

NEUROFIBROMA OF THE THIGH WITH COMPRESSION OF THE SCIATIC NERVE IN A CHILD

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Abstract

Peripheral nerve sheath tumors account for nearly 12% of the benign and 7-8% of the malignant soft tissue neoplasms. Neurofibromas and schwannomas constitute the benign category, while the malignant peripheral nerve sheath tumor (MPNST) comes under the malignant category.

A 5 year old male patient was admitted to the hospital having a painful tumor of approximately 10 cm diameter on the posterior left thigh with sciatic neuropathy and flexion contracture of the knee.

The patient was living in the rural area under poor social conditions. A neurofibromatosis type I was diagnosed and the child underwent successful surgical removal of the tumor which was compressing the sciatic nerve without relapse at follow-up 18 month after surgery. The histology confirmed the suspicion of a neurofibroma. After postoperative physiotherapy he could resume walking.

Keywords: neurofibromatosis type I (NF I), sciatic nerve, neurofibroma, sciatic neuropathy, child

Introduction

Peripheral nerve sheath tumors account for nearly 12% of the benign and 7-8% of the malignant soft tissue neoplasms. Neurofibromas and schwannomas constitute the benign category, while the malignant peripheral nerve sheath tumor (MPNST) comes under the malignant category.

Neurofibromatosis, also named von Recklinghausen disease for the man who described the disease in 1882, was formerly considered a single disease but is now known to be at least two clinically and genetically distinct diseases. The more common disease, formerly known as the peripheral form of neurofibromatosis, is designated neurofibromatosis 1 (NF1), whereas the less common disease, formerly known as the central form, is designated neurofibromatosis 2 (NF2) (bilateral vestibular schwannoma). A common genetic disease, NF1 affects 1 in every 3500 individuals. It is inherited as an autosomal dominant trait with a high rate of penetrance. Because only half of the patients with this disease have affected family members, the disease in the remaining patients represents new mutations. In the typical patient, NF1 becomes evident within the first few years of life when café au lait spots develop. These pigmented macular lesions resemble freckles, especially during the early stage when they are small. Typically, they become

much larger and darker with age and occur mainly on unexposed surfaces of the body.

Neurofibromas, the hallmark of the disease, make their appearance during childhood or adolescence after the café au lait spots. The time course varies greatly: some tumors emerge at birth, and others appear during late adult life.

They may be found in virtually any location and, in rare instances, may be restricted to one area of the body (segmental neurofibromatosis). Unusual symptoms have been related to the presence of these tumors in various organs such as the gastrointestinal tract. The tumors are usually slowly growing lesions. Acceleration of their growth rate has been noted during pregnancy and at puberty. A sudden increase in the size of one lesion should always raise the question of malignant change.

Case presentation

A 5 year old male patient was admitted to the hospital having a painful tumor of approximately 10 cm diameter on the posterior left thigh with signs of sciatic neuropathy and flexion contracture of the knee (Figure 1). The patient was living in the rural area under poor social conditions.

On clinical exam multiple café au lait spots were present. After a careful anamnesis a neurofibromatosis type I was suspected. The father of the patient turned out to have also multiple criteria for neurofibromatosis I.

The patient was then investigated and a neurofibromatosis I was confirmed. The patient had a optic glioma and 3 Lisch nodules. He also had palpable neurofibromas on the left arm, but without an obvious symptomatology.

X-rays of the left thigh showed no abnormalities. Ultrasound of the tumefaction revealed a non-compressive, anechoic mass with a marginal Doppler signal. Magnetic resonance imaging (MRI) showed a tumor of the thigh with dimensions of 10,4x8,9x6,8 cm with compression of the sciatic nerve with low signal intensity on T1 images, high signal intensity on T2 images, and a heterogeneous pattern.

Foot weakness with primarily tibialis anterior weakness (foot drop) was seen in the patient. Intensity of pain as a symptom of sciatic neuropathy was difficult to evaluate in the young patient, but the parents were able to report that in the last 2 weeks before presenting to the hospital ambulation was not possible at all. Muscular knee contraction was present at clinical examination. A surgical excision of the neurofibroma was performed under general anesthesia (Figure 2, Figure 3).

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Figure 1 – Clinical aspect of the neurofibroma.

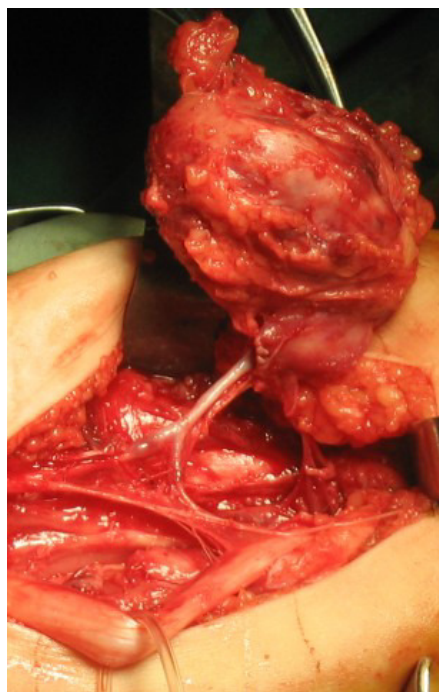


Figure 2 – Intraoperative aspect.

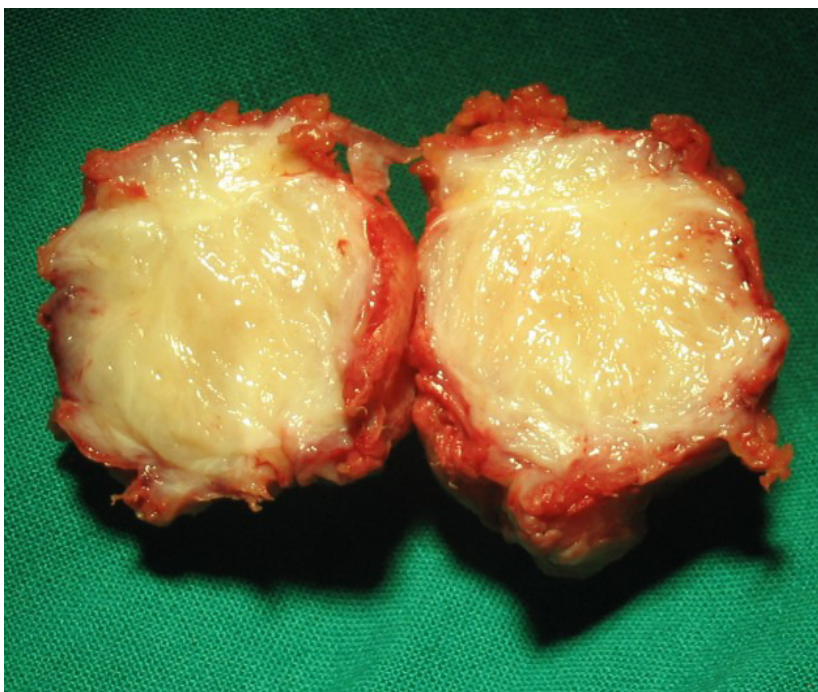


Figure 3 – Removed neurofibroma.

Histological examination confirmed the diagnosis (Figure 4).

There were no complications in the postoperative period. The patient was released from the hospital on the 6th postoperative day. After postoperative physiotherapy he was able to resume walking.

No local relapse of the tumor at follow-up 18 month after surgery was noticed clinically and on control MRI.

Unfortunately the patient was lost from our evidence because the family emigrated to a foreign country.

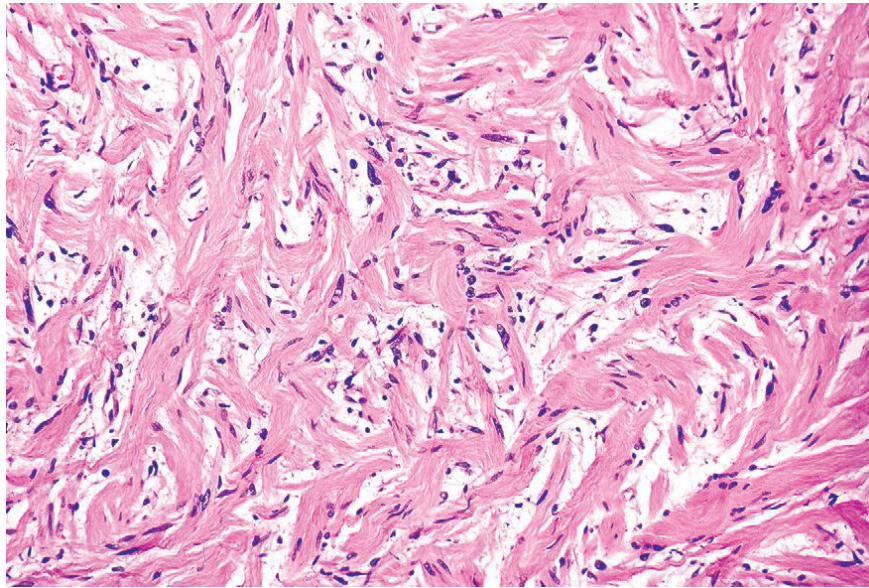


Figure 4 – Histological aspect.

Discussions

A neurofibroma is a benign tumor of the connective tissue, specifically the endoneurium, of peripheral nerves [1]. It is generally associated with neurofibromatosis type I; however, it can occur in isolation, in which case it is called a solitary neurofibroma. This tumor is rare, unlike

a schwannoma, which is the most frequent tumor of the peripheral nerves; the prevalence of schwannomas reached 55 % of all tumors in some series [2]. Both tumors result from the nerve sheath, which in turn originates from the neuroectoderm and neural crest. The distinction between a schwannoma and neurofibroma is important. Neurofibromas deeply invade the nerve, and total resection often has functional consequences; in contrast, schwannomas arise from Schwann cells and form a macroscopically smooth, rounded, yellowish, encapsulated proliferation, the enucleation of which is easy and without loss of continuity [1].

In addition to peripheral neurofibromas, patients with NF1 also develop central nervous system tumors, including optic nerve glioma, astrocytoma, and a variety of heterotopias. Vestibular

schwannoma, the hallmark of NF2, is virtually never encountered in NF1.

Ultrasound can help in detecting and diagnosing nerve tumors. In their classic forms, peripheral nerve tumors are well-defined, round, oval or spindle hypoechoic masses [3] as eccentric masses with a homogeneous structure, acoustic posterior enhancement, intratumoral cystic modifications, and intratumoral vascularity on color Doppler examination. In contrast, neurofibromas appear echogenic and hypovascular on Doppler examination [3,4,5]. MRI is a gold standard for evaluating nerve tumors.

The signal intensity of a normal nerve on MRI is of intermediate to low on T1-weighted sequences becoming

slightly higher on T2-weighted and other fluid-sensitive sequences.

In entrapment syndromes of peripheral nerves hyperintense signal of the denervated muscle

is usually identified when entrapment is acute, and fatty infiltration and muscle atrophy are the signs of chronic neuropathy in longstanding cases [6,7].

In most cases, MRI permits differentiation between neurofibromas and schwannomas. A rounded appearance, peripheral location, and more or less homogeneous, central low signal intensity on T2-weighted images are the characteristics of schwannomas; neurofibromas are usually heterogeneous on both T1 and T2 images [8,9].

Plexiform neurofibromas as well as neurofibromas of larger nerves have a significantly higher risk of malignancy than solitary neurofibromas of smaller nerves [10,11]. This is why it is important to have an exact diagnosis, which sometimes ultimately requires surgery and histopathological examination as in the case of our patient.

MRI following the administration of intravenous contrast materials improves contrast resolution in evaluation of soft-tissue tumors [10]. Imaging following contrast is particularly important with masses that have high water content or are composed of prominent necrotic/hemorrhagic foci allowing identification and differentiation of these regions from enhancing solid cellular tissue [12].

Surgical treatment for the nerve sheath tumors such as schwannoma or neurofibroma is primarily reserved for the symptomatic tumors.

McMillan et al. have reported 7 cases of sciatic neuropathy due to tumor [13]. In 3 children, this was due to infiltration of the nerve by sarcomas external to the nerve, an osteosarcoma of the femur in 2 patients and a rhabdomyosarcoma of the pelvis in one child.

Venna et al. reported a case of a child with unilateral chronic progressive sciatic nerve dysfunction found to be due to nerve entrapment in the thigh by a fibrovascular band. Sectioning of the band was followed by marked improvement in the nerve function. They concluded that compression by a band is a rare but treatable cause of sciatic neuropathy [14].

Conclusions

Sciatic neuropathy is an uncommon mononeuropathy in children and the causes of sciatic neuropathy are varied and

unique. Peripheral nerve tumors of the sciatic nerve is a rare condition. Especially, primary tumors of the sciatic nerve are rare. A palpable mass is usually the only presenting symptom of peripheral nerve tumors. Other clinical symptoms include radicular pain, paresthesia, hypoesthesia and motor deficiencies. Sciatic neuropathy due to neurofibroma of the thigh in a child 5 years of age is also a very rare condition, which to the best of our knowledge has never been described in the medical literature.

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