Painful 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, aged 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 evertors 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 hypertonic. Straight leg raising to 90° produced no pain or discomfort. Lumbar spine 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 maximii 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). Urodynamic 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 fashions such as pseudoclaudication, foot ulceration, subarachnoid haemorrhage, papilloedema, and sensory ataxia, but it is generally accepted that pain in the back and lower limbs is nearly always the most important and early presenting symptom. Pain may be of sudden or gradual onset, usually tends to become constant, and characteristically worsens at night.

Allen and Spiller reviewed the literature of cauda equina tumours and both found only one case of cauda equina compression with no pain, reported by Volhard. Campbell 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, only one had no pain and presented with left leg weakness for seven years; but finally he developed severe lumbar pain. In an Oxford 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 lumbar-sacral region.

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, and indicated a lesion of the cauda equina and, therefore, mandated myelography. This emphasizes the potential diagnostic significance of proper electromyographic study in patients with progressive neuromuscular disease.

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References


Use and misuse of the Roussy-Levy eponym

Sir: On a clinical and electrophysiological basis only, Yudell, Dyck and Lambert in 1965 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 onion bulbs in their peripheral nerves and were therefore labelled hypertrophic Charcot-Marie-Tooth. Four of the nine patients in this kinship 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. The use of this eponym for such patients seems to us to be illogical since in 1906 P Marie had described a family with a dominantly inherited peroneal muscular atrophy, hypertrophic nerves and essential tremor of great amplitude. Boveri in 1910 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
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*J Neurol Neurosurg Psychiatry* 1982 45: 938
doi: 10.1136/jnnp.45.10.938

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