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(CASE REPORT)

Unusual localization of a digital neurofibroma: About a case and review of the literature

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Abstract

Neurofibromas are benign tumors of peri-nervous origin. may occur as a single lesion (solitary neurofibroma) or as part of a neurofibromatosis type 1. We present a case of a 44-year-old man with a painful neurofibroma on the dorsal surface of the 3rd right finger, revealed by a painful mass on its posterolateral aspect. Treatment was by excisional biopsy, with histopathological examination confirming the diagnosis. In the study by Lincoski et al, 24 benign nerve tumours accounted for 11.5% of the total number of soft-tissue tumours in the hand and forearm. Isolated neurofibromas are rare in the hand and normally grow slowly over many years. MRI provides precise information on the anatomical relationships of the tumour. Even with a correct diagnosis, the therapeutic decision must be carefully considered.

Keywords: Neurofibroma; Digital; Benign tumors; Case

1. Introduction

Neurofibromas are benign tumors of peri-nervous origin, most commonly localized on the face, shoulders, arms and peri-ungual regions of the feet [1, 2].

Neurofibromas may occur as a single lesion (solitary neurofibroma) or as part of a neurofibromatosis type 1.

A dozen or so publications have been published on this benign tumor, because it is an uncommon lesion that predominates in young adults around the age of 40, but can occur at any age without gender predominance [3, 4].

Treatment is indicated for functional and/or cosmetic reasons, and is essentially surgical [5, 7].

Isolated neurofibroma in the hand are rare and pose both diagnostic and treatment challenges for the surgeon. Imaging can be very useful to guide the diagnosis as well as determining its relationship with the surrounding soft tissues.

The aim of this paper is to present a rare case of a solitary neurofibroma of the dorsal surface of the 3rd right finger.

2. Clinical case

A 44-year-old woman with no specific pathological history presented with a painful mass on the dorsal surface of the 3rd right finger with paresthesia that had been present for 9 months.

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Clinical examination revealed a mass on the posterolateral aspect of the 3rd right finger, opposite P2 on the right hand, with a firm consistency, painful to palpation, 1.5 cm long, fixed in the deep plane and mobile in relation to the superficial plane.

The swelling was interfering with his grip of tools and his daily routine work. There was no neurological deficit.

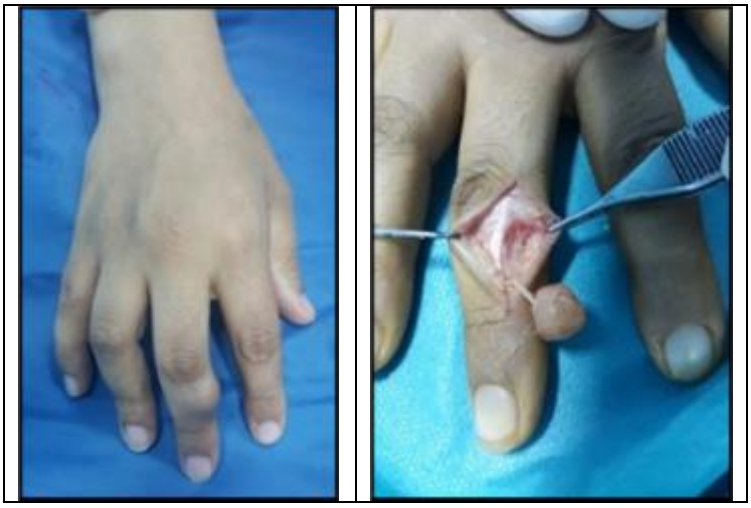

Figure 1 Clinical appearance of the tumor opposite the posterolateral aspect of P2 on the 3rd finger of the right hand

Radiography of the hand revealed no associated bone lesions, but Ultrasonography revealed a well-limited hypoechoic mass opposite P2 measuring 1.5 cm, with no associated signs of malignancy.

Figure 2 X-ray of the right hand, showing no bone reaction to the mass

Treatment was by excisional biopsy, with histopathological examination confirming the diagnosis of a Neurofibroma, and the evolution was favorable at 2-year follow-up without recurrence.

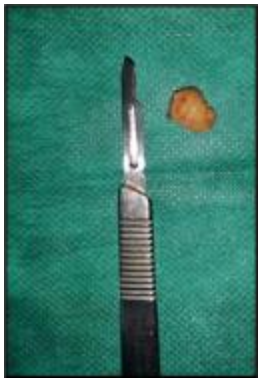

Figure 3 Intraoperative appearance and size of the tumor mass

3. **Discussion**

Neurofibromas are benign nerve tumors of neuro-ectodermal origin. They are classified into localized neurofibromas, which originate from a single fascicle, and plexiform neurofibromas, which originate from several fascicles that usually develop in the nerve trunk [6].

Neurofibromas are often found as part of neurofibromatosis type I (Von-Recklinghausen disease), an autosomal dominant disorder characterized by multiple neurofibromas and café-au-lait spots [6].

Neuronal hand tumours are rare in clinical practice. According to numerous articles, they account for a small percentage of soft tissue tumors of the hand [1-5].

Our patient has presented a rare localization of neurofibroma. in the study by Lincoski et al [3], 24 benign nerve tumours accounted for 11.5% of the total number of soft-tissue tumours in the hand and forearm (208 tumours excised). hand and forearm (208 tumours excised over a 16-year period). Kehoe et al [7] reported 104 cases of benign nerve tumours, 25 cases of schwannomas and only 3 cases of neurofibromas in the hand and wrist. In the series by Goski et al [1], neurofibromas were relatively evenly distributed in the palm and fingers, while schwannomas were located downstream of the metacarpophalangeal joints.

Isolated neurofibromas are rare in the hand and normally grow slowly over many years [8]. Although our patient's disease history dates back only 9 months, it is likely that the lesion developed unnoticed for years.

Clinically, neurofibromas are characterized by a well-circumscribed, firm, often solitary or sometimes bilobed, painless nodule contrasting with the painful mass in our case, averaging 2 cm in size (0.5 to 10 cm), with a slow evolution (median evolution of three years), located in the skin (dermis), subcutaneous (extremities) or deep (soft tissues, mammary gland, orbit, etc.). Palmar involvement of the fingers is exceptional [10].

Diagnosis is based on the presence of a collagenous component made up of thick, sinuous, short fibers, cells with curved oval nuclei, associated with mast cells and lymphocytes, sometimes with aspects of specific differentiation. Histopathological and immunohistochemical studies support the diagnosis [5].

Pain, neurological symptoms, enlargement and/or functional discomfort are the main indications for surgical treatment. However, the majority of benign nerve tumors are asymptomatic.

Diagnostic biopsy may be considered for a definitive diagnosis. In our case the diagnostic biopsy was not performed prior to surgery, as the decision had been made to excise the mass due to its benign aspect and its functional impairment.

Ultrasonography is a non-invasive examination that is capable of providing basic information about the number and size of tumours. It is definitely the first-line imaging examination in the diagnostics of peripheral nerve tumours.

Ultrasound can show the morphology and location of soft-tissue tumours with great precision, but remains limited in terms of specificity [6].

MRI was not done by our patient because of the sufficiency of the ultrasound, and also due the its high cost. However when asked, MRI provides precise information on the anatomical relationships of the tumour, and therefore plays an important role in preoperative planning.

Neurogenic tumours usually show enhancement on T1-weighted images after intravenous administration of the contrast agent. Certain characteristic features of neurofibromas can be observed on MRI. These include fat split sign (on T1- weighted images), dural tail sign, target sign and fascicular sign (on T2-weighted images).

Even with a correct diagnosis, the therapeutic decision must be carefully considered. Isolated, well-localized neurofibromas are usually easily resectable without sequelae. However, resection of a plexiform neurofibroma cannot be performed without resection of the incriminating nerve, which is sometimes only visible intraoperatively [4,9,11].

In the preoperative period,, some features such as tumour size above 5 cm and the history of fast tumour growth can indicate the malignant character of the lesion. [11, 12]. However these characteristics were absent in our patient, which motivated a direct tumor resection.

4. Conclusion

A benign tumor of rare peri-nervous origin in young adults, it is usually solitary, although it may be of multiple localization or reappear after an initial incomplete excision. It should be considered in the differential diagnosis of an extremity tumor.

Compliance with ethical standards

Disclosure of conflict of interest

The authors declare no conflict of interest in relation to the writing of this this article.

Statement of ethical approval

Ethical approval for this study was obtained from the Institutional Ethics Committee

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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