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Superficial siderosis of the central nervous system: report of three cases and review of the literature

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Key words: Superficial siderosis; Central nervous system; Magnetic resonance imaging; Cerebrospinal fluid

Summary

We present 3 cases and a review of the literature to demonstrate the current state of clinical diagnosis and therapy of superficial siderosis of the central nervous system. Typical symptoms were progressive cerebellar ataxia, spasticity and hearing loss. Repeated subarachnoid hemorrhage was indicated by persistent xanthochromia of the cerebrospinal fluid and confirmed by the presence of erythrophages, siderophages and iron-containing pigments. Deposition of free iron and hemosiderin in pial and subpial structures leads to intoxication of the central nervous system and represents the pathophysiological mechanism of superficial siderosis. Hypointensity of the marginal zones of the central nervous system on T2 weighted MR images indicates an iron-induced susceptibility effect and seems pathognomonic for superficial siderosis. In 39 of the 43 previously described cases superficial siderosis was verified by biopsy or autopsy. To day magnetic resonance imaging enables diagnosis at an early stage of the disease. Therapeutic management requires the elimination of any potential source of bleeding. In patients with unknown etiology no proofed therapy is yet available.

Introduction

Superficial siderosis (SS) of the central nervous system (CNS) is an uncommon, clinically often underdiagnosed disorder. The responsible pathogenetic mechanism comprises intoxication of neuronal structures by free iron and hemoglobin breakdown products (Jackson 1949; Iwanowski and Olszewski 1960; Koeppen and Borke 1991). Siderosis is most likely caused by recurrent or chronic subarachnoid hemorrhage of various etiologies (Table 1).

Although in advanced cases clinical manifestations are quite characteristic, in vivo diagnosis has been made in only 6 of the 43 reported cases (Pinkston et al. 1983; Gomori et al. 1985; Katsuragi et al. 1988; Koeppen and Dentinger 1988; Zwarts et al. 1988). Persistent xanthochromia of the cerebrospinal fluid (CSF) is a typical, but not specific finding and biopsy or autopsy have been necessary to ensure the diagnosis so far. Nowadays characteristic signal abnormalities on magnetic resonance imaging (MRI) yield early evidence of SS of the CNS (Gomori et al. 1985). We report 3 cases with emphasis on clinical, MRI and CSF findings.

Patients and methods

Between 1989 and 1991 SS of the CNS was diagnosed in 3 male patients aged 58, 59, and 77 years at our hospital. All of them underwent repeated clinical and neurological examinations. The following laboratory tests were performed: erythrocyte sedimentation rate, complete blood count, blood coagulation and platelet function tests, serum electrolytes, liver function tests, cholesterol, triglyceride, thyroid function tests, serum electrophoresis and immunoglobulins, autoantibodies, immune complex screen and lues serology. CSF analysis from repeated lumbar punctures included cell count, total protein, CSF glucose, ferritin, transferrin, free iron, and bilirubin; cells were examined cytologically (Table 2).

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TABLE 1

REPORTED CNS LESIONS CAUSING SUPERFICIAL SIDEROSIS (INCLUDING THE PRESENT 3 CASES)

Lesion	Authors
Intracranial neoplasm	
Glioblastoma	Braham and Wolman (1965)
Ependymoma	McGee et al. (1962); Tomlinson and Walton (1964); Kott et al. (1966); Gomori et al. (1985); Koeppen and Dentinger (1988); Willeit et al. (1992)
Oligodendroglioma	Rosenthal (1958); Koeppen and Dentinger (1988)
Pinealoma	Dastur and Sinh (1962)
Meningeosis carcinoma	Noetzel (1940)
Angioma/aneurysm	Cammermeyer (1947); McGee et al. (1962); Foncin et al. (1967); Hughes and Oppenheimer (1969) (2 cases); Pinkston et al. (1983); Koeppen and Dentinger (1988)
Intracerebral hematoma	Neumann (1948); Rosenthal (1958) (2 cases); Jacob and Goachet (1959)
Hemispherectomy	Ulrich et al. (1965); Hughes and Oppenheimer (1969) (4 cases);
Subdural hematoma	Noetzel (1955); Hughes and Oppenheimer (1969); Koeppen and Dentinger (1988)
Meningitis hemorrhagica	Noetzel (1940);
Cerebral trauma	Jacob and Goachet (1959); Hughes and Oppenheimer (1969)
Unknown etiology	Lewey and Govons (1942); Neumann (1956); Garcin and Lapresle (1957); Rosenthal (1958); Jänisch and Weiss (1964); Braham and Wolman (1965); Castaigne et al. (1967); Hughes and Oppenheimer (1969); Koeppen and Barron (1971); Katsuragi et al. (1988); Zwarts et al. (1988); Willeit et al. (1992) (2 cases)

Iron was measured on a centrifugal analyser using ferrozine as chromogen, Ferritin was determined by radioimmunometric assay. Transferrin measurement was done by nephelometry and bilirubin measurement was performed colorimetrically.

Non-enhanced and enhanced cerebral CT scans and digital-subtracted 4-vessel angiography were done in all cases. Spinal angiography was performed in patients 1 and 2. MR-scans of the brain and spinal cord were performed at a 1.5 Tesla system and included T2 (TR = 2400 ms, TE = 90 ms) and T1 (TR = 500 ms, TE = 15 ms) weighted images. T1 weighted images were obtained before and after administration of Gd-DTPA (0.1 mmol/kg/BW).

Results

Clinical presentation

(1) Patient T.J., age 58 years, complained about unsteady gait, dizziness and lower extremity weakness since 1986. Subsequently he developed hearing loss, lack of concentration and a mild forgetfulness with difficulty to recall recent events. He never complained

TABLE 2

CSF FINDINGS IN 3 PATIENTS WITH SUPERFICIAL SIDEROSIS OF CNS

	Patient 1	Patient 2	Patient 3	Normal values
	1/1989 10/1990	3/1991 4/1991	1/1991 5/1991	
Protein (mg/dl)	160 163	125 115	640 62	< 50
Bilirubin (mg/dl)	0.9 0.7	0.5 0.6	1.4 1.5	Not present
Iron (nmol/l)	3.4 3.0	2.6 3.0	15 14	1–3
Ferritin (ng/ml)	98 85	102 105	242 156	10–80
Transferrin (mg/dl)	< 10 < 10	< 10 < 10	41 12	Not present
Cells/mm^3	18 40	45 24	20 28	< 5
Cytology	Monocytes with hemosiderin	Monocytes with hemosiderin	Monocytes with hemosiderin	

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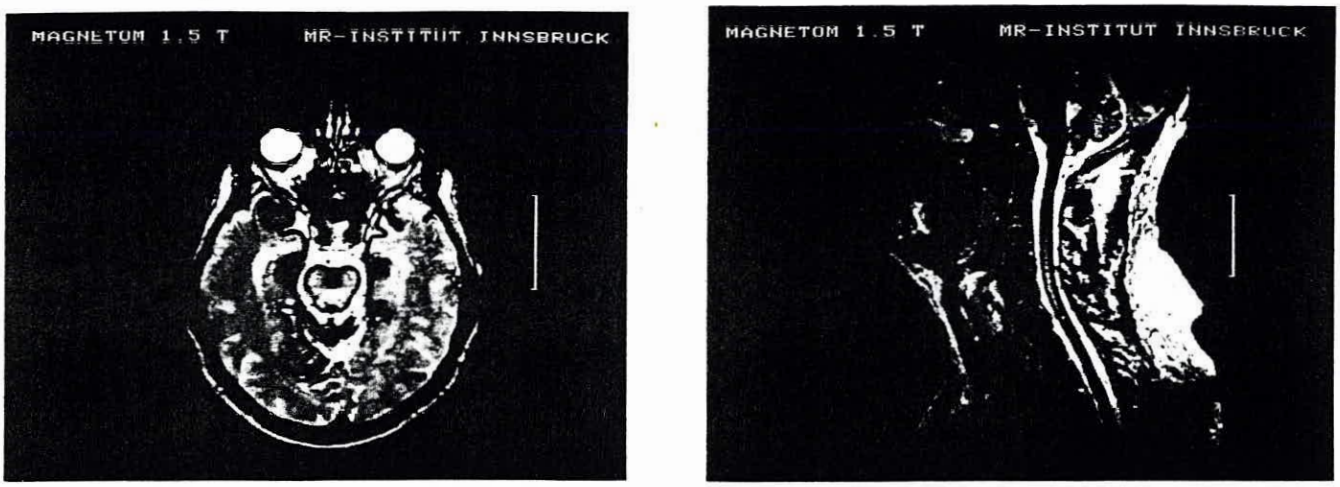

Fig. 1. T2 weighted MR images of the brain and cervical spinal cord in a 58-year-old patient suffering from superficial siderosis of the CNS. The axial image (A) reveals a signal void of the temporal lobe surface, the brain stem and vermis cerebelli. The sagittal image (B) of the cervical spine confirms the abnormal T2 shortening over the vermis cerebelli, brain stem and cervical spinal cord.

of headache. Clinical controls showed marked progression of the disease.

(2) Patient H.H., age 77 years, started to develop unsteady gait, vertigo and hearing loss in 1989. Six months later he suffered from generalized weakness and recent memory deficiency. The symptoms gradually progressed and on admission in 1991 he was unable to walk anymore.

(3) Patient Z.J., age 59 years, complained of dizziness, unsteady gait, and hearing loss since 1989. One year later he became unable to feel the passage of urine on micturition and had occasional loss of bowel control. In addition there were erection and ejaculation failures. He never had had headache or lower back pain.

The *clinical examination* of all 3 patients revealed cerebellar symptoms, spasticity of the lower extremities, hearing loss. Patients 1 and 2 showed signs of mild dementia with impairment of short-term memory and difficulty in defining words and concepts. In patient 2 marked atrophy of the distal skeletal muscles and decreased proprioceptive muscle reflexes could be found as well. Patient 3 had additional hypesthesia related to the sacral dermatomas S1–S4 and bladder and bowel dysfunction.

Blood chemistry findings of all 3 patients were essentially normal. In none of them there was evidence of hemorrhagic diathesis or vasculitis.

CSF findings are presented in Table 2. All patients showed xanthochromic CSF and cytological examina-

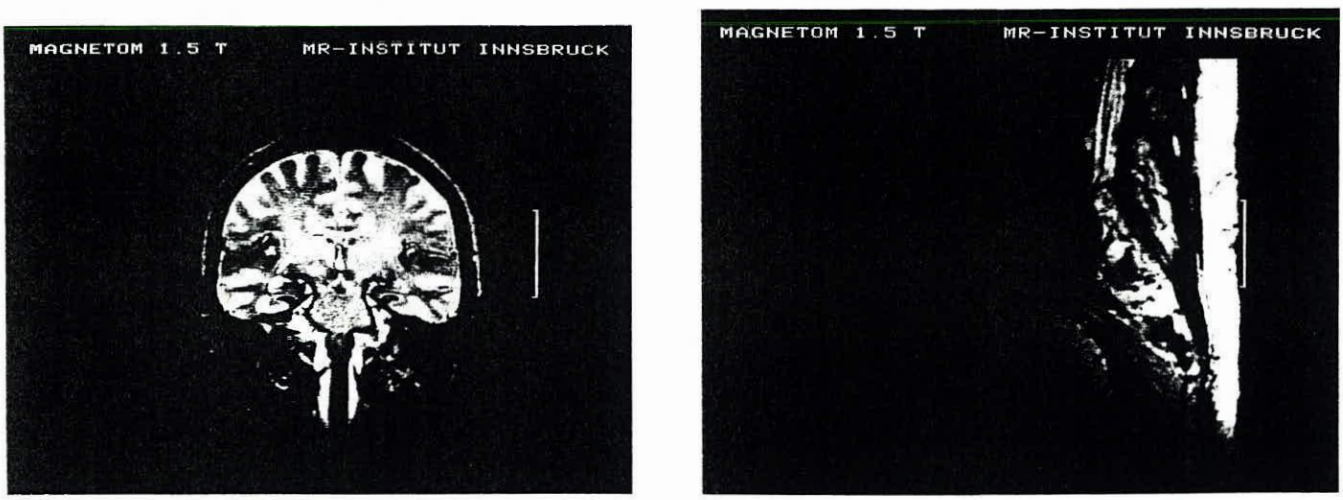

Fig. 2. T2 weighted MR images of the brain (A) and lumbosacral spine (B) in a 59-year-old patient with a symptomatic form of superficial siderosis of the CNS. The coronal screen (A) through the temporal lobes shows signal loss of the cortical surface in the insular region, the mesiobasal temporal lobes, the brain stem and spinal cord. The sagittal image (B) of the lumbosacral spine revealed a tumor extending from L5 to S5 with widening of the spinal canal and signs of previous hemorrhage (arrow).

tion revealed erythrophages and siderophages. Iron concentration was abnormally increased and bilirubin was present. Two months after surgical removal of the ependymoma in patient 3 the concentration of iron (2.4 nmol/l) and ferritin (40 ng/l) was considerably lowered and bilirubin and siderophages were no longer detectable.

Neuroradiological findings

Pre- and postcontrast CT of the brain were normal in all 3 patients. Four-vessel angiography excluded cerebral vascular malformations in all of them and segmental spinal angiography was normal in patients 1 and 2.

In all patients T2 weighted MR images revealed characteristic findings. The surface of the brain and spinal cord showed hypointensity, which was prominent in the basal temporal lobes and the insular regions (Figs. 1A, 2A), the brain stem and cerebellum (Figs. 1A and B), especially in the vermis, and in the entire spinal cord (Figs. 1B, 2A). There was no Gd-DTPA enhancement and T1 weighted images were normal as well. In patient 3 MRI revealed a tumor of the filum terminale (Fig. 2B), histologically proven to be an ependymoma.

TABLE 3

CLINICAL FINDINGS IN 13 CASES OF SS WITH UNDETECTED BLEEDING SOURCE

Clinical characteristics	$n = 13$ (present series: $n = 2$ literature: $n = 11$)
Sex	
male	10
female	3
Age at onset (yr)	5–73
Symptoms	
cerebellar ataxia	13
nerve deafness	12
spasticity	12
dementia	10
convulsions	3
radiculopathy	3
headache	0
CSF ($n = 9$)	
xanthochromatoous	7
CT ($n = 3$)	
normal 3	
MRI ($n = 3$)	
T2 shortening	2
cerebellar atrophy	2
Died	9 (in average 11 years after onset)

Discussion

Superficial siderosis of the CNS is caused by repeated episodes of subarachnoid hemorrhage and can be reproduced in animal models by injection of blood or iron-containing substances into the subarachnoid space (Jackson 1949; Iwanowski and Olszewski 1960). In 32 of the 43 reported cases the responsible lesion of siderosis was found (Table 1). In the remaining patients, in whom no source of hemorrhage could be disclosed, increased permeability of meningeal vessels may produce recurrent bleeding (Hughes and Oppenheimer 1969; Katsuragi et al. 1988).

Progressive cerebellar ataxia, hearing loss and spasticity are the characteristic symptoms of SS (Table 3). Other neurologic deficits such as convulsions and radiculopathy may occur, dementia commonly develops in the later stage of the disease. In contrast to acute subarachnoid hemorrhage, headache usually is not present. The identified underlying pathology contributes to the clinical presentation and determines long-term outcome. The prognosis of patients without detectable bleeding source is poor: 9 of 13 patients died after a mean period of 11 years (Table 3).

Until now SS has rarely been diagnosed in vivo. In 37 of the 43 reported cases diagnosis was made neuropathologically. In 2 patients SS was proven by repeated CSF examinations in connection with clinical findings (Koeppen and Dentinger 1988; Zwarts et al. 1988). Two other cases were confirmed at biopsy (Pinkston et al. 1983; Katsuragi et al. 1988). In 1 case the diagnosis was strengthened by CT scanning (Koeppen and Dentinger 1988), in another case it was confirmed by MRI (Gomori et al. 1985).

In our patients diagnosis of SS was based on CSF and MR findings. Repeated CSF-examinations revealed xanthochromia and erythrophages and siderophages indicating recurrent or persistent extravasation of blood into the subarachnoid space (Table 2). In patient 3, xanthochromia may have been additionally produced by the high protein content of CSF (Table 2).

Conventional neuroradiological examinations were normal, whereas T2 weighted MR images revealed hypointensive superficial zones of the brain and spinal cord (Figs. 1 and 2). The T2 shortening of the MR signal is a result of susceptibility effects caused by iron and hemosiderin deposition in the marginal zones of the CNS (Gomori et al. 1985). The neuropathologic evidence of rusty-brown superficial staining, particularly of the temporal lobe, brain stem, cerebellum, and spinal cord correlates with areas of signal void on MRI. A similar iron-induced susceptibility effect leads to a decreased signal in the rim of chronic intracerebral hematomas (Thulborn et al. 1990). In accordance with a recent report on recurrent hemorrhage from an

ependymoma, the effect of T2 shortening is more prominent at higher magnetic field strength (Gomori et al. 1985). This may explain the normal MRI findings in an other reported case (Zwarts et al. 1988). MRI offers high sensitivity and seems specific for parenchymal iron deposition. Therefore, diagnosis of SS can be established by typical MRI and CSF findings even without biopsy.

The toxicity of free iron and iron-containing pigments on neuronal structures has been known for years (Jackson 1949; Iwanowski and Olszewski 1960). The pathogenesis of SS has recently been eluciated by Koeppen and Borke (1991). As hemoglobin delivers relatively little iron to the cerebral surface, symptoms of SS develop exclusively in the case of recurrent or chronic subarachnoid hemorrhages. Persistent hemorrhagic CSF stimulates microglia cells in the subpial parenchyma inducing an accelerated ferritin biosynthesis. Iron-containing heme pigments and free iron molecules are taken up from the CSF by Bergmann glia and are stored in macrophages and glial cells. The first iron storage takes place in the form of iron-ferritin and this is later transformed to hemosiderin, which causes the rusty-brown staining of the brain surface. When the iron-binding capacity is exceeded, free ionic iron induces lipid peroxidation leading to parenchymal damage and neuronal cell death with secondary gliosis. Immunohistochemical findings reveal large numbers of socalled 'ovoid bodies' in the astroglial cells, which corresponds to neuroaxonal dystrophy (Katsuragi et al. 1988).

Not only the location of bleeding, as suggested by Hughes and Oppenheimer (1969), is responsible for the different involvement of CNS structures in SS, but also local tissue factors are of great importance. The selective vulnerability of the cerebellar cortex can be explained by the high vascularity of this region and by the fact, that Bergmann glia come into direct contact with CSF stimulating the uptake of iron and the biosynthesis of ferritin. Purkinje cells are known to be highly sensitive to toxic agents.

Therapy of SS requires elimination of the source of bleeding. In patient 3 the tumor of the cauda equina was surgically removed. Two months later lumbar puncture showed clear, colorless CSF, both siderophages and erythrophages were no longer detetable.

There is no specific treatment for those cases of SS, in which the lesion for recurrent hemorrhages could not be defined. The use of antioxidants and chelating substances as well as ventriculo-atrial shunting are discussed, their efficacy remains to be determined (Koeppen and Borke 1991).

MRI is helpful not only in the early diagnosis of SS but may also represent a potential tool to monitor treatment effects.

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