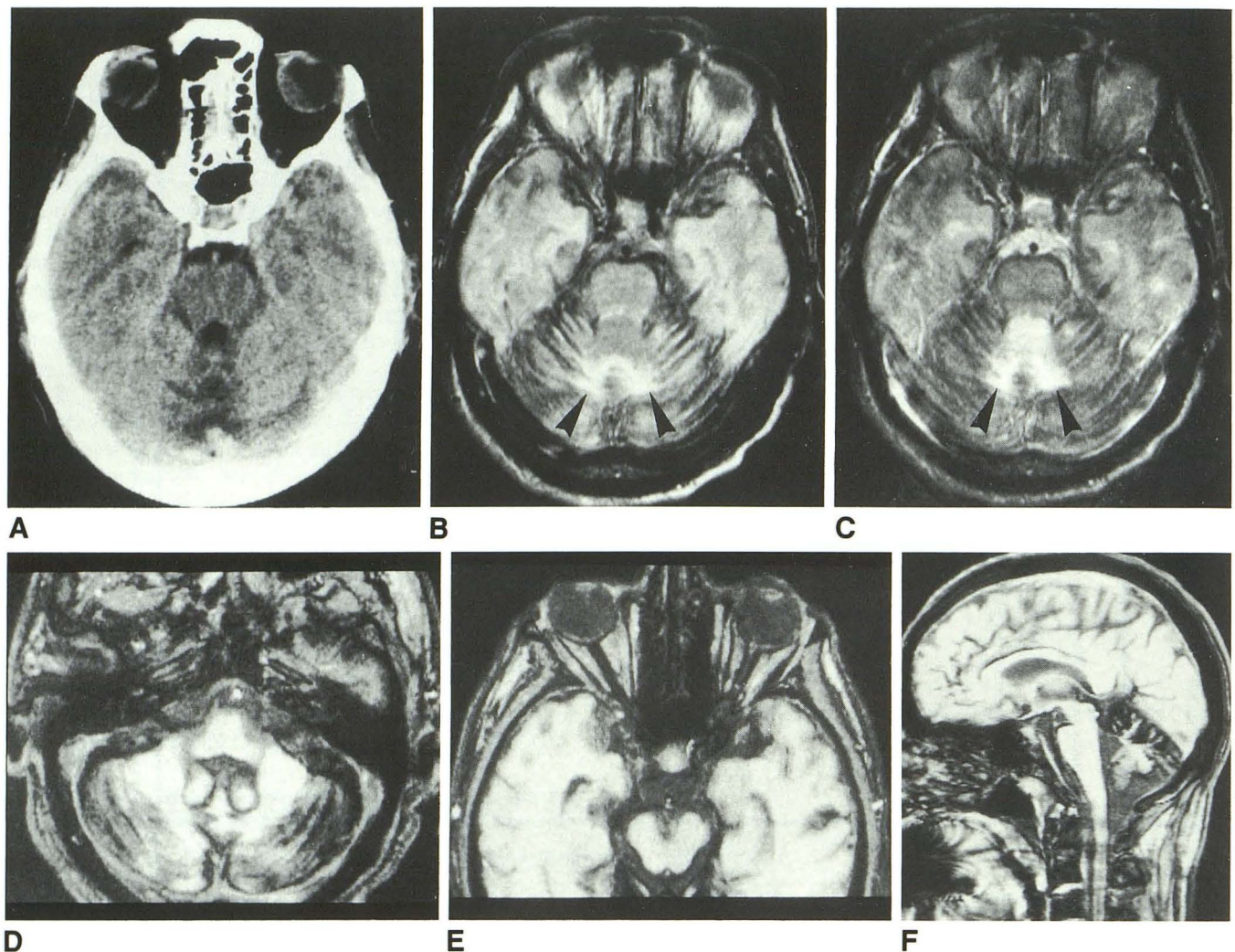


Fig. 1. Case 1.

A, Noncontrast CT scan shows superior vermian hypodensity consistent with severe atrophy.

B-F, MR (0.5 T).

B and C, (SE 2100/50-100) sections at the same level show a superficial rim of hypointensity and subjacent vermian and paravermian hyperintensity (*arrowheads*) perhaps due to gliosis and spongiotic changes.

D-F, (fast field echo 410/20/90°). Hypointensity due to paramagnetic effect of hemosiderin is visible on the right seventh-eighth cranial nerve complex (D) and on both optic nerves in the orbits (E). Despite this dramatic appearance, the patient's visual acuity was apparently normal; peripheral visual field was not tested. Sagittal section (F) shows partial loss of folia of the superior vermis and a conspicuous rim of hypointensity.

CT examinations showed atrophy of the cerebellum in five cases. In three cases (cases 1, 2, and 4), marked enlargement of the cisterns and lack of demonstration of superior vermian folia indicated a focal atrophy (Fig. 1A). In two cases (cases 4 and 7), a hyperdense rim around the brain stem was seen. In case 4 (Figs. 2A and 2B), CT study was performed prior to MR and suggested the possibility of SS, subsequently confirmed by MR (Figs. 2C and 2D); in case 7, CT was performed after MR; a prior CT performed in 1983 was reported as normal.

In all the cases, MR studies showed a rim of marked hypointensity in T2-weighted images around the brain stem, particularly at the level of the quadrigeminal plate, around the cerebellum, mainly at the superior vermis, and in the basal cisterns (Figs. 3A and 3B). Hypointensities along the cranial nerves were sometimes demonstrable (Figs. 1D, 1E, and 4). Atrophy of posterior fossa structures was also evident in all the patients, particularly at the level of the superior vermis of the cerebellum. In this area, complete loss of folia or destruction of the crowns of the folia were

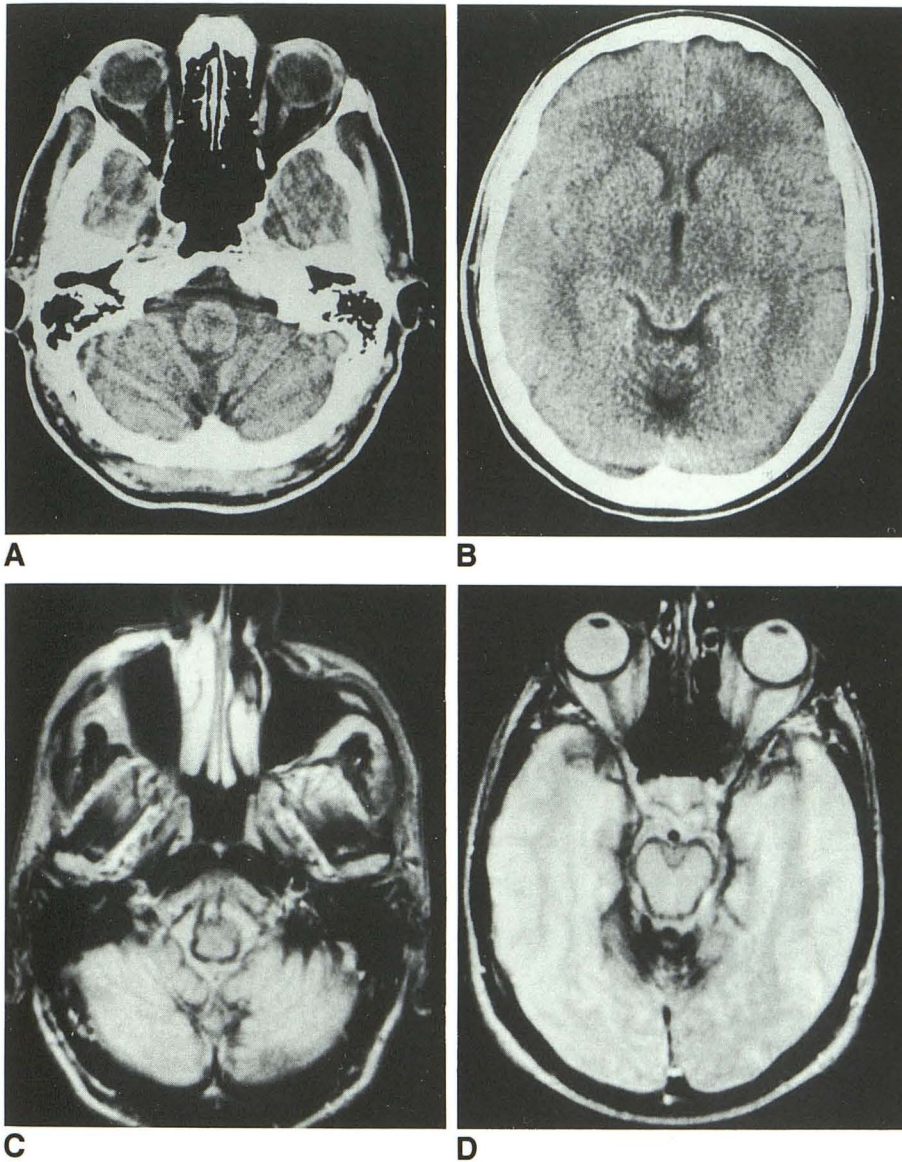


Fig. 2. Case 4.

A and B, plain CT shows high attenuation values around the medulla (A) and the midbrain (B), compatible with large amounts of hemosiderin encrusting the surface of the brain stem.

C and D, MR (0.5 T; SE 2000/70) at the same levels shows marked hypointensity compatible with hemosiderin deposits. Reprinted with permission from Daniele et al (9).

frequently seen in T1-weighted images (Figs. 1F and 5C-5E). In four cases, the subjacent cerebellar tissue also presented an increased signal intensity in intermediate and T2-weighted images (Figs. 1B, 1C, and 5E). In the supratentorial compartment, the rim of hypointensity was particularly marked in the sylvian and interhemispheric fissures, and in the chiasmatic region, while only a few sulci over the convexities were involved (Figs. 3C and 3D). No hypointensity in T2-weighted images was present on the wall of the ventricular system (Fig. 3C). In the patient with a history of head trauma (case 2), there were focal areas of increased signal intensity in T2WI in the frontal lobes, which were considered the result of cerebral contusion. This patient also had a small extradural leptomenigeal cyst in the posterior fossa (Fig. 5E). This same patient showed thick-

ening and enhancement of the dura on postcontrast study (Figs. 5A and 5B). The other three postcontrast examinations (cases, 3, 4, and 5) were normal. Atrophy in the supratentorial compartment was present in four cases.

MR of the spine (performed in six cases) demonstrated that the whole spinal cord was surrounded by a hypointense rim in T2-weighted images in five cases (Figs. 3E and 3F); in two of these cases, there was also hypointensity at the bottom of the dural sac.

The angiographic studies were negative for vascular malformations, aneurysms, or any other lesion in all the patients.

Myelography was performed in four patients and showed right C8 root avulsion with a small dural laceration in case 5 and a large pseudomeningocele at C6-C7 level on the right in case 7; in

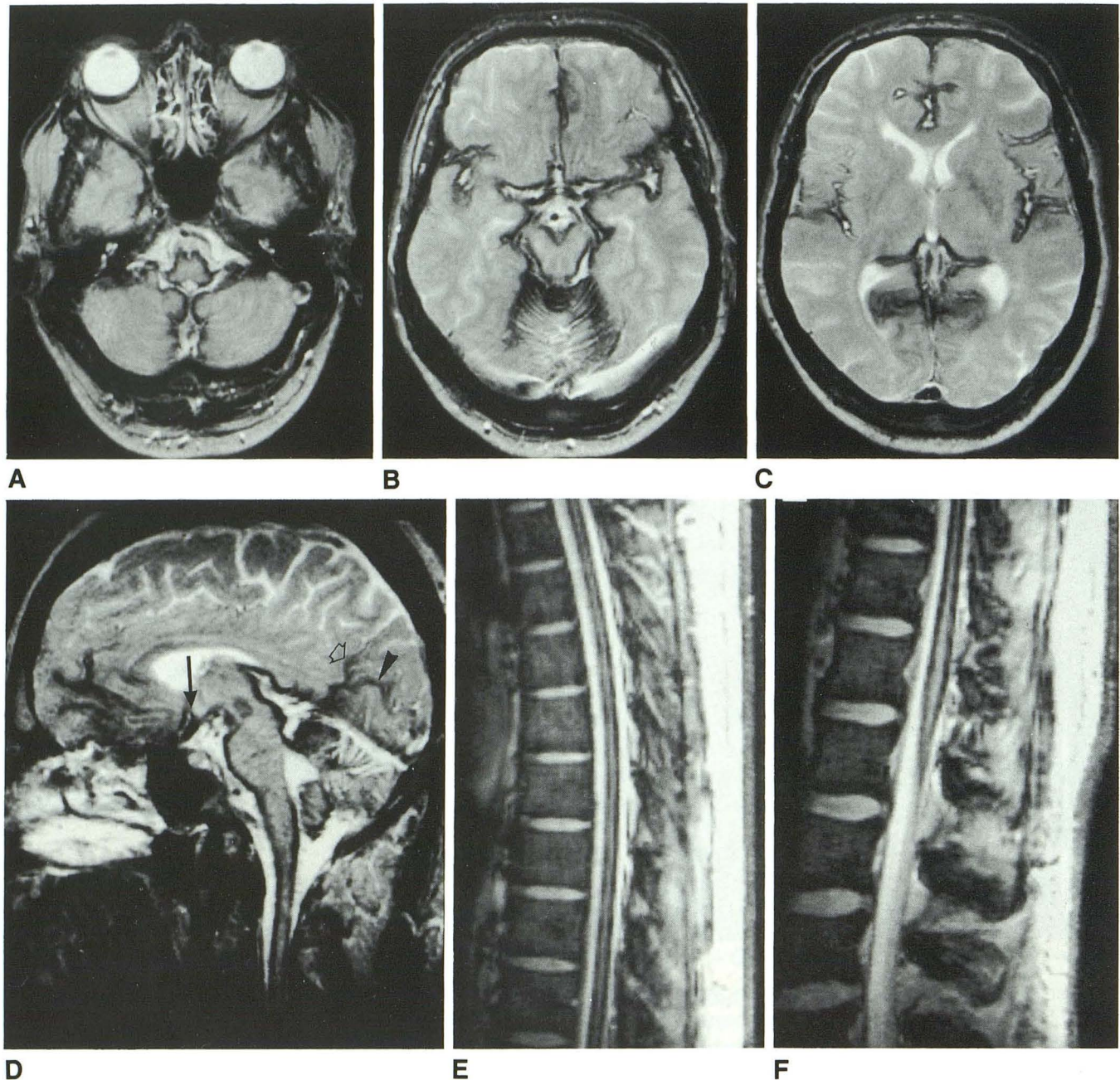


Fig. 3. Case 5.

A–D, MR of the head (1.5 T; SE 2400/90). A rim of hypointensity surrounds the medulla oblongata, the tonsils (A), the midbrain, superior cerebellum, optic tracts, and mesial temporal surface (B). The surface of the ventricles is normal (C), while marked deposits of hemosiderin are visible on the surface of the brain facing the sylvian, interhemispheric, and retropulvinar fissures (C). In sagittal section (D), hypointensity around the chiasm (*arrow*) and the cervical spinal cord is also evident. Hypointensity is present in frontal interhemispheric, calcarine (*arrowhead*), and parieto-occipital (*open arrow*) fissures.

E and F, MR of the spine (1.5 T; FLASH 400/18/15°) shows a rim of hypointensity around the spinal cord, down to the conus, while the roots of the cauda equina are normal.

case 3, there was leakage of contrast medium in a thin anterior epidural cyst extending in the cervical-upper dorsal region. Even this patient had a previous cervical trauma. In the fourth case (case 4), myelography was negative. No spinal vascular malformations were detected.

## Discussion

There are two major points of discussion. The first regards the diagnosis of this disease by MR; the second regards the disease itself, its course, and its possible pathogenetic mechanism.

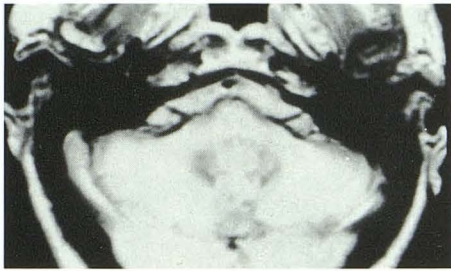


Fig. 4. Case 7. MR (1.5 T; SE 2000/30) shows hypointensity along the seventh–eighth cranial nerve complex. In pathologic reports, only the eighth nerve seems to be affected.

There is no question that MR will increase the number of diagnoses of siderosis of the CNS. The exquisite capability of MR to demonstrate the rim of hypointensity in T2-weighted images on the surface of the CNS (Fig. 3) because of the magnetic susceptibility effect of iron, makes this imaging modality the best diagnostic tool for siderosis. A limitation to diagnosis is the unawareness of this disease; in fact, a subtle rim of hypointensity may be easily overlooked.

Magnetic susceptibility effects are proportional to the square of magnetic field strength (10); therefore, a much more striking demonstration of SS is obtained at high field intensity. However, in our series, the amount of iron present on the surface of the brain was great enough to be easily detected also with 0.5-T magnets (Figs. 1, 2, and 5) and, in two cases, with CT (Figs. 2A and 2B). The preferential T2 proton relaxation enhancement caused by hemosiderin makes T2-weighted images the most demonstrative images for SS. Gradient-echo (GRE) (T2) sequences should be even more sensitive than SE (T2), but in our case (case 5), there was no noticeable difference. With T1-weighted GRE images at 0.5 T, we could also demonstrate, in one case, a rim of hypointensity not clearly visible in SE T1-weighted images (Figs. 5C-5E).

Regarding the second point of discussion, first we have to distinguish between milder degrees of siderosis that are occasionally seen in patients with previous single episodes of hemorrhage, and a chronic progressive disorder likely due to repeated hemorrhages of unknown origin or from oozing tumors or subdural or posthemispherectomy membranes.

Some degree of SS of the CNS is occasionally seen in patients with previous hemorrhages or operations. Two cases of neonatal subependymal and superficial hemosiderosis with striking MR findings, resulting from a single massive bleeding,

have been reported by Gomori et al (11). However, there is no evidence of long-term persistence of hemoglobin degradation products in the CSF in postnatal subependymal hemorrhage nor in other cases of single episodes of subarachnoid hemorrhage. Very likely, this kind of SS remains a self-limited condition without further clinical deterioration, except, perhaps, for the possible development of posthemorrhagic hydrocephalus.

Iwanowski and Olszewski (12) and Nöetzel and Ohlmeier (13) experimentally reproduced SS of the CNS only by repeated subarachnoid injections of blood or iron-containing pigments. In our patients, CSF was always bloody, with xanthochromia, and it was so in three different occasions in one patient. Therefore, repeated, even minimal, hemorrhages seem to be a necessary condition.

Prior to the introduction of MR, the reported cases of SS of the CNS were more frequently due to bleeding tumors (3). It is conceivable that the ability to detect tumors by modern neuroradiologic techniques, either CT or MR, has decreased the number of these cases. A bleeding or oozing tumor, either intraventricular or of the cauda equina, which may remain silent or cause very few signs, is now detected at a much earlier stage and chances that it may cause SS are greatly diminished: in the same period of time in which we collected the seven cases reported here, we observed only two cases of SS secondary to tumors, although minor degrees of siderosis associated with tumors may have been overlooked. MR now makes it possible to detect "idiopathic" cases of SS of the CNS, in which no obvious causes of bleeding may be found in spite of MR examination of brain and spine, cerebral angiography, and myelography. Therefore, the problem of the origin of hemorrhages remains unsolved.

SS is described as being a rather frequent complication of hemispherectomy, a neurosurgical procedure performed mainly for intractable epilepsy of childhood (6, 7). Neuropathologic studies have been performed in these cases, demonstrating the presence of posthemispherectomy membranes with fragile capillaries (7). These membranes are histologically indistinguishable from the membranes of a chronic subdural hematoma: they differ in that their edges are continuous with the brain tissue (7). The innermost layers of these membranes, especially near the edges, present multiple bleeding points (7). This is a kind of "bad reparation phenomenon" that we could theoretically expect also in other major neurosurgical procedures and in traumas with

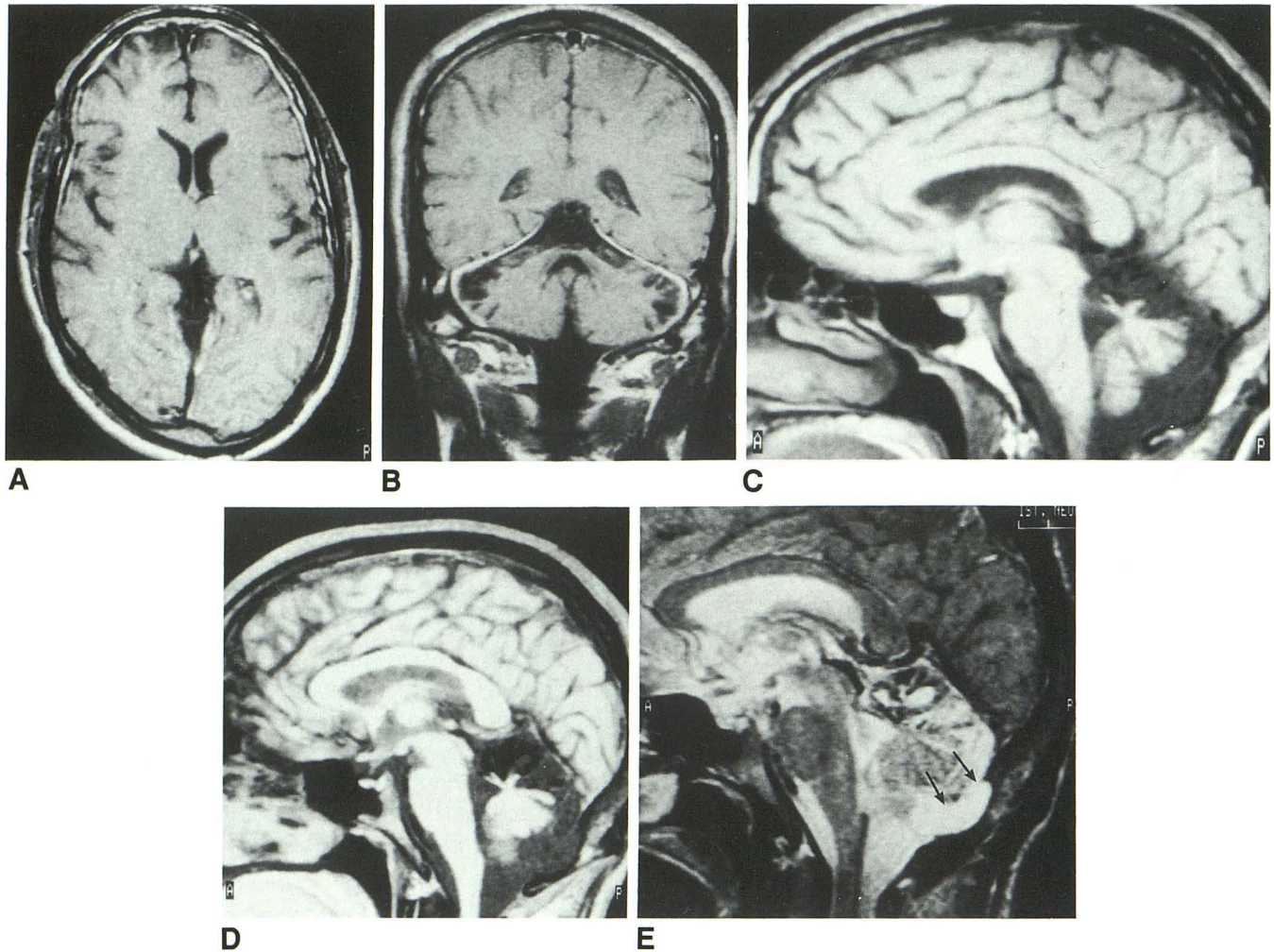


Fig. 5. Case 2. 42-year-old man with head trauma at age 12; MR (0.5 T).

A and B, Gadolinium-enhanced MR (See 680/30) shows a diffusely thickened and enhancing dura, particularly evident in the frontal areas (A), and on the tentorium (B).

C–E, Sagittal sections: C (SE 500/30), D (FFE 395/17/90°), E (SE 2100/100). SE (T1) image (C) only shows loss of folia of the superior vermis, while the GRE image (D) also shows a rim of hypointensity, which is better demonstrated in SE (T2) image (E). The arrows in E indicate an extradural posterior cyst, better demonstrated in more lateral sections.

meningeal lacerations. The fact that there is scant evidence of cases of SS in the literature could be due to the rarity of this phenomenon, but could also be related to the difficulties of diagnosing SS, because of the long latency of the symptoms. In our series, we do not have postsurgical cases. The only possible association we have found in our patients is with previous trauma: we have two cases of cervical root avulsion, one case with cervical trauma with epidural cyst, and one case with previous frontal contusions and posterior fossa arachnoid cyst. In all these cases, the trauma was remote; however, we know from the literature that, no matter what the cause of the chronic bleeding, a long delay is necessary before the symptoms become evident (3). The presence,

at least in three of our cases, of documented disruption of meningeal layers (root avulsions and traumatic epidural cyst), suggests that irregular scarring processes could be the source of repeated bleedings, similar to those demonstrated in posthemispherectomy patients. The association of SS with previous spinal cord injury has also been recently reported by Nudelman J, Seltzer S, Cohen WA (paper presented at the annual meeting of the American Society of Neuroradiology, Washington, June 1991). The thickening of the dura with marked post-Gd-DTPA enhancement, observed in one posttraumatic case (case 2, Fig. 5), could just represent fibrosis of the meninges, due to repeated hemorrhages



(14), rather than the expression of abnormalities prone to bleed easily.

We do not have any proof that posttraumatic changes may be responsible for repeated hemorrhages; however, patients with "idiopathic" SS should be carefully questioned for previous, even old, traumas, and patients with previous traumas who develop deafness or ataxia, even many years after the injury, should be examined by MR.

However, both in the literature and in our series, there are idiopathic cases with no previous history of trauma. Among the different theories proposed to explain the idiopathic cases, increased permeability of meningeal arteries with passage of RBCs into the CSF (15) seems to be an acceptable hypothesis. Another possibility is repeated bleeding from angiographically undetectable superficial microscopic vascular malformations. Reduction of normal phagocytosis of RBCs due to leptomeningeal fibrosis (16) or lack of iron-binding protein in the CSF (15) seems to us to be groundless; moreover, these hypotheses do not take into account that in most of the cases, as in our series, there is indeed blood and xanthochromia in the CSF, indicating minor repeated hemorrhages.

The final point to consider is the pathogenetic mechanism of the destructive changes of the brain and nerves in SS. This problem has been addressed by Koeppen and Detinger with histochemical studies (5). There is a probable active mechanism of phagocytosis of the blood by-products in the CSF from the glial cells on the surface of the CNS. The microglia is able to biosynthesize, within the hemosiderin matrix, ferritin, which is toxic to the cells themselves. The areas that are more severely affected are the superior vermis, the surface of the brain stem, particularly the quadrigeminal plate, the brain tissue surrounding the sylvian cisterns, and the first, second, and eighth cranial nerves. The selective involvement of these nerves is explained by their structure. The fibers of the first and second cranial nerve are enveloped by *central* myelin and glia for the whole extent of the nerve in the cisterns. All the other cranial nerves, except the eighth, present the transition point from *central* myelin to *peripheral* myelin very close to the brain stem. The eighth cranial nerve presents this transition point near the internal acoustic meatus rather than near the brain stem. Therefore, the eighth cranial nerve has a long cisternal tract with the *central* structure, and in this tract it is separated from the subarachnoid space only by a

continuation of the thin and delicate pia mater (2). Koeppen and Detinger using a staining technique with glial fibrillary acid protein demonstrated that in the cranial nerves the extent of siderosis corresponds exactly to the extent of *central* myelin and glial cells, ending precisely where Schwann cell covering begins (5). The distribution of SS of the CNS corresponds to the main distribution of blood observed in diffuse subarachnoid hemorrhages, and to the distribution of radioisotope demonstrated in cisternographic studies. That the distribution of SS is simply related to the amount of blood present in the cisterns is supported by the following observation. In one case reported in the literature (6), in which the bleeding originated in the ventricles, the inferior vermis, which was conceivably more "bathed" than the superior vermis by the blood coming out of the fourth ventricle, exhibited a greater destruction of its folia than the superior vermis. The severity of involvement of the cerebellum always seems more marked than that of the cerebral hemispheres; it is possible that the cerebellar glia may be particularly vulnerable to the blood by-products compared to the cerebral glia because of its specific ability to synthesize ferritin (5). When we observed the destructive changes of the crowns of the folia of the superior vermis in the first of our cases (Fig. 1), we were convinced that this was the result of bleeding from a vascular malformation that had "burned" itself and caused local destruction of cerebellar tissue. The absence of clinical episodes of subarachnoid hemorrhage, the absence of angiographic demonstration of vascular malformation, the constance of this finding, also observed in the other MR series (17) as well as in pathologic specimens (2, 6, 15, 16), leads to the conclusion that the severe destructive changes of the superior vermis do not reflect the focal cause of SS but are rather the result of greater vulnerability of the superior vermis and of pooling of blood in the surrounding cisterns.

The clinical symptoms and signs and the evolution of SS of the CNS are quite uniform and reflect the distribution of iron deposits and the related destructive changes. The main clinical signs are sensorineural hearing loss and ataxia. Myelopathy may follow in advanced cases. Dementia is an inconstant feature, probably related, in most of the reported cases, to the concomitant hydrocephalus. In fact, improvement of the mental status has been reported in cases of SS after shunt (2, 14), but dementia may be associated

with severe siderosis of the temporal lobes (8). Loss of sense of smell has been rarely reported, probably because it does not disturb the patient, but probably it is often present. It is surprising that visual problems are seldom mentioned in spite of the marked involvement of the chiasm, optic nerves and optic tracts by iron deposits. An explanation may be found in the central location in the optic nerves of the macular fibers, which may therefore remain uninvolved, thus maintaining a normal visual acuity.

The slow progression of the characteristic clinical features, ie, deafness and ataxia, simulating a degenerative disease, should alert the neurologist, who is, however, rarely aware of this disease. Many times, therefore, SS of the CNS will remain a neuroradiologic diagnosis made on an MR study. It is to be hoped that the collection of other cases will help to clarify the major problem of SS: where the bleeding comes from.

A tentative treatment with iron chelating agents has been mentioned in the literature (5) and has been instituted in two of our cases, but it is too early to predict whether it will stop or slow down the course of the disease.

### Acknowledgments

We thank C. A. De Fanti, MD, and D. Testa, MD, for allowing to report cases under their care.

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