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Case report

Clinical case: giant solitary neurofibroma of the thigh

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Abstract

The solitary neurofibroma, a benign ectodermal tumor reaching the nerve sheaths, is very rare both in its frequency and its location. Its diagnosis is histo-immunological and its treatment, surgical, consists of a complete resection of the mass due to a degenerative risk tumor. The authors report the case of a 36-year-old patient, admitted for a large tumor of the anterior aspect of the right thigh. Ultrasound and CT revealed the presence of a soft tissue tumor measuring 28 x 15 cm. The resection was performed without damage to the adjacent noble elements. The tumor weighed 7 kg. The histology found a neurofibroma. Benign tumors of the nerve sheath are rare. The neurofibroma is a benign tumor, which can occur in two forms: solitary in young adults or multiple in the context of a Recklinghausen disease. The majority of cases of giant neurofibromatous tumors reported, were almost always isolated localization. TDM or echo-guided biopsy is the only way to confirm preoperative histological diagnosis. The treatment is surgical. However, the tumor infiltrates the fasciculi of the nerve along its course, which could make its resection difficult and dangerous. Thigh localization of the neurofibroma is rare. Complete excision with negative margins is the treatment of choice. In the case reported, one year after surgery, the patient was in good general condition with normal locomotor function.

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Introduction

Benign nerve sheath tumors in patients with or without neurofibromatosis type 1 are very rare [1-3]. They include neurofibromas and schwannomas. We report a case of solitary giant neurofibroma of the thigh. It is a benign tumor that develops in and along the nerves and nerve sheaths. Its presence is a cardinal sign of neurofibromatosis type 1 or Von Recklinghausen's disease. We will discuss its diagnostic, therapeutic and evolutionary aspects.

Patient and observation

Mrs. O.A, aged 35, consulted in May 2016 for a voluminous tumor of the right thigh, shiny, painless, with no other functional sign, progressively evolving over the last ten years. The clinical examination found a patient in good general condition, apyretic. Hisblood pressure was 130/80 mmHg. Neurological assessment and skin examination were normal. The anterior part of the right thigh was occupied by a voluminous, shining mass, extending from the inguinal region to the knee. It was ovoid in shape, measuring 35 cm long (Figure 1). The rest of the somatic examination was normal; in particular there was no adenopathy. The biological check-up was normal. The standard radiograph of the femur showed no bone abnormalities. Ultrasound had discovered a voluminous echogenic mass, poorly limited, more than 30 cm in diameter. MRI showed a subcutaneous oblong tissue mass in hyposignal T1 and hypersignal T2. An echo-guided trans-parietal biopsy with histological analysis allowed the diagnosis of neurofibroma to be made.

By an anterior vertical approach of the thigh, evacuating punctures of the areas of necrosis have been carried out in order to reduce hypertension of the tumor. Then a gradual dissection allowed to enucleate the tumor which molded around the femur without adhering to it. The Quadriceps was laminated, fibrillar and the bone was widely exposed (Figure 2). The macroscopic study of the mass found a whitish tumor, sometimes firm, sometimes soft, and discretely fasciculated, with necrotic-haemorrhagic changes, measuring 33 x 20 x 17 cm and weighing 7 kgs.

In histology, the tumor was composed of intersecting beams of fusiform cells with dark and undulating nuclei, mixed with collagen bands. The cells did not exhibit mitotic activity. On immunohistochemical examination, the tumor cells expressed the S100 protein, concluding with a neurofibroma. The postoperative follow-up was simple. No additional treatment was given. At one and a half years of the intervention, no recurrence was noted. The patient was autonomous with moderate stiffness of the knee (95° flexion) and amyotrophy of the thigh with a muscular force rated at 3/5 (Figure 3).

Discussion

Benign tumors from the nerve sheaths (neurofibroma, benign schwannoma) are rare ectodermal tumors [2-5], affecting mainly the cutaneous nerves of the extremities, the nape and the trunk, which can sometimes be part of Von Recklinghausen's disease [6-9]. Neurogenic tumors can develop in all parts of the body: thorax, retroperitoneum, sinus, maxilla, limbs [1, 8, 10, 11]. In the majority of cases of giant neurofibromatous tumors reported, localization was almost always isolated. According to the study by Bories-Azeau and Guivarc'h [2], ectodermal tumors are more frequent in children (56%), whereas they are only seen in 12% in adults. However, the distribution of histological types is different according to age. In adults, nerve sheath tumors are the most frequent, whereas those from the sympathetic (ganlioneurome, ganglioneuroblastoma) or parasympathetic (paraganglioma) are seen more in children. Neurofibroma is a benign tumor that can occur in two forms: solitary encountered in young adults [3, 10-12] (our observation) and the multiple form associated with neurofibromatosis type 1 or Von Recklinghaus disease.

On the anatomo-pathological level [1, 11-15], the neurofibroma constitutes a non-encapsulated tumor encompassing the entire nerve. The tumor infiltrates the fascicles of the nerve along its course, which could make its resection difficult and dangerous. In our observation the nerve involved was probably the lateral cutaneous nerve of the thigh. We did not find it and it was probably resected with the tumor. Preoperative diagnosis of neurofibroma is difficult. Indeed, due to a fat overload of Schwann cells or a transformation of fibroblasts into adipocytes, the appearance of the tumor can orient towards a lipomatous origin. The differential histological diagnosis [1, 3, 11, 15] is presented with benign schwannoma, which is an encapsulated tumor, made exclusively of Schwann cells, whose proliferation takes place in the endon-fever of a nervous fascicle. It contains no nerve fiber.

Benign tumors of the nerve sheathare clinically silent in almost 60% of cases and the clinical signs, of compressive type, of the adjacent structures, are the fact of the masses of large dimension. They manifest themselves in deaf pains, sometimes in the form of gravity [1, 10, 15]. The coexistence of a neurofibromatosis, often of type 1, directs the diagnosis, especially if the masses are multiple. Usually, the neurofibroma appears, in ultrasound [15, 16], hypoechoic with posterior reinforcement. Combined MRI and CT scans play a key role in the diagnosis of benign tumors of the nerve sheath [7, 10, 16]. The correlation between anatomopathology and imaging should allow a better histological specificity of the imaging, although the difference between a benign tumor and a malignant tumor is still tainted by uncertainties [1, 7, 14, 16]. Transparietal biopsy, echo or TDM guided is the only means of preoperative histological diagnosis. But some authors advise against it because of the cellular pleiomorphism of tumors of large dimensions [1, 8, 10, 11], as was the case of our patient. In the isolated neurofibroma, surgery is the first-line treatment to be implemented given the risk of recurrence and malignant transformation. The approach must allow an accurate assessment in case of multiple neurofibromas [15] and easy control of large vessels. The complete excision of the tumor with extemporaneous examination of the sectional slices is the treatment of choice [1, 2, 8, 11,13]. It can sometimes lead to neurosensory deficits.

Conclusion

Neurofibromas are ectodermal tumors with rare thigh localization and histological diagnosis. They are clinically mute and can take on important dimensions. Imaging helps with topographic assessment and the lack of reliability of the biopsy, given the difficulties of histological interpretation, makes it of little use. Complete excision with negative margins is the treatment of choice.

Competing interests

The authors declare no competing interests.

Authors' contributions

Dr Salam Ouedraogo was the second surgeon during the operation. Dr Alassane Drave, neurologist physician assisted in diagnosing neurofibroma. All other authors contributed to the writing and proofreading of the manuscript. All authors have read and agreed to the final manuscript.

Figures

Figure 1: The tumor
Figure 2: Tumor molded around the femur
Figure 3: Patient at 3 months postoperative

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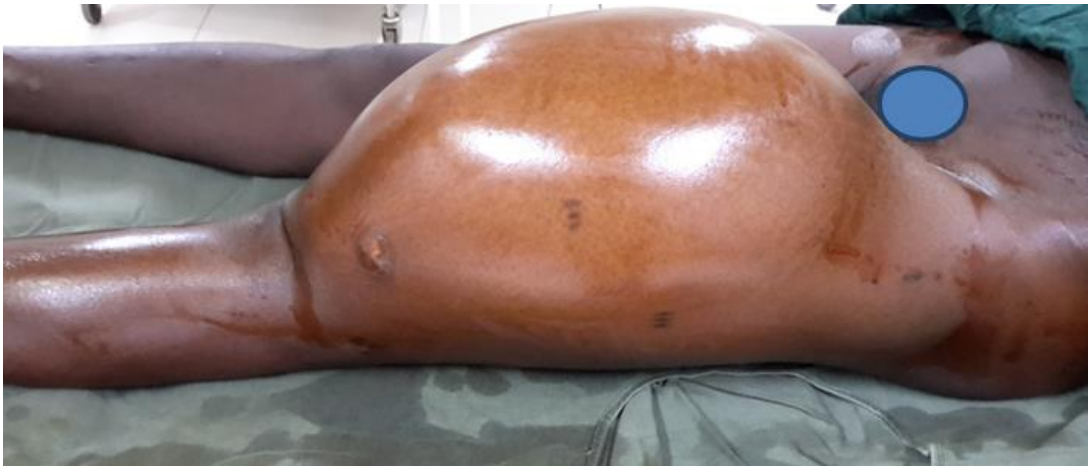

Figure 1: The tumor

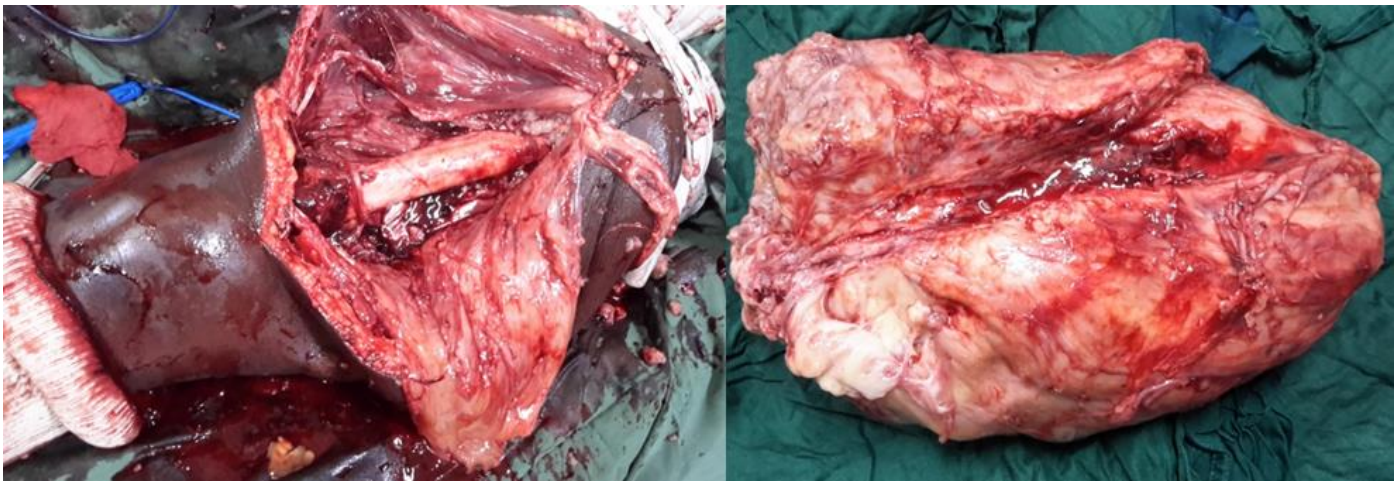

Figure 2: Tumor molded around the femur

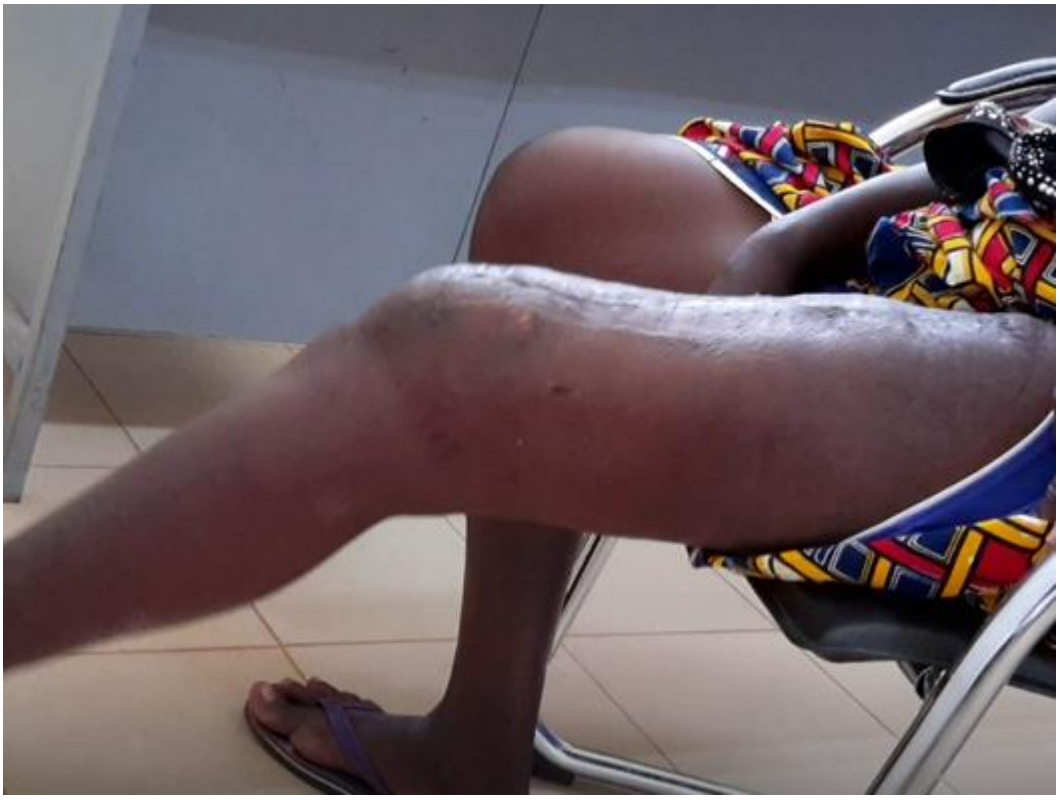

Figure 3: Patient at 3 months postoperative