Short report

Femoral pain of solitary neurofibromatous origin: a report of three cases

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SUMMARY Three patients, each with a solitary neurofibroma in the thigh, are described. The presenting complaint in each case was pain. A neurofibroma of a branch of the femoral nerve was found in two cases and an intraosseous neurofibroma of the femoral shaft was present in the third. An excellent recovery was observed in each of these three patients following surgical excision of the tumour. Attention is drawn to the unduly long interval between the onset of painful symptoms and the achievement of a clinical diagnosis leading to a surgical cure in each instance.

Involvement of peripheral nerves by tumours is one of the usual features of von Recklinghausen's disease, but this may be absent, particularly in central neurofibromatosis.¹ However, solitary neurofibroma without any other evidence of von Recklinghausen's disease is uncommon, rarely painful and most frequently observed in the upper limbs. We describe three such cases of solitary neurofibroma in the thigh.

Case reports

Case 1

A 35-year-old business man was admitted to Royal Perth Hospital in August 1970, complaining of pain and weakness in the right thigh for 11 years. He felt that his right leg had not fully regained its strength after a meniscectomy 15 years previously. Four years later he began to experience recurrent sharp, stabbing pains for 10 to 20 minutes at a time, between the right mid calf and groin. The pain kept him awake at night and was unaffected by posture, simple analgesics or local heat. Over the ensuing years his right thigh became weaker and he sought advice from numerous clinicians, often being labelled as neurotic. There was no family history of von Recklinghausen's disease. Examination revealed an exquisitely tender area below the right inguinal region, making it difficult to palpate a definite mass. There were five or six small, firm subcutaneous lumps in the anterior abdominal wall which were painless and mobile. There was no cafe-au-lait pigmentation. The

right quadriceps muscle was quite wasted and weak and the right knee jerk was absent. There was loss of light touch sensation over the proximal anterior right thigh. On investigation, myelography disclosed minor degenerative disc changes in the lower thoracic spine, and partial unilateral wedging of the right tenth thoracic vertebra. There was electromyographic evidence of chronic partial denervation in the right quadriceps muscle, but normal right femoral nerve distal motor conduction. Histological analysis of one of the abdominal wall lumps which was biopsied, revealed adipose tissue with occasional nerve fibres and collagen. At surgical exploration on 21 August 1970, a large tumour was found attached to a deep branch of the femoral nerve. The tumour measured 5 cm by 3 cm after its extirpation. Microscopically, it consisted of interlacing bundles of collagen tissue and cells with elongated nuclei, some of which were large and hyperchromatic. The tumour was well vascularised and there were areas of recent and past haemorrhage with many pigment-laden macrophages. Some areas were densely cellular and others consisted of loosely packed fibres. There were no mitotic figures. These findings confirmed the clinical impression of neurofibroma. He was assisted by physiotherapy postoperatively and when last examined some months later the bulk and strength of the quadriceps had increased greatly and he was completely free of pain.

Case 2

This 55-year-old male presented to Royal Victoria Infirmary, Newcastle-upon-Tyne on 13 January 1961. Thirteen years earlier he developed a tender swelling on the outer aspect of the right thigh. This persisted despite an attempt at removal three years later, but it had not altered in size. For some time he had also been aware of a painful lump on the inner aspect of the right thigh, and in the past 10 years the right thigh had become progressively weaker and more wasted. As a keen mountain climber, he was now unable to undertake this sport. He could not kick a ball

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with the right leg and had difficulty mounting stairs. Examination was entirely negative, except for marked wasting, fasciculation and weakness of the right quadriceps muscle, but there was no weakness of other muscles in either lower limb. All the deep reflexes were symmetrical. Palpable in the substance of the right vastus lateralis was a tumour which was removed on 8 February 1961, through a vertical incision, the fascia lata being divided at a point 2 cm above the previous incision. The tumour had been lying on an intermuscular septum laterally. It was smooth, slightly lobulated, completely encapsulated, measuring 3 cm by 2 cm and arising from a small nerve which was probably a genicular branch of the femoral nerve. Histology confirmed the surgical impression of a neurofibroma. He was seen on 17 September 1961, when it was noted there was a remarkable increase in the power and bulk of the right quadriceps muscle which was not now fasciculating. He was finally reviewed on 21 August 1967, when there was very mild wasting and weakness of the right quadriceps muscle but his functional recovery was complete.

Case 3

This man presented to the Regional Neurological Centre, Newcastle-upon-Tyne on 4 February 1964, with a history that for nine years he had suffered intermittent pain in the right knee with the knee giving way. For 18 months he had complained of a dull ache along the right groin, which would often awaken him at night. In addition, there had been progressive weakness of the right leg over the past three years and he had developed a limp. For two years he had noticed a tender spot on the inner aspect of the right mid thigh, pressure over which reproduced his stabbing pain. Examination showed no café-au-lait patches and no palpable tumours, but he was tender over Hunter's canal on the right side and the clinical diagnosis was of a neurofibroma on a twig of the femoral nerve in Hunter's canal. Radiographs of the lumbar spine were normal. The cerebrospinal fluid was normal with a protein level of 0.26 g/l. Exploration of Hunter's canal was undertaken on 11 February 1964, and no abnormality was discovered. The operation made no difference to his symptoms and he continued to complain of severe pain. At review, on 4 September 1968, there was wasting of right quadriceps and some apparent weakness of the right hip flexors. Radiographs of the right femur showed an eccentric, cystic lesion with a well-defined margin in the region of the mid shaft and expanding the cortex slightly on its medial aspect, but with no evidence of periosteal reaction. The medullary trabecular pattern above the level of the lesion appeared normal. On 8 October 1968, a histologically proven neurofibroma was evacuated from the shaft of the right femur. After operation, he regained full power and enjoyed complete relief of his pain.

Discussion

Neurofibromata of peripheral nerves are often observed in von Recklinghausen's disease. Borchardt,² in 1927, first reported a solitary tumour not related to von Recklinghausen's disease. Since

1962, solitary neurofibromata have become recognised as a distinct entity following Heard's³ report on 46 such peripheral tumours, which included 35 superficial and 11 deep lesions. However, Russell and Rubinstein⁴ doubted the truly solitary occurrence of these tumours. Harkin and Reed⁵ suggested the existence of a forme fruste of multiple neurofibromatosis, particularly as the majority of patients with solitary neurofibromata of the skin have no other evidence of von Recklinghausen's neurofibromatosis. However, when a diagnosis of neurofibroma is made, the clinician should be obliged to search for other manifestation of generalised neurofibromatosis.⁶ The common presentation of pain in these three cases of apparently solitary peripheral neurofibroma in the absence of stigmata of von Recklinghausen's disease is quite unusual,^{3,7-9} especially in the thigh. Peripheral neurofibromata most frequently involve the nerve trunks of the upper extremities, particularly the ulnar and radial nerves,^{1,7,8} and are usually found in patients with von Recklinghausen's disease.

Rarely have intraosseous cystic neurofibromata been reported^{1,10,11} since the original description of subperiosteal bone cyst formation by Brooks and Lehman.¹² However, all these cases were associated with multiple neurofibromatosis. Solitary neurofibroma of bone is extremely rare. Even though the presence of nerve fibres in bone is generally accepted, only a few cases have been reported in which there was conclusive evidence of endosteal neurofibromatosis.^{13,14} This report emphasises the long interval elapsing between the onset of pain and the diagnosis.

References

- Canale D, Bebin J. Von Recklinghausen's disease of the nervous system. In: Vinken PJ, Bruyn GW, eds. *The Phakomatoses. Handbook of Clinical Neurology*. Amsterdam: North Holland 1972;14:132-62.
- Borchardt M. Zur kenntnis der neurinome. *Beitr Klin Chir* 1927;138:1-38.
- Heard G. Nerve sheath tumours and von Recklinghausen's disease of the nervous system. *Ann R Coll Surg Engl* 1962;31:229-48.
- Russell DS, Rubinstein LJ. *Pathology of Tumours of the Nervous System*. London: Edward Arnold, 1977: 389-97.
- Harkin JC, Reed RJ. Tumours of the peripheral nervous system. In: *Atlas of Tumour Pathology*. 2nd Ser, Fasc 3. Washington DC, Armed Forces Inst: Path 1968.
- Weller RO, Cervos-Navarro J. *Pathology of Peripheral Nerves*. Butterworths, 1977:169-77.
- Kramer W. Tumours of nerves. In: Vinken PJ, Bruyn GW, eds. *Diseases of Nerves. Part II. Handbook of Clinical Neurology*. Amsterdam: North Holland 1970;8:412-512.

- ⁸ Seddon HJ. *Surgical Disorders of the Peripheral Nerves*. 2nd ed. Edinburgh: Churchill Livingstone, 1975:165.
- ⁹ Tognetti F, Poppi M, Gaist G, Servadel F. Pudendal neuralgia due to solitary neurofibroma. *J Neurosurg* 1982;**56**:732–3.
- ¹⁰ Klatte EC, Franken EA, Smith JA. The radiographic spectrum in neurofibromatosis. *Semin Roentgenol* 1976;**11**:17–33.
- ¹¹ Sane S, Yunis E, Greer R. Subperiosteal or cortical cyst and intramedullary neurofibromatosis—uncommon manifestations of neurofibromatosis. A case report. *J Bone and Joint Surg* 1971;**53A**:1194–2000.
- ¹² Brooks B, Lehman EP. The bone changes in von Recklinghausen's neurofibromatosis. *Surg Gynec Obstet* 1924;**38**:587–95.
- ¹³ Dahlin DC. *Bone Tumours: General Aspects and an Analysis of 2776 Cases*. Springfield, Illinois: Charles C Thomas, 1957.
- ¹⁴ Friedman MM. Neurofibromatosis of bone. *A M J Roentgenol* 1944;**51**:623–30.