

haustable ankle clonus and extensor plantar reflex, all on the right side. After a 15 minutes rest, upper-motor neuron signs and symptoms had disappeared. Routine laboratory tests including syphilis serology and complement studies were normal or negative. ANA and rheumatoid factor were negative. The CSF showed high IgG and one extra oligoclonal band at iso-electric focusing. Myelography, spinal angiography and cranial CT scan were normal. MRI showed multiple hemispherical white matter lesions, suggesting multifocal demyelination. Visual-evoked-responses (VER) and somatosensory-evoked-responses (SSER) showed increased latencies.

Patient 2: Six years before admission, this 58 year old book-keeper repeatedly noticed brief jerks of his right leg after walking for 10 minutes, which disappeared after resting for 5 minutes. One and a half years later he was troubled by a heavy feeling in the right arm with numbness of the first three fingers, on using it extensively. These symptoms disappeared after 30 minutes rest. Four years after onset he noticed weakness of the legs on walking for 5 minutes, which disappeared during rest. These symptoms forced him to work for half days only. Six months later, walking felt stiff and awkward with pain and tingling of the legs and an imperative micturation developed. When lying in bed he noticed jerking movements of the legs. Six years after onset of the symptoms he was admitted to our hospital. Neurological examination revealed no abnormalities except for absent abdominal reflexes. His gait was normal initially, but became spastic after walking for 1 hour and a brisk left biceps jerk, exaggerated ankle jerks and bilateral extensor plantar reflexes appeared. After a 10 minute rest upper-motor neuron signs and symptoms disappeared. Routine laboratory tests were normal. CSF examination revealed high IgG and multiple extra oligoclonal bands at iso-electric focusing. Myelography was normal. MRI revealed multiple paraventricular white matter lesions consistent with multifocal demyelination. VER showed prolongation of P100 on stimulation of the right eye, SSER showed absent cortical responses on bilateral tibial nerve stimulation and electro-oculography showed square wave jerks on eye closure.

The main features of both case histories are intermittent, exertion-induced, upper-motor neuron symptoms and signs, disappearing after rest. During rest or mild exercise both patients were subjectively and objectively asymptomatic. Normal "vasculitis tests", angiography, myelography and MRI of the spinal cord made vascular

lesions or compression of the spinal cord most unlikely. Based on the results of CSF studies, multimodality evoked potentials tests and MRI, in both cases a diagnosis of "laboratory-supported-definite multiple sclerosis" was made, according to the criteria of Poser *et al.*⁷ In the literature on neurogenic intermittent claudication, multiple sclerosis is not generally cited as a cause for IPC.^{1,2} On the other hand, textbooks on multiple sclerosis mention IPC as "a frequent early symptom of multiple sclerosis", however, without referring to well documented observations.^{4,5} Descriptions of signs and symptoms are variable and sometimes vague.^{4,5,8,9} The period during which upper-motor neuron signs and symptoms really are intermittent (that is, they completely disappear after rest) appears to be short.^{4,9} Only three short case histories could be found in the literature.^{6,10} McAlpine⁶ described two patients who, as early feature of multiple sclerosis, showed IPC for several weeks. In both cases this disappeared for about a year and then recurred as a prelude to a progressive course of the disease. Godlewski¹⁰ mentioned a multiple sclerosis patient who had IPC for 4 months, before permanent spastic paraplegia developed. Multiple sclerosis presenting with slowly progressive IPC as isolated symptom for nearly 6 years has, to the best of our knowledge, not been described before. The pathophysiological mechanism of IPC in multiple sclerosis is believed to be related to diminished motor conduction in demyelinated axons, resulting from even a slight rise of body-temperature caused by exercise.^{4,5,11} A temperature increase of only 0.5°C appears to be sufficient to produce reversible conduction-block in demyelinated fibres.¹² Indeed in our first patient, walking for 30 minutes increased tympanic temperature by 0.5°C. Obviously, comparison of pre- and post-exertion corticospinal tract conduction time¹³ in patients with IPC, may give better insight in the pathophysiological mechanism.

IPC is generally attributed to transient spinal ischaemia.¹ However, actual evidence for such causal relationship is sometimes poor.^{3,14} Conduction or angiographic evidence are rarely found. In retrospect, some of these cases might in fact have been caused by multiple sclerosis.

In conclusion, these two case histories illustrate that IPC may be a solitary manifestation of multiple sclerosis for protracted periods. A simple exertion-provocation-test may be essential to reveal the neurological substrate of the complaints and to establish the diagnosis.

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Unexplained chronic subarachnoid bleeding and a slowly progressive neurological syndrome

Sir: Superficial haemosiderosis of the central nervous system (CNS) (subpial cerebral

siderosis) is the result of chronic and repeated leakage of blood into the subarachnoid space. The syndrome is unique in that the symptoms are mainly caused by the deposition of iron-containing pigments (haemosiderin) in the marginal zones of the CNS¹ resulting in gliosis and neuronal loss. The cardinal feature is the finding of xanthochromic cerebrospinal fluid (CSF) on repeated lumbar punctures. The clinical picture is quite varying, but one or more of the following signs are usually present: cerebellar ataxia, nerve deafness, dementia and spasticity.² These signs do not indicate the site and cause of the bleeding. Two patients (adults) have been reported who were diagnosed during life;^{3,4} we report one young patient with a remarkable benign course over 4 years.

A female child was admitted to hospital at the age of 5 years because of abdominal pain, vomiting and failure to thrive. After thorough investigations no cause was found for the abdominal complaints. An EEG unexpectedly showed generalised bilateral synchronous spike and waves. A lumbar puncture was then performed which showed xanthochromic CSF with a raised protein content of 2 g/l (see table). One day later she complained of headache, consciousness became impaired, and signs of meningism developed. A CT scan was normal.

On transfer to our hospital, examination revealed a slightly stuporous child with signs of meningism, without lateralising neurological signs and with normal fundi. In the course of the next days she recovered spontaneously. Repeated EEG showed bilateral synchronous epileptic discharges of short duration. Four-vessel angiography revealed no abnormalities. Repeated lumbar punctures disclosed persistent xanthochromia, elevated protein, methaemoglobin and bilirubin (see table). A month after admission slight cerebellar ataxia became evident. At discharge 2 months later the CSF abnormalities were still present. Six months later she developed absences. At the age of

6 years superficial haemosiderosis of the CNS was suspected. Neuropsychological examination showed a discrepancy between the verbal IQ (114) and the performance IQ (77) on the WISC-R scale. An audiological examination was normal. At the age of 8 years she was readmitted after two generalised convulsions and an increase of her cerebellar signs. A new finding at neurological examination were absent ankle jerks. The CSF was still xanthochromic. Myelography and MRI of the brain and spinal cord were normal. Neuropsychological examination showed no deterioration in comparison to the examination of 2 years before. She was discharged and 3 years later her clinical state is unaltered.

In this 9 year old child CSF xanthochromia has been present for 5 years with elevated protein, methaemoglobin and bilirubin, indicating continuous blood leakage in the subarachnoid space (table). Since other causes of the neurological signs have been excluded, the syndrome is very likely to be caused by superficial haemosiderosis of the CNS. Apart from an initial period after the first lumbar puncture, there have been no signs of meningism, so the bleeding has been chronic and slow.

The clinical picture consisted of bilateral cerebellar ataxia, seizures, a low performance IQ and absent ankle jerks. There has been no evidence of spasticity or nerve deafness. From the patients described by Tomlinson and Walton² it can be concluded that none of the clinical features is a sine qua non for superficial haemosiderosis of the CNS. Absent ankle or knee jerks have been described^{1,6,7} as likely caused by the siderosis as by a tumour of the cauda equina with repeated bleeding. Epilepsy was reported in one patient.² The dementia, if present, occurs in the terminal stage of the disease as a result of necrosis of the cortex due to the haemosiderosis; in addition hydrocephalus may be a causative factor. Only one author⁸ has reported on necroses in two children with this syndrome: a 12

years old boy and a 7 month old baby. In our patient the disease has lasted 5 years with only a minimal progression of the symptoms.

Despite an extensive search no cause or place of the bleeding was found. This is not too much of a surprise since even at necropsy in approximately half of the patients no cause of the bleeding has been found.⁸ In the remaining cases a tumour in contact with the CSF was usually found and sometimes a vascular malformation.

The presence of methaemoglobin in the CSF is remarkable and not reported before in superficial haemosiderosis. It is usually formed in longstanding collections of encapsulated blood, due to the lack of oxygen.⁵ It could be argued that a relative lack of oxygen arises in the condition because of the chronic inflammation of the meninges resulting in the formation of methaemoglobin.

In our patient haemosiderin deposition in macrophages in the CSF was seen supporting the diagnosis of superficial haemosiderosis. We suggest that this clinical syndrome is underreported as it is only mentioned in two major textbooks^{9,10} and in the literature of the last 13 years we found only one publication⁴ paying attention to this syndrome.

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Table CSF findings in the reported patient

	October 1981	November 1981	December 1981	May 1984	June 1984
Total protein (mg/dl)	1.67	2.15	1.96	2.2	1.7
Bilirubin (mg/dl)	0.7	0.9	0.9	0.6	0.8
Methaemoglobin (mg/dl)	11.8	16.2	14.3	15.8	15.7
Cells/mm ³	6 white	11 red	—	8 white	7 white
Cytology	Monocytes macrophages	—	—	Monocytes macrophages some with haemosiderin	—

Normal values: protein 0.15-0.45 mg/dl; bilirubin not present; methaemoglobine not present.

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Complete gaze palsy in pontine haemorrhage

Sir: Henn *et al*¹ have recently shown that in monkeys lesions confined to the reticular formation of the pons (paramedian pontine reticular formation, PPRF) cause abnormalities of vertical as well as horizontal gaze. In man it has long been recognised that pontine lesions cause horizontal gaze defects but vertical palsies have rarely been recorded. We report a patient with a pontine haemorrhage in whom there was both horizontal and vertical gaze palsy.

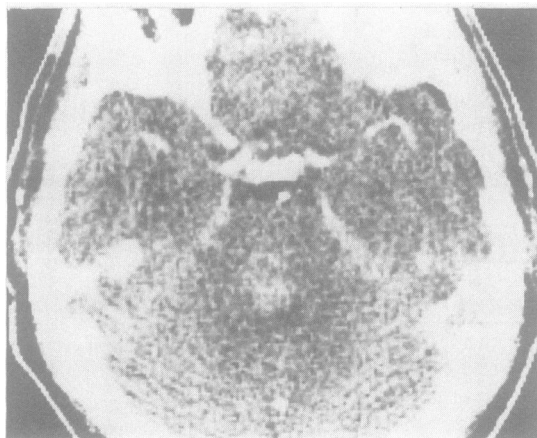
A 55 year old hypertensive male was admitted to hospital with sudden onset of severe walking difficulty and left sided paraesthesia. On examination his blood pressure was 230/140 mm Hg. He was co-operative and alert. Visual acuity and clinical visual field testing were normal. His fundi showed Grade II retinopathy. There was bilateral symmetrical miosis, but the pupils reacted to light and the eyelids were normally posi-

tioned. The patient's eyes were in the primary position and he could not move them on command or in response to stationary or moving visual targets. Doll's head manoeuvre in the horizontal plane, (vestibulo-ocular reflex), as well as caloric irrigation with water at about 20°C failed to induce any eye movement. There was a right sided lower motor neuron facial weakness. The patient was unable to walk because of severe gait ataxia but sensorimotor examination of the limbs while lying down was normal. Tendon reflexes and plantar responses were also normal.

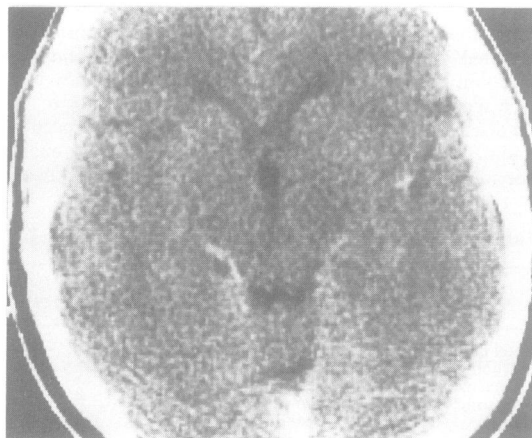
Investigations included normal CSF examination. A CT scan (fig a) showed a pontine haemorrhage occupying the medial tegmental area, slightly more on the right, extending up to the ponto-mesencephalic junction. Mesencephalic and mesencephalon-diencephalic junction sections (fig b) were normal. The patient's hypertension was controlled with propranolol (160 mg a day) and nifedipine (40 mg a day). On the third day the patient noticed diplopia and oscillopsia and examination showed he had recovered a few degrees of conjugate vertical eye movements and abduction of the left eye, accompanied by left beating nystagmus on attempted gaze to that side. After a week the patient's gait improved and he was able to walk with help although he suffered from vertigo on standing. During the second week vertical gaze and convergence were restored. Abduction in the left eye was partially recovered. There was a conjugate, torsional nystagmus to the left (clockwise) and a superimposed horizontal pendular nystagmus in the left eye. By the eighth week the haematoma had resolved on CT scanning.

When examined 2 years later the patient had mild cerebellar signs in the left limbs and an unsteady, broad based gait with left lateropulsion. He complained of positional vertigo and horizontal oscillopsia which was worse when viewing with the left eye alone. There was a horizontal pendular nystagmus with an elliptical trajectory more marked on the left eye, a constant clockwise torsional nystagmus in both eyes, a severe right VI nerve palsy, bilateral horizontal gaze palsy much worse towards the right, and a right internuclear ophthalmoplegia (one and a half syndrome); these horizontal gaze defects were less marked on pursuit and doll's head manoeuvre. The rest of the eye movements were normal.

Henn and co-workers showed that restricted caudal lesions in the PPRF can produce permanent abolition of horizontal and vertical saccades and quick components of nystagmus in the monkey.¹ Clinical examples of documented central pontine lesions in cooperative patients are not very common and the effects on vertical gaze have been variable and contradictory.¹⁻⁶ For instance, no case reported had a permanent vertical gaze palsy; some of the patients showed normal vertical eye movements whereas others had only slowed vertical saccades. The case reported here had clinical and radiological evidence of a pontine lesion. However, the lesion was not restricted to the PPRF, since smooth pursuit, saccades and the vestibulo-ocular reflex were also affected. It is possible that involvement of other neighbouring pontine structures such as the VI nuclei, MLF and/or vestibular pathways may partly account for this.



(a)



(b)

Fig (a) Enhanced CT scan showing tegmental haemorrhage in the pons. (b) Normal appearance of the enhanced CT scan at the level of the superior colliculi.