

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.

C GENY
J P N'GUYEN
P CESARO
Department of Neurosciences,
Hôpital Henri Mondor, 94010 Créteil, France.
C GOUJON
P BRUGIERES
J D DEGOS
INSERM U421, 8 avenue général Sarrail,
Faculté de médecine, 94010 Créteil, France.

Correspondence to: Dr C Geny, Service de Neurologie, Hôpital Henri Mondor, F-94010 Créteil, France.

- Carrea RME, Mettler FA. Function of the primate brachium conjunctivum and related structures. *J Comp Neurol* 1955; 102:151-322.
- Benabid AL, Pollak P, Gervason C, *et al*. Long term suppression of tremor by chronic stimulation of the ventral intermediate thalamic nucleus. *Lancet* 1991;337:403-6.
- N'Guyen JP, Degos JD. Thalamic stimulation and proximal tremor: a specific target in the vim nucleus. *Arch Neurol* 1993;50:498-500.
- Hallett M. *Differential diagnosis of tremor*. In: Van Bruyn, ed. *Handbook of neurology*. Vol 5. Amsterdam: Elsevier Science, 1986: 583-95.
- Lonjon M, Roche JL, George B, Mourier KL, Paquis Ph, Lot G, Grellier P. Les cavernomes intracranéens. *Presse Med* 1993; 22:990-4.
- Hirai T, Miyazaki M, Nakajima H, Shibazaki T, Ohye C. The correlation between tremor characteristics and the predicted volume of effective lesions in stereotaxic nucleus ventralis intermedialis thalamotomy. *Brain* 1983; 106:1001-18.
- Ohye C, Shibazaki T, Hirai T, Wada H, Hirato M, Kawashima Y. Further physiological observations on the ventralis intermedialis neurons in the human thalamus. *J Neurophysiol* 1989;61:488-500.

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

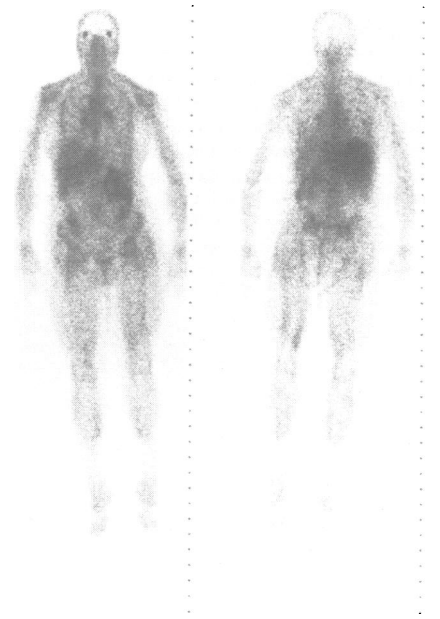


Figure 1 Gallium-67 total body scan 72 hours after giving gallium-67 citrate, showing the increased uptake in the hilar region and lacrimal gland, together with diffuse muscular uptake which is especially prominent in the trapezius, deltoid, and gluteal muscles, and in most of the muscles of the lower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

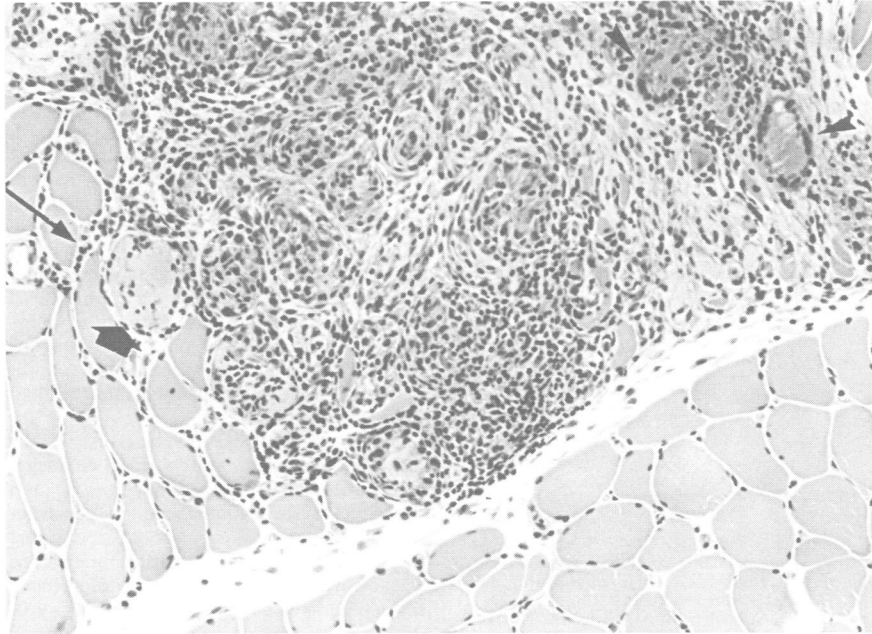


Figure 2 Biopsy of the gastrocnemius muscle showing multiple granulomas in an interfascicular septum. Multinucleated giant cells are present (arrowheads). A lymphocytic infiltrate invades the surrounding muscle fibres (long arrow). A necrotic fibre around the granuloma is shown (thick arrow).

of about 50% of patients with sarcoidosis, a clinical myopathy is seen in less than 1% of them, corresponding to 12% to 26% of cases of neurosarcoidosis.¹ Rarely, a myopathy is the presenting feature: Stern *et al* found only one such presentation out of 649 patients.¹ Chapelon *et al* described three patients with a myopathy as the only clinical manifestation of sarcoidosis during a follow up of 104 months.²

Although the course of sarcoid myopathy can be acute or subacute, a chronic course with atrophy (or pseudohypertrophy) seems to be more frequent. It almost always presents as a proximal myopathy, sometimes affecting the bulbar muscles. A distal myopathy, as seen in our patient, is an exceptional presentation of this already unusual disease. Dyken reported one case with proximal weakness in the legs, and with mild distal involvement of the upper limbs although the shoulder girdle muscles were normal.³ Chapelon *et al* reported one patient with a proximal myopathy who also had distal involvement.² Wolfe *et al* (patient 2)⁴ and Meyer and Regli⁵ each reported one patient with possible distal involvement, but the coexisting peripheral neuropathy made the interpretation difficult.

The pathogenesis of sarcoid myopathy is uncertain. The inflammation is clearly concentrated in the connective tissue department of the muscle, affecting muscle fibres as "innocent bystanders". Our finding, however, that the infiltrate readily invades muscle fascicles, suggests that a true myositis may contribute to the clinical weakness.

Unfortunately, although inflammation is thought to be a reversible cause of muscle disease and several authors report a favourable effect of steroids on sarcoid myopathy,^{1,4} we were unsuccessful in treating the patient with steroids. The chronicity of the inflammation accompanied by fibrosis is probably a major factor in this steroid resistance.

In summary, the present case suggests that sarcoidosis should be considered in the differential diagnosis of a distal myopathy

and that a chronic myopathy with distal predominance can be the sole manifestation of sarcoidosis for a period as long as four years.

W ROBBERECHT
P THEYS

Department of Neurology,
University Hospital Gasthuisberg,
Leuven, Belgium

M LAMMENS
Department of Pathology,
University Hospital Gasthuisberg,
Leuven, Belgium

J LEENDERS
Department of Neurology,
General Hospital St.-Dimpna,
Geel, Belgium

- 1 Stern B, Krumholz A, Johns C, Scott P, Nissim J. Sarcoidosis and its neurological manifestations. *Arch Neurol* 1985;42:909-17.
- 2 Chapelon C, Ziza JM, Piette JC *et al*. Neurosarcoidosis: signs, course and treatment in 35 confirmed cases. *Medicine* 1990; 69:261-76.
- 3 Dyken PR. Sarcoidosis of skeletal muscle. A case report and review of the literature. *Neurology* 1962;12:643-51.
- 4 Wolfe SM, Pinals RS, Aelion JA, Goodman RE. Myopathy in sarcoidosis: clinical and pathological study of four cases and review of the literature. *Sem Arthr Rheum* 1987;16: 300-6.
- 5 Meyer HJ, Regli F. Über die Beziehungen der Polymyositis granulomatosa Boeck zur late onset myopathy. *Dtsch Z Nervenheilk* 1965; 186:547-70.

Unilateral proptosis due to cerebellar stroke

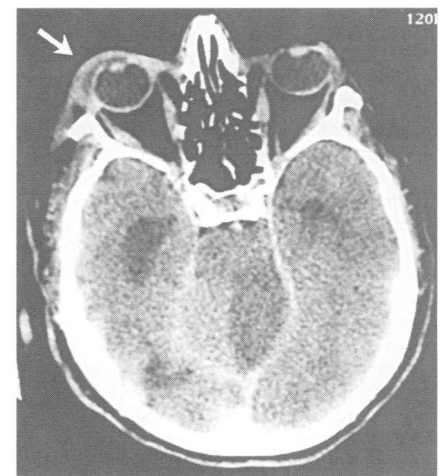
We report a case of unilateral proptosis resulting from a cerebellar stroke causing acute hydrocephalus. Ventriculostomy to alleviate the increased intracranial pressure resulted in regression of the proptosis.

A 73 year old woman was admitted to hospital for right sided hemiparesis that appeared on the morning of her admission. Her medical history included mitral regurgitation, chronic atrial fibrillation, and hypertension. Despite recurrent transient ischaemic attacks in the past she had never received anticoagulant or antiplatelet treatment.

On admission the patient was afebrile, her pulse was 107 beats/min and irregular, her blood pressure was 180/100 mm Hg. An apical systolic murmur was heard. She was lethargic but responded to verbal stimuli. The right pupil was slightly wider than the left, both equally reactive to light. Right sided hemiparesis and an extensor plantar response were noted. Brain CT obtained 48 hours after her admission showed a hypodense lesion within the left cerebellar hemisphere, with surrounding oedema, consistent with a recent stroke in the territory of the left anterior inferior cerebellar artery. On the fourth day in hospital she became somnolent, responding only to painful stimuli. Ptosis and proptosis of the right eye with pronounced chemosis appeared. Oculocephalic response was normal on the left, but she had seventh and partial third nerve palsies on the right. The pupil was dilated and unresponsive to light. Repeated CT showed a large cerebellar infarction with massive oedema, compression of the fourth ventricle, and hydrocephalus (figure). A continuous pressure controlled ventricular drainage system was inserted, with prompt improvement in the patient's alertness and regression of the ocular findings within 24 hours. Her subsequent hospital course was complicated by staphylococcal septicaemia and she died on the 12th day after her admission.

Unilateral proptosis is almost invariably the result of acute or chronic disorders within or around the orbit.¹ Rarely, it is caused by chronic increase in intracranial pressure.^{2,3} We are unaware of a previous report of proptosis as the consequence of a posterior fossa stroke complicated by acute hydrocephalus. Nor has proptosis been associated with other causes of acutely increased intracranial pressure. The postulated mechanism by which intracranial hypertension may lead to exophthalmus is transmission of increased pressure to the orbital veins, resulting in orbital tissue oedema. Proptosis caused by this mechanism is usually symmetric but due to the highly variable drainage of the cavernous sinuses it could be unilateral.⁴

Compression of the fourth ventricle is a recognised complication of cerebellar strokes and surgical interventions may be indicated for relieving hydrocephalus and brain stem



Enhanced axial CT showing hypodensity of the left superior vermis with mass effect compressing the pons and aqueduct. There is secondary enlargement of the temporal horns. The right eye is proptotic with preseptal oedema (arrow).