Painless cauda equina schwannoma simulating Charcot-Marie-Tooth disease

Sir: Pain is almost always the principal symptom of patients who have tumours of the cauda equina. Painless tumours at this site, although rare, do occur and should be considered in the differential diagnosis of pain free patients presenting with progressive weakness in the lower limbs. The following case report demonstrates this point.

A man, age 38 years, was admitted with 10 years’ history of progressive weakness of the legs, the left being more severely affected. There was no history of any pain or paresthesia in his back or legs. Other symptoms included a recent constant feeling of urgency, occasional nocturia, constipation, and weak penile erection. Previously he was admitted to another hospital because of a left ankle sprain, leg weakness, and a gait disturbance, at which time a diagnosis of peroneal muscular atrophy was made. The abnormal neurological signs were confined to the lower limbs. Moderate to severe weakness and wasting, especially of hip extensors, foot dorsiflexors, extensors of toes, foot dorsiflexors, and invertors, were present with bilateral foot drop. Knee jerks, ankle jerks, and plantar responses were normal. There were no sensory changes. The rectal sphincter was hypotonic. Straight leg raising to 90° produced no pain or discomfort. Lumbar spiné radiographs were normal. Nerve conduction studies with standard techniques showed normal maximum conduction velocities in right median, ulnar, tibial, and sural nerves, and borderline conduction velocities in left tibial nerve. Supramaximal percutaneous stimulation of the peroneal nerves produced no muscle action potential. Needle electromyography showed extensive denervation potentials in glutei maximus and in the muscles innervated by both sciatic nerves but no abnormality in paraspinal muscles. Myelography revealed a mass at the cauda equina region. Spinal fluid protein was elevated (1.28 g/l).

Urodynamical studies were consistent with a spastic neurogenic bladder. Laminectomy of D12 and L1 vertebrae was performed and a large Schwannoma was removed totally.

Cauda equina tumours are rare. Occasional cases present with unusual clinical features such as pseudoclaudication,¹ foot ulceration,² subarachnoid haemorrhage,³ papilloedema, and sensory ataxia,⁴ but it is generally accepted that pain in the back and lower limbs is nearly always the most important and early presenting symptom.⁵ Pain may be of sudden or gradual onset,⁶ usually tends to become constant, and characteristically worsens at night.⁷ Allen⁸ and Spiller⁹ reviewed the literature of cauda equina tumours and both found only one case of cauda equina compression with no pain, reported by Volhard.¹⁰ Campbell¹¹ could find clinical details of only two cases of painless tumour of the cauda equina and presented one of his own. In a series of 20 patients from the National Hospital, Queen Square,¹² only one had no pain and presented with left leg weakness for seven years; but finally he developed severe lumbar pain. In an Oxford¹³ series of 70 patients, six complained of painless progressive weakness of legs; only three had bilateral leg weakness, and pain was the presenting symptom in 31 out of 34 patients with neurofibromas of the lumbo-sacral region.³

In sum, a case is reported of a patient with a Schwannoma of the cauda equina who carried the diagnosis of Charcot-Marie-Tooth for 10 years. There was slowly progressive motor dysfunction without pain or sensory loss during this time. Electromyography excluded Charcot-Marie-Tooth disease,¹²-¹³ and indicated a lesion of the cauda equina and, therefore, mandated myelography. This emphasises the potential diagnostic significance of proper electromyographic study in patients with progressive neuromuscular disease.

F ROOH
AW COOK
H CLARKE
R TORN
M ZANDIEH
Department of Neuroscience,
The Long Island College Hospital,
340 Henry Street,
Brooklyn, New York 11201, USA

References


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Use and misuse of the Roussy-Levy eponym

Sir: On a clinical and electrophysiological basis only, Yudell, Dyck and Lambert in 1965¹ described a family with a dominantly inherited form of peroneal muscular atrophy which they called Charcot-Marie-Tooth. As this group of patients showed substantially reduced motor conduction velocity they were assumed to have one bulb in their peripheral nerves and were therefore labelled hypertrophic Charcot-Marie-Tooth disease. Four of the nine patients in this kindship had a disorder of movement similar to essential (familial) tremor of unusually great amplitude. This association of signs was considered to be an example of the Roussy-Levy disease.² The use of this eponym for such patients seems to us to be illogical since in 1906 P Marie³ had described a family with a dominantly inherited peroneal muscular atrophy, hypertrophic nerves and essential tremor of great amplitude. Boveri in 1910⁴ provided the post mortem material of the member of the kinship who had the tremor of the largest amplitude. Since that time this type of hypertrophic neuritis has been known as the Pierre Marie-Boveri type. P Marie did not say that his patient had the hypertrophic variety of Charcot-Marie-Tooth disease associated with essential tremor and

Letters