

LETTERS TO THE EDITOR

Swelling at the site of a skull defect during migraine headache

Goltman¹ reported a patient with a cranial bone defect who had noted that the affected area became depressed before her migraine headache started and bulged during the headache phase. A patient's description of the same phenomenon prompts this brief case report.

A security patrolman aged 33 had been subject to headaches since he was 14 years old. These had recurred every six to 12 months until he reached the age of 31 when the frequency increased to twice each month. At the age of 18 he had fallen from a building site, sustaining a comminuted fracture of the skull that necessitated the removal of a bony fragment from the occiput.

His headaches were preceded by blurred vision associated with the impression of "bubbles like those in a fish tank, floating in circles" which persisted for 20-60 minutes. The headache then started as a pain in his right temple that radiated backwards to the right occiput. As the headache increased in severity, he became nauseated and sensitive to sound but not to light or smells. He then became aware that his bony defect had swollen and felt firm "like a large marble or small egg". It became larger as the headache developed and later subsided as the headache eased. He had not observed any change in the skull defect during his visual aura. On examination, the defect, roughly 3 × 2 cm, was palpated in the midoccipital region but no other relevant abnormality was detected.

It is odd that 60 years have elapsed since Goltman's paper without similar descriptions being published. Nevertheless, these findings indicate that, in some patients at least, intracranial pressure increases during migraine headache. Possible mechanisms include dilatation of intracranial arteries,² the release of vasodilator peptides such as calcitonin gene related peptide (CGRP),³ a perivascular "sterile inflammatory response",⁴ and, possibly, breakdown of the blood-brain barrier.

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Thalamic stimulation for severe action tremor after lesion of the superior cerebellar peduncle

Severe postural cerebellar tremor (SPCT) generally involves cerebellar outflow tracts. Experimental lesions of the superior cerebellar peduncle produce severe SPCT in monkeys,¹ but clinical and pathological correlations are rarely available in humans. This kind of tremor is particularly disabling. Many drugs have been used without success. Stimulation of the ventral intermediate nucleus of the thalamus has been recently considered to be efficient in parkinsonian and essential distal tremors but not in proximal postural tremors.² Interest in this surgical technique has grown with the use of a multiple point electrode which offers the possibility of stimulating different areas within the ventral intermediate nucleus.³ We report a patient with severe SPCT secondary to a cavernous angioma limited to the superior cerebellar peduncle who was dramatically improved by thalamic stimulation.

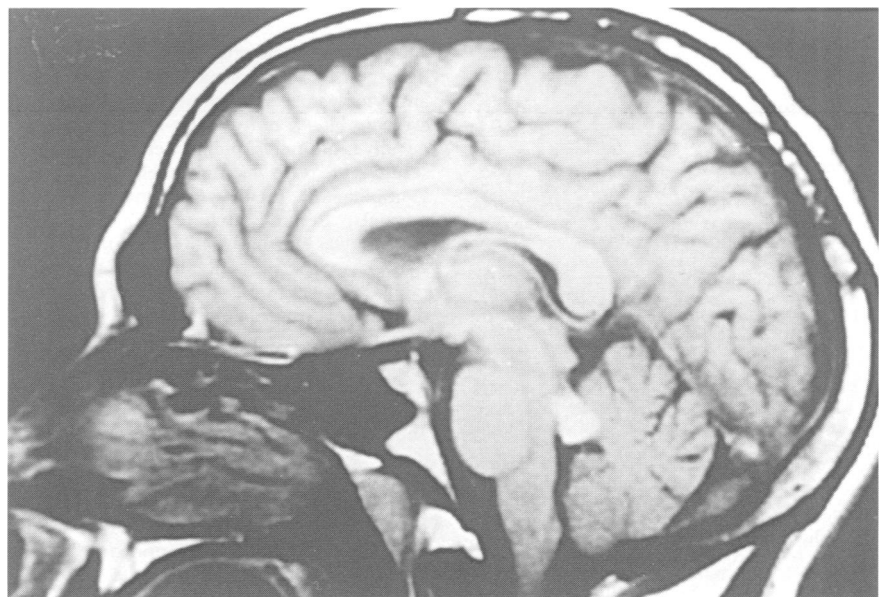
A 33 year old man had an acute paresis of the right arm with mild dysarthria, transient vertigo, and left thoracobrachiofacial paraesthesia in January 1990. Two days later, he complained of severe tremor of his right arm. Neurological examination disclosed left thermoalgic hypoesthesia, rotatory nystagmus, right Horner's syndrome, and severe action tremor of his right arm. His gait was mildly disturbed. Magnetic resonance imaging showed a small haematoma in the right superior cerebellar peduncle (figure). The patient was operated on in October 1990 and the diagnosis of cavernous angioma was confirmed histologically after neurosurgical excision. The tremor disappeared for one week only.

The patient continued to complain of a severe tremor of his right arm and was admitted to our unit in May 1991. The amplitude of the tremor was greatest (more than 10 cm) when the patient tried to keep his forefinger near his nose and was least (less than 2 cm) when he stretched out his arms. It was so disabling that the patient had no functional use of his right arm and was unable to perform daily activities such as

drinking, writing, or eating. A mild postural tremor of the right leg was present without functional consequences. No rest or head tremor was seen. Clinical examination showed a slight hypotonia of the upper and lower right limb, right patellar pendular reflex, and hypoesthesia of the left face. Somatosensory evoked potentials showed normal latencies.

The patient underwent stereotactic implantation of a brain electrode with four contact points (Medtronic®).³ Contrast ventriculography was used to identify the anterior and posterior commissures. The target was defined as the intersection of the inter-commissural line and the rostral part of the ventral intermediate nucleus located according to Guiot's geometric diagram. The electrode was introduced to place the three distal contacts in the left ventral intermediate nucleus. During the operation, the patient was awake and the abolition of the tremor confirmed the adequate placement of the electrode. On another occasion, the electrode was connected to a Ytrell II stimulator (Medtronic®). With optimal stimulation (amplitude: 2.5 V; frequency 130 Hz, pulse width 270 μs, negative plots 2, 1, and 0, positive case), the patient was able to drink from a full glass, catch an object, and write with moderate difficulties. No adverse effect was seen. Examination when the stimulator was on disclosed a right kinetic cerebellar syndrome with hypermetria, adiadococinaesia, and slight hypotonia. In 1993, the patient continued to be improved by the stimulation.

The tremor in our patient affects proximal more than distal muscles and could be considered according to classic terminology as a severe postural cerebellar tremor.⁴ This has been classically associated with lesions of the superior cerebellar peduncle. This kind of tremor was initially described in ischaemic or tumorous lesions of the brainstem, but the most common aetiologies are multiple sclerosis and severe brain trauma.⁴ In these conditions, the lesions are not usually limited to the superior cerebellar peduncle. Our finding is interesting because of the limited character of the lesion and illustrates the role of the superior cerebellar peduncle



T1 weighted (TR/TE = 600/20) MRI shows a small haematoma in the right superior cerebellar peduncle.

- 1 Goltman AM. The mechanism of migraine. *J Allergy* 1935/36;36:351-5.
- 2 Thomsen LL, Iversen HK, Olesen J. Cerebral blood flow velocities are reduced during attacks of unilateral migraine without aura. *Cephalalgia* 1995;15:109-16.
- 3 Goadsby PJ, Edvinsson L. The trigeminovascular system and migraine: studies characterising cerebrovascular and neuropeptide changes seen in humans and cats. *Ann Neurol* 1993;33:48-56.
- 4 Moskowitz MA. Interpreting vessel diameter changes in vascular headaches. *Cephalalgia* 1992;12:5-7.

in the production of ipsilateral severe postural tremor. Cavemous angiomas are congenital vascular malformations of small size usually located in the supratentorial compartment. The commonest infratentorial site is the pons. The involvement of the cerebellar peduncle seems not to be exceptional in the series published recently.⁵ The clinical features of these patients are only briefly described but could include abnormal movements. The description of these movements, however, is incomplete and effective treatment is not reported.

Distal tremors are dramatically improved by chronic stimulation of the lower part of the ventral intermediate nucleus.² Using stereotaxic thalamotomy, Hirai *et al* showed that a large coagulative lesion is necessary to relieve proximal or action tremors and suggested a kinaesthetic somatotopic organisation in the ventral intermediate nucleus.⁶ In particular, the neurons corresponding to the upper arm are located in the upper part of the ventral intermediate nucleus.⁷ The stimulation by an electrode with multiple contact points is a promising technique. It is possible to adjust the site of stimulation according to the different type of postural tremor. This technique has been effective in patients with various causes of tremor including multiple sclerosis and trauma.³ When the stimulation was on, the amplitude of arm tremor decreased dramatically. The improvement of arm function was also important. Activities of daily life such as eating and drinking were dramatically improved. Fine tasks such as writing or manipulation of small objects were possible but difficult. This residual handicap could be explained by the fact that thalamic stimulation does not modify the underlying kinetic cerebellar syndrome. Despite this restriction, the benefit found after thalamic stimulation is important and this treatment should be suggested to patients with this type of tremor.

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Distal myopathy as the presenting manifestation of sarcoidosis

Distal weakness with atrophy is an unusual presentation of a myopathy other than myotonic dystrophy. Inflammatory muscle disease is an exceptional cause of a distal myopathy; only inclusion body myositis affects the distal muscles of the upper limbs. In the present report, we describe a patient with a chronic distal paresis of the lower limbs caused by granulomatous myositis as the presenting and solitary clinical manifestation of sarcoidosis.

A 52 year old woman was investigated for bilateral drop feet. Four years earlier, she had been examined for the same complaint: no diagnosis was made but an unusual induration of the left anterior tibial muscle was noted and peroneal nerve entrapment was excluded.

In 1994, she consulted a neurologist because of progression of the problem. She complained of stiffness of the muscles in the lower limbs, loss of strength, and tripping. She denied cramps or muscle pains. She had no sensory complaints. Her personal medical and family history were unremarkable.

On neurological examination, a symmetric 3/5 dorsiflexion paresis of the feet and 4/5 paresis of plantar flexion was found. A slight 4+/5 paresis of the proximal muscles of the four limbs was noted. All tendon reflexes were present, including both Achilles tendon reflexes. The idiomuscular responses assessed by direct muscle percussion were absent. No fasciculations were seen. On palpation, the gastrocnemius muscle was unusually firm on both sides, but no nodules could be felt. No sensory abnormalities were found. The rest of her physical examination was unremarkable.

Motor and sensory nerve conduction studies were normal. A pronounced induration of the anterior tibial muscle was again noted on EMG needle insertion. The EMG disclosed moderately frequent fibrillation potentials and positive sharp waves in all muscles studied of upper and lower limbs, both distal and proximal, and the paravertebral muscles. The contraction pattern was characterised by small polyphasic motor unit potentials.

C Reactive protein, erythrocyte sedimentation rate, serum creatine kinase, blood cell count, serum electrolytes, and renal and liver tests were normal. Antinuclear and anticytoplasmic antibodies, rheumatoid arthritis latex, and Waaler-Rose tests were negative. No paraproteins were found in the serum on immunoelectrophoresis. Complement (CH50, C4, and C3a) was normal. Serum angiotensin converting enzyme was normal (54 U/l; normal 17-55 U/l). Urinary sediment and calciuria (64.6 mg/24 h) was normal. Fecal cultures showed no enteropathogenic organisms.

Pulmonary function tests showed an obstructive pattern: the forced vital capacity was normal (2.72 l, which is 98% of the value predicted on the basis of sex, length, and age), but the forced expiratory volume in one second was 1.78 l (76% of predicted value), with a forced expiratory volume between 25 and 75% of vital capacity of only 1 l/s (33% of predicted value). The diffusion capacity for carbon monoxide was at the lower limit of normal (5.9 mmol/kPa/min, which is 77% of the predicted value). A bronchoalveolar lavage was refused by the patient.

High resolution CT of the chest disclosed multiple, slightly enlarged (1.5 cm in diameter) pretracheal, precarinal, and infracarinal and para-aortic lymph nodes. No interstitial changes in the lungs were noticed.

A total body gallium-67 scan was performed 72 hours after giving gallium-67 citrate (fig 1). It showed an abnormal uptake of the tracer over the lung hilus on both sides and possibly over the pulmonary fields. Abnormal tracer uptake was clearly present in the muscles of both upper and lower limbs, and in the lacrimal gland.

A biopsy of the gastrocnemius muscle was performed. Multiple non-caseating granulomas were present (fig 2). They were composed of typical epithelioid histiocytes and multinuclear giant cells of the Langhans type, intermingled with lymphocytes and plasmocytes. Some of the granulomas were located in the perimysium and endomysial septa, but they spread out in the muscular fascicles. A slight interstitial lymphocytic infiltrate was present around the granulomas. Individual necrotic fibres and basophilic fibres were present in the immediate vicinity of the granulomas. No eosinophilic granulocytes were found. Staining for fungi, acid fast bacilli, or other bacteria or parasites was negative. There was no grouping of fibre types.

The diagnosis of sarcoidosis was made. Oral corticosteroids (methylpredisone (32 mg twice daily)) did not improve muscle strength or pulmonary function during a follow up period of five months. The corticoid treatment was discontinued at the patient's request. She has been stable over the past six months.

This chronic distal granulomatous myopathy, involving the anterior compartment of the lower legs more severely than the posterior compartment, seems to be the presenting manifestation of systemic sarcoidosis in this patient. Although sarcoid granulomas can be found in muscle biopsies

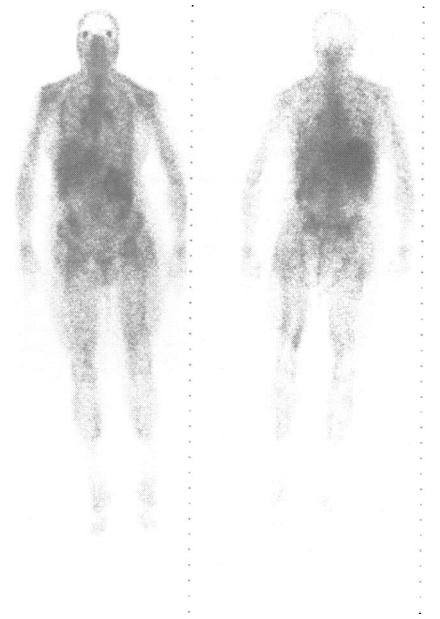


Figure 1 Gallium-67 total body scan 72 hours after giving gallium-67 citrate, showing the increased uptake in the hilar region and lacrimal gland, together with diffuse muscular uptake which is especially prominent in the trapezius, deltoid, and gluteal muscles, and in most of the muscles of the lower limbs.