

thesis, but had normal CSF albumin and normal plasma proteins. Similar changes often occur in multiple sclerosis and other chronic and subacute inflammatory diseases of the nervous system but are usually of lesser degree than those observed in our patient.¹⁶ Elevations of CSF gamma globulin to levels higher than 30% of the total protein are very unusual in neurological disorders other than subacute sclerosing panencephalitis and chronic rubella panencephalitis.¹⁶ Oligoclonal bands, however, often seen in chronic inflammatory disorders of the nervous system, were not observed in our patient.

This patient emphasises the protean clinical manifestations of Behçet disease and suggests that this diagnosis should be entertained in young patients with cerebrovascular disease and evidence of a chronic noninfectious inflammatory CNS process.

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VICENTE J IRAGUI,
ELIAS MARAVI,
Department of Neurosciences,
University of California,
and Veterans Administration Medical Center,
San Diego, CA
Residencia V. del Camino,
Pamplona,
Spain

Address for correspondence: Vicente J Iragui, MD, Ph.D. Department of Neurosciences (M-008), University of California, San Diego, La Jolla, CA 92093, USA.

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Lhermitte's "sign" due to thoracic cord compression

Sir: Lhermitte's "sign", really a symptom, is a sensation of an electric shock radiating down the body and into the limbs on flexion of the neck. Lhermitte described the condition in 1924¹ and believed it was pathognomonic of multiple sclerosis. Marie *et al*² had described a similar symptom in a patient who had injured the cervical spine. The symptom occurs in 25-33% of patients with multiple sclerosis³ but has been reported in many other conditions including cervical spondylosis, tuberculosis of the cervical spine, arachnoiditis, radiation myelitis, tumours of the cervical cord, Behçet's disease⁴ and subacute combined degeneration of the cord.³ Clinical opinion is that "this phenomena denotes a lesion of the cervical cord and occurs most frequently but not exclusively in patients with demyelinating disease".⁵ Most

patients in whom it has been reported, however, have no compressive or structural disease that is easily localised to the cervical cord. Although the arms are spared in the majority of patients,³ the possibility that pathology in the thoracic cord might give rise to this clinical phenomenon has not been considered.

A 23-year-old woman presented to her general practitioner describing a tingling sensation, which began in her feet and ascended both legs over 3-4 weeks, being associated with numbness in both legs. At the same time she described the feeling of an "electric shock" radiating down her back and into both legs on flexion of her neck. Because of the presence of Lhermitte's "sign" it was assumed that she had suffered an episode of demyelination and no further investigation was undertaken. Apart from a brief episode of improvement her condition became progressively worse and she was referred for a neurological opinion 6 months later. In addition to the sensory disturbance, she now also described a two month history of weakness of the legs, which impaired her running. She was admitted for investigation by which time she had developed urinary urgency and occasional incontinence. The major complaint was, however, Lhermitte's phenomenon, which radiated to the legs but not to the arms. For this reason the suspected clinical diagnosis was one of demyelinating disease.

On examination she was a fit young woman. Neurological examination revealed a mild spastic paraparesis with a sensory level to superficial sensation at the level of D7. The reflexes were symmetrically brisk and the plantars were extensor. Routine haematological and biochemical investigations were normal. A myelogram showed a complete block at D5 caused by extradural compression. The texture of the adjacent vertebral body appeared abnormal. A D5 laminectomy was performed and a hard calcified tumour was removed. The histology was that of a cavernous haemangioma. Post-operatively the Lhermitte's "sign" resolved, bladder function returned to normal and there was no weakness in her legs.

Although Lhermitte's "sign" is most commonly seen in multiple sclerosis³ it should not be regarded as pathognomonic. It does occur with other pathology affecting the cervical cord. This case demonstrates that symptoms, without radiation to the upper limbs, can be caused by pathology in the thoracic cord. In this case the pathology was extrinsic, but it is possible that similar symptoms might arise from intrinsic demyelinating lesions in the thoracic cord. In

a rather simplistic way clinicians have assumed that neck flexion will cause mechanical stresses only to the cervical cord. Movement of the cord relative to the thoracic spine may also occur during neck flexion, and could be a mechanism of this woman's symptomatology. For this reason it is important that myelography include examination of the entire spinal cord in patients describing this symptom, in whom electric shock sensations radiate only to the legs.

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RN BALDWIN,
D CHADWICK,

Mersey Regional Department of Medical
and Surgical Neurology,
Walton Hospital,
Rice Lane, Liverpool, L9 1AE,
UK

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A tonic pupil with Horner's syndrome

Sir: The conjunction of a Horner's syndrome and features of a tonic pupil with an apical carcinoma in the superior pulmonary sulcus and paravertebral gutter produces unusual and misleading physical signs. We report such a case.

A 71-year-old female patient presented with a 2 year history of progressive tingling and numbness over the ulnar aspect of her left forearm and hand associated with shooting pains triggered by touch. Over a two month period she had noticed increasing pain, weakness and muscle wasting of her

left hand with intermittent blurring of vision in her left eye. There was no past history of visual disturbance or neurological disorder.

On the left she had moderate ptosis, a pupil which was slightly larger than the right in daylight, and conjunctival injection. The left pupil was smaller than the right when examined in dim light. Infrared television pupillography¹ revealed an abnormally small resting darkness diameter of 3.7 mm (right eye: 5.2 mm) with almost no response to light (<0.2 mm constriction); accommodative effort to near vision resulted in a 0.9 mm constriction which was abnormally slow in both onset and offset, characteristic of a tonic pupil. Slit lamp examination revealed slight segmental movement in the upper part of the left iris in response to near accommodation. The right pupil was normal for age in all respects. Ocular movements were full and other cranial nerves were intact. In her left upper limb the skin was dry and there was wasting of forearm and hand muscles with weakness of triceps, finger extension, wrist and finger flexion and all small muscles of her hand. The triceps jerk was absent. Sensation was impaired over the C7, C8 and T1 dermatomes. No motor, sensory or reflex abnormality was found in other parts of the body. There was fullness in her left supraclavicular fossa although no mass was palpable.

Segmental electromyography demonstrated denervation of C7, C8 and T1-innervated muscles. Nerve conduction studies showed an absent left ulnar sensory action potential and a median sensory action potential of 15 μ v. There was an absent flare response following intradermal injection of 0.016 ml histamine acid phosphate 1 mg/ml to the inner aspect of her left forearm; the flare was preserved on the right. Sweat testing with quinzarin powder was inconclusive. Radiographs of her cervical spine showed degenerative change but other radiological investigations, including chest radiography, AP tomography of the mediastinum and cervical spine, cervical myelogram and CT scan of neck, were all negative. Examination of the cerebrospinal fluid was normal and syphilis serology was negative.

Responses of both pupils to topical drug applications were tested on four occasions each separated by at least 3 days. The findings were:

Drug	
Phenylephrine 2%	
Hydroxyamphetamine 0.5%	
Cocaine 4%	
Pilocarpine 0.05%	

The affected pupil was moderately supersensitive to the direct-acting sympathomimetic phenylephrine, dilated normally to hydroxyamphetamine, which causes noradrenaline release from the sympathetic nerve terminal, and was unresponsive to cocaine, which blocks noradrenaline re-uptake. These findings are consistent with a pre-ganglionic sympathetic nerve lesion. The affected pupil was also supersensitive to the constrictor action of the direct-acting cholinomimetic pilocarpine, which is indicative of a parasympathetic nerve lesion.

Exploration (Mr K Burnand) revealed extensive tumour in the left para-vertebral gutter; the T1 root was oedematous and ran through the tumour mass. Biopsy of the tumour showed anaplastic carcinoma.

The left pre-ganglionic sympathetic lesion was caused by tumour infiltration in the region of the T1 root.^{2,3} The ocular features were unusual for Horner's syndrome in a number of respects, namely that the pupil was slightly dilated when examined in normal room lighting, that there was a minimal light reflex and that accommodative effort produced a large but very slow response. In view of the segmental iris movement and supersensitivity to dilute pilocarpine, it seems likely that these atypical features were due to a concomitant pre-existing post-ganglionic parasympathetic lesion such as is seen in the Holmes-Adie pupil. There was no evidence of local infiltration by tumour of the ciliary ganglion, meninges or central nervous system. It was conceivable that the drug responses in this patient were influenced by a change in corneal permeability but we know of no evidence that this was the case.

Patients with Horner's syndrome associated either with ipsilateral accommodative paresis or with other disorders of accommodation have been described in the past.⁴ However, this unusual conjunction of sympathetic and parasympathetic lesions has not, to our knowledge, been previously reported and it exemplifies the value of pupillography and pharmacological testing in the diagnosis of pupillary dysfunction.

P ST J TREND,
SE SMITH
CM WILES,

Department of Neurology,

St Thomas' Hospital, London SE1 7EH, UK

Diameter change (mm)

Right pupil	Left pupil
2.50	3.40
2.00	1.82
0.90	0.08
0.75	-0.93