

**Neurofibroma of the hand
(case report)****Neurofibrome de la main
(cas clinique)**

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RESUME

Nous rapportons un cas de neurofibrome sporadique du 5ème doigt de la main droite chez un militaire âgé de 41 ans. La localisation au niveau de la main d'un neurofibrome isolé est rare et pose des problèmes à la fois diagnostiques et thérapeutiques pour le chirurgien. La diversité de la présentation clinique peut rendre le diagnostic difficile en préopératoire. La contribution de l'imagerie reste non claire et le diagnostic définitif n'est souvent fait que par l'anatomo-pathologiste. Le traitement chirurgical du neurofibrome est complexe et controversé; un équilibre doit être trouvé entre la résection complète et le risque de morbidité.

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MOTS-CLÉS : main; neurofibrome;
diagnostic; résection chirurgicale

ABSTRACT

We report the case of a sporadic neurofibroma arising in the right fifth finger of a 41 year old military. Isolated neurofibroma in the hand are rare and pose both diagnostic and treatment challenges for the surgeon. The variation in clinical presentation may make preoperative diagnosis difficult, the contribution of preoperative imaging remains unclear and the final diagnosis may often lie in the hands of the pathologist. The surgical treatment of neurofibromas is demanding and controversial, and a balance must be found between complete resection and significant morbidity.

Keywords: Hand tumor; neurofibroma;
diagnosis; imagery; surgical resection.

INTRODUCTION

Neurofibromas are uncommon in the hand [1,2]. Although making an accurate preoperative diagnosis of neurofibroma is usually difficult, it is arguably more important to do so than it is for other benign tumours because of the postoperative problems [3]. Furthermore, The surgical treatment of neurofibromas is demanding and controversial.

CASE REPORT

A 41 year old man, right hand dominant,

soldier, presented with a nodule in the volar face of the proximal phalanx of the right fifth finger. The nodule had been present for the past 4 months and was growing in size with dysesthesia along the fifth finger. He was otherwise medically fit and well, took no regular medications and was a non smoker.

Surgical exploration revealed a limited swelling at the digital nerve from the ulnar nerve (fig. 1). However we bear in mind the existing dysesthesia, an attempt was made to dissect the nerve from the tumour. The lesion was carefully dissected free from both digital nerve and soft tissue attachments and excised with loupe magnification (fig. 2).

Histological examination showed cells with small, elongated, wavy nuclei with regular chromatin and indistinctive cytoplasm. The stroma is loose, it consists of eosinophilic, shredded collagen bundles and it is punctuated occasionally with mononuclear cells. A diagnosis of neurofibroma was made (fig. 3).

Postoperatively, the patient recovered well, and the surgical wound fully healed after two weeks. The patient developed hypoesthesia in the cutaneous distribution of the digital nerve which persisted for 6 weeks. Normal sensation returned later. There has been no recurrence 1 year postoperatively.



Fig 1: Neurofibroma appears as a limited swelling at the digital nerve from the ulnar nerve.



Fig 2: Dissection of the digital nerve free from the surrounding tumor.

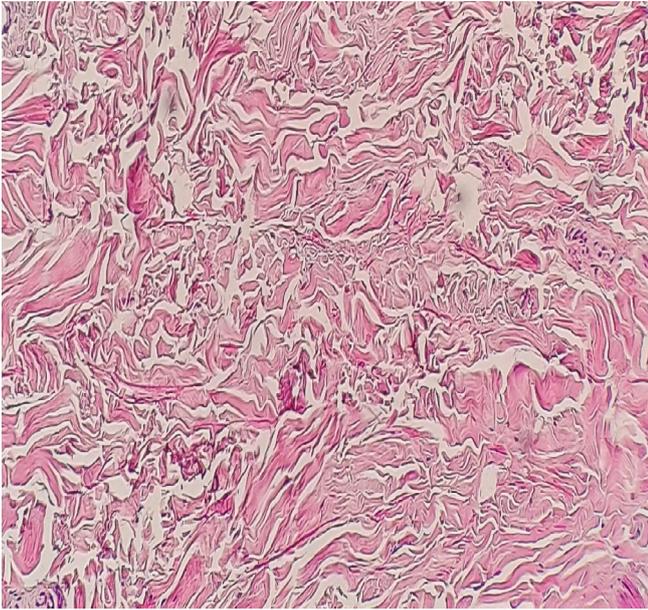


Fig 3: Cells with small, elongated, wavy nuclei with indistinct cytoplasm in a background of eosinophilic shredded collagen.

DISCUSSION

Neurofibromas in the hand are rare, and reports in the literature are scanty. Bogumill et al. [1] reported one case of digital nerve neurofibroma out of 129 tumours. Strickland and Steichen reported one out of six tumours of nerve origin, which constituted 0.8% of hand and arm tumours [4]. Rinaldi reported one neurofibroma in a digital nerve and one in an ulnar sensory branch [5]. Neurofibromas are normally slow growing over many years. Whilst the reported history in the case described is shorter, it is likely the lesion had grown unnoticed for years previously.

Neurofibromas arise from the connective tissue of the nerve sheath and consist of Schwann cells, fibroblasts, and cells with perineural properties with occasional residual nerve fibers [6]. The majority of neurofibromas occur sporadically, but multiple tumors or specific morphologic variants can be associated with genetic

syndromes, the most common of which is the neurofibromatosis type 1 NF1 (von Recklinghausen disease). NF 1 is reported to occur in 10 to 15% of cases [2,4]. It is usually associated with cutaneous neurofibromatosis, café-au-lait spots, optic glioma, specific bone lesions such as pseudarthrosis or sphenoid dysplasia, lipomas, sebaceous adenomas, intestinal tumours, and plexiform nerve lengthening [7]. The gene associated with NF1 is a tumor suppressor called neurofibromin 1 and resides on chromosome 17q11.2. Loss of neurofibromin 1 function leads to the development of both sporadic and NF1-associated neurofibromas, but neurofibromas can gain additional mutations and give rise to malignant peripheral nerve sheath tumors MPNSTs [8].

Neurofibromas may present as a diffuse thickening of the nerve trunk, due to overgrowth of the endoneurium along the length of a nerve, as a localized overgrowth affecting branches of a single nerve (plexiform neuroma), as a cutaneous lesion, or as a solitary lesion, which forms an encapsulated tumour adjacent to a nerve.

Some investigators, have described, like our case, a clinical picture of a painful, firm, solitary nodule, while other cases have been entirely asymptomatic. Review of the literature reveals no consistent histological difference between symptomatic and asymptomatic lesions. The variation in clinical presentation may make preoperative diagnosis difficult and the final diagnosis may lie in the hands of the pathologist. In a series of 208 soft tissue tumours of the hand and forearm by Lincoski et al., only one case of the 24 nerve sheath tumours reported had a correct preoperative diagnosis and 4 (33.3%) of the 12 patients with neurofibromas had neurologic symptoms [9]. Then, the diagnosis of neurofibroma should be kept in mind when approaching any hand

tumor, so that the patient may be warned about the possibility of residual numbness.

The role of imagery in the diagnostic of neurofibromas is unclear. Ultrasound, whilst accurately demonstrating the morphology and location of a soft tissue mass, is likewise limited in terms of specific diagnosis [10]. Neurofibromas are hypodense on CT scan, whereas on MRI, they present as hypointense on T1-weighted images and hyperintense on T2-weighted images with increased contrast uptake. T2 MRI sequence images show a target sign that represents a dense collagen core [11]. Indeed, MRI gives accurate information regarding the anatomical relationships of the tumour and is thus helpful for surgical planning, but is often not diagnostic. Moreover, MRI does not always provide 100 % accuracy in delineating benign from malignant masses [12].

Diagnostic biopsy may be considered for a definitive diagnosis. In this case the diagnostic biopsy was not performed prior to surgery, as the decision had been made to excise the mass due to it causing functional impairment. Microscopically, neurofibromas appear as hypocellular arrangements of cells showing small, elongated, wavy nuclei with indistinctive cytoplasm in a background of eosinophilic, shredded collagen [13]. Mast cells are commonly seen within these lesions but do not have prognostic implications. All neurofibromas demonstrate immunohistochemical reactivity for S100 (Schwann cells), although the extent of staining varies depending on the proportion of accompanying reactive cells (S100 negative). This is in contrast to the uniform S100 immunoreactivity in schwannomas [14]. Neurofibromas lack EMA staining due to the absence of true perineural cells within the center of the lesion. However, approximately 50% of all neurofibromas will show neuronal staining as they can contain a varying percentage

of entrapped neurons and axons [8].

The surgical treatment of neurofibromas is demanding and controversial. Neurofibromas can be cured by total surgical resection because they lack the ability to metastasize, but resection may be difficult because of their anatomic location and may lead to significant morbidity because of damage to associated nerves.

Wide surgical exposure is required to fully visualize the tumor. Most neurofibromas, are intimately associated with nerve substance and require some sacrifice of fascicle integrity for complete removal. This is in contrast to other benign nerve tumors, such as neurilemmomas, which can often be shelled out of the nerve without fascicular sacrifice. Even with the correct diagnosis, treatment decisions must still be judiciously considered. Like our case, well circumscribed, localised neurofibroma can often be carefully excised with minimal morbidity. Fascicles involved with the neurofibroma require meticulous dissection under loupe or microscope magnification in a bloodless field to preserve maximal nerve continuity and postoperative sensibility [15]. The true involvement of the affected nerve may only become apparent at surgery [16].

CONCLUSION

Neurofibromas are uncommon in the hand. The majority of neurofibromas occur sporadically, but multiple tumors or specific morphologic variants can be associated with genetic syndromes. The variation in clinical presentation may make preoperative diagnosis difficult and the role of imagery in the diagnostic of neurofibromas is unclear. Then, the diagnosis of neurofibroma should be kept in mind when approaching any hand tumor. Diagnostic biopsy may be considered for a definitive diagnosis. The

surgical treatment of neurofibromas is demanding and controversial, and a balance must be found between complete resection and significant morbidity.

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