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CASE REPORT

Neurilemoma of the popliteal fossa : Report of two cases with long subclinical course and misleading presentation

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The authors report two cases of neurilemoma localised in the popliteal fossa. Both patients experienced non-specific symptoms, such as painful numbness and burning dysaesthesia, involving the lower extremity. Tinel's sign was positive over the popliteal fossa. The patients sought medical advice and underwent conservative treatment without any relief, for a long time before the right diagnosis was made. Magnetic Resonance Imaging revealed in both patients a well- circumscribed mass posterior to the sciatic nerve, occupying the popliteal fossa. Following surgical excision of the neurilemoma, the patients experienced immediate relief of their chronic symptoms. In similar situations, ultrasound or magnetic resonance imaging of the whole sciatic nerve should be performed if this is indicated by detailed physical examination. Once the diagnosis is made, neurilemomas should be surgically removed, in order to exclude malignancy, prevent neurologic deficits and provide relief of symptoms.

Keywords : neurilemoma ; popliteal fossa.

INTRODUCTION

Although uncommon, sciatic nerve sheath tumours are usually benign, with neurilemoma (schwannoma) and neurofibroma being the most representative (*3*). Separating schwannoma from neurofibroma clinically, may be difficult. Schwannomas are typically solitary, circumscribed, and encapsulated tumours eccentrically located on proximal nerves or spinal nerve roots. Neurofibromas, by contrast, are not infrequently multiple, are usually but not always non-encapsulated, and are often subcutaneous or produce fusiform enlargement of distal nerves (*6*). Malignant nerve sheath tumours or malignant transformation of benign sheath tumours is unusual (*3*), and is much rarer in schwannomas. A variety of unusual peripheral nerve tumours have been reported in the literature, such as perineuroma, lymphangioma, haemangioma, desmoid tumours, granular cell tumours, ganglion cysts, lipoma, chondroma, osteoma, angioma, haemangioblastoma, haemangiopericytoma, sarcoma and metastatic tumours (*3*). Microscopically, schwannomas have regions of high and low cellularity called Antoni A and B areas, respectively. In the Antoni A tissue there may be foci of palisaded nuclei called Verocay bodies,

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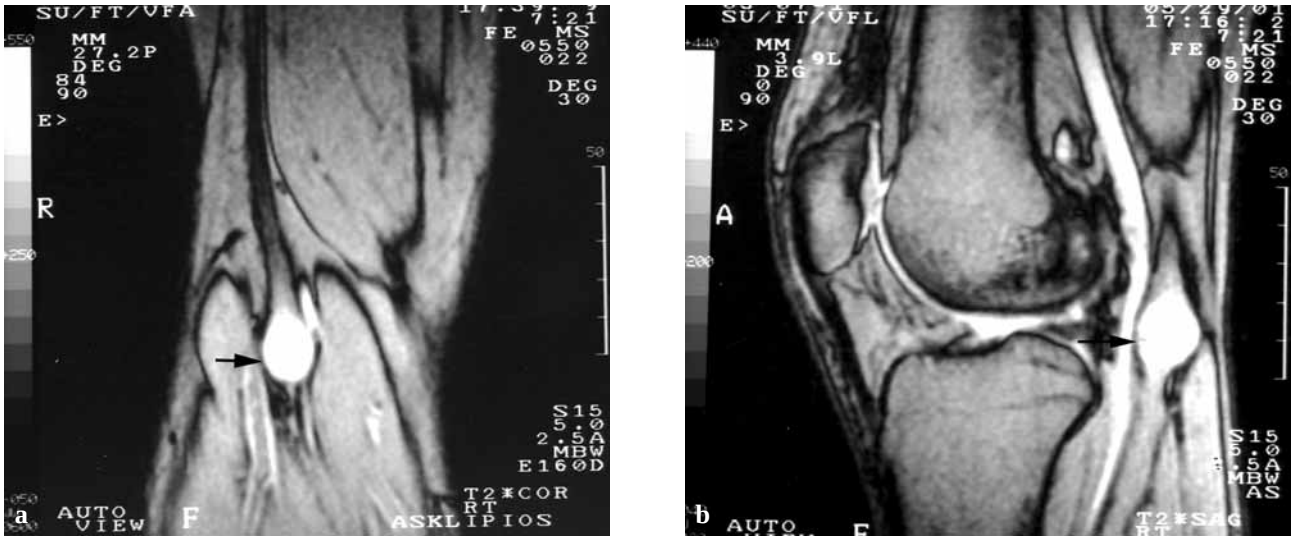

Fig. 1. — a. Coronal T2-weighted MRI scan displays a well-circumscribed neurilemoma ($2 \times 2 \times 1.8$ cm) of the right popliteal fossa of the patient in case 1 ; b. Sagittal T2-weighted MRI scan illustrates the homogeneously enhanced neurilemoma of the same patient, posterior to the sciatic nerve bifurcation.

and the blood vessels in schwannomas often have hyaline thickening around which there may be pseudopalisading of tumour nuclei. Neurofibromas usually consist of a loose pattern of interlacing bands of delicate spindle cells with elongated, slender, and sometimes wavy nuclei (*6*). Although neurilemomas may be found throughout the body, involvement of the sciatic nerve is uncommon. In a total of 557 neurilemomas, reviewed by several authors (*1, 2, 8, 10*), only 34 (5.9%) were localized in the thigh.

CASE REPORTS

Case 1

A 42-year-old man reported painful numbness involving the right lower extremity from the calf to the sole of the foot and the toes. The initial symptoms started two years previously, persisted and progressed during the last 4 months. At the time of his admission the patient described the numbness as a “painful electricity sensation” shooting down from the calf to the sole, exacerbated in the sitting position. He denied any back symptoms or any increase in pain during coughing, sneezing or back movements. The patient also reported diminished muscle strength during dorsiflexion of his right foot. Physical examination revealed only diminished sensation of the lateral plantar surface and diminished Achille’s tendon reflex. Muscle strength was normal. The straight leg raising test was negative. Tinel’s sign was produced by tapping on a painless but palpable mass in the popliteal fossa. Electrophysiologic examination showed normal nerve conductions. Magnetic Resonance Imaging (MRI) revealed a well-circumscribed mass ($2 \times 2 \times 1.8$ cm), just posterior to the sciatic nerve bifurcation, occupying the popliteal fossa (fig 1a, 1b). Surgical excision was decided upon. The patient was placed in a prone position and an S-shaped incision was made over the lesion. A well-circumscribed tumour was identified, originating from the sciatic nerve, just proximal to its bifurcation. The sciatic nerve fascicles were incorporated in the tumour capsule and they spread like a sheath circumferentially. A careful circumferential dissection was performed according to microneurosurgical principles. The tumour was resected and the functioning fascicles were preserved within the outer sheet. Histologic examination revealed a neurilemoma. The patient experienced immediate relief of his

chronic painful numbness. Tinel's sign disappeared. Normal strength of the foot musculature was preserved postoperatively. In less than one month the patient returned to full time job. He remains free of symptoms three years later.

Case 2

A 58-year-old-man presented with a 7-year history of numbness and a painful burning dysaesthesia distributed in the dorsal surface of his left foot. The symptoms persisted and exacerbated during the last two years, involving also the second toe and the first metatarsal joint. The patient sought medical advice several times during this period. Physical examination showed no neurologic deficit. Back mobility was adequate and painless. Lasègue's sign was negative. Laboratory tests and radiographs of the left hip were performed, with normal findings. Electrophysiologic examination showed normal nerve conductions. No clear diagnosis was made, and the patient underwent non-steroidal anti-inflammatory medical treatment and repeated intra-articular steroid injections, but the symptoms were not relieved. A more detailed physical examination was performed and it was noted that finger percussion over the left popliteal fossa caused paraesthesia and dysaesthesic pain with similar distribution (positive Tinel's sign). Local ultrasound examination revealed a well-defined mass with homogeneous echogenicity. MRI showed a well-circumscribed mass ($2.9 \times 2.6 \times 3.5$ cm), posterior to the sciatic nerve, that occupied the popliteal fossa. The patient underwent surgical resection of the tumour according to microneurosurgical principles. He was placed in a prone position and a longitudinal incision was made over the left popliteal fossa. A well-delineated tumour was identified, originating from the sciatic nerve proximal to its bifurcation. The sciatic nerve fascicles were incorporated peripherally on one side of the tumour capsule. The tumour was dissected and resected, with preservation of the functioning fascicles within the outer sheet. Histologic examination revealed a neurilemoma. The patient experienced immediate relief of his chronic dysaesthesias. Some numbness on the dorsal surface of the foot persisted for six months. No neurologic deficit was noted postoperatively. The patient experienced full recovery after three months, without any limitation to mobilisation. He remains free of symptoms three years later.

DISCUSSION

Although rare, schwannomas are the most common benign nerve sheath tumours of the sciatic nerve (*3*). Most are solitary lesions, and multiplicity or malignant transformation occurs rarely (*3*). These tumours grow slowly in the periphery, along the nerve and do not generate symptoms, unless they grow within a confined space (*4*). Pain, not responding to rest or activity (*4*), sensory and motor dysfunction along the nerve distribution are the most common manifestations (*3*). The provocative tension root and mechanical back tests are negative. Other symptoms may occur from local pressure on adjacent tissues due to growth of the tumour (*4*). Nevertheless, clinical presentation is not always obvious and sciatica has occasionally been attributed to plantar nerve neuropathy due to nerve entrapment at the ankle, discopathy, psychogenic or central origin of pain (*3*) and peripheral arthritis as in case 2. A peripheral nerve neurilemoma may also present as a painless, palpable mass with a positive Tinel's sign (external pressure may cause tenderness and paraesthesia), as in case 1. Current reports demonstrate that neurilemomas often have a long subclinical course and their clinical presentation is usually misleading. Patients often seek medical advice and undergo conservative or invasive treatment without any relief, before the right diagnosis is set. The solution suggested to this diagnostic problem, through our cases, is detailed history and physical examination in order to exclude usual causes of sciatica and raise suspicion of peripheral nerve pathology. Sciatica not responding to rest or anti-inflammatory treatment, absence of lumbar contracture, a negative Lasègue's test and a positive Tinel's sign should lead to an ultrasound and if indicated, MRI imaging of the whole leg, to confirm the diagnosis of a peripheral nerve tumour. If these diagnostic procedures prove to be negative, further investigation in

order to exclude other causes of the sciatica should be performed.

Surgical treatment for neurilemomas is recommended, especially, when pain or neurologic deficit is present (*5*). It is reported in the literature that the majority of peripheral nerve schwannomas can be excised with good results (*5, 7*). In an analysis of 16 patients with peripheral nerve schwannomas that underwent surgical excision by Oberle *et al* (*7*), total or partial pain relief and complete or partial recovery of motor deficit was observed in 100%. New motor and sensory deficits developed predominantly, in patients with large tumours or longstanding symptoms. In our case 2, the patient experienced complete relief of his painful dysaesthesias postoperatively, despite the longstanding course of symptoms and the large size of the tumour, demonstrating that surgical intervention, even late, should always be performed. Nevertheless, early surgical intervention, according to microneurosurgical principles, should be the treatment of choice, in order to prevent neurologic deficits and exclude the possibility of malignancy. This report supports others in the literature, demonstrating that surgical treatment is the treatment of choice (*5, 7*).

In schwannomas, no nerve fibers are present in the body of the tumour, contrary to neurofibromas in which nerve fibers are scattered throughout the tumour mass. The distribution of nerve fibers within these tumours has some practical significance, as the peripheral position of a schwannoma on one side of the nerve of origin leaves the possibility to remove the tumour without requiring transection of the nerve (*9*), an option not available for neurofibromas in which the entire nerve is involved in the tumour (*6*). A low incidence of recurrence is reported in the literature (*4*) ; in certain cases, recurrence is considered to be a consequence of incomplete resection Nevertheless, Oberle *et al* (*7*) did not report any tumour recurrence in 16 patients with neurilemomas, during the follow-up period of 23 months. Both patients in the current report remain free of symptoms, three years after total removal of their sciatic schwannoma.

To conclude, the diagnostic workup for peripheral nerve sheath tumours, and especially neurilemomas of the sciatic nerve, should include a) detailed history, b) careful physical examination, c) sonography and if indicated, d) magnetic resonance imaging of the whole sciatic nerve. Once the diagnosis is made, surgical excision should be performed, in order to exclude malignancy, prevent neurologic deficits and provide relief of symptoms.

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