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Benign Neurofibroma of a Digital Nerve in A 21-Year-Old Basketball Player: A Case Report

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1. Abstract

Although ganglion cysts are a common type of benign soft tissue tumours in the hand, primary neural tumours in the hand are rare. Typically, benign nerve tumours in the hand present as neurilemomas, while neurofibromas are even rarer. Patients usually present with a painless mass, and specific treatment is not always necessary due to their benign nature. However, if the patient experiences pain during daily activities, surgical excision may be recommended. This almost always results in complete removal of the lesion and improvement of symptoms, with recurrence being uncommon. This article describes the case of a 21-year-old male who presented with a nodular subcutaneous swelling on the palmar ulnar side of the fourth digit of his right hand. The patient underwent operative excision, and the diagnosis was a neurofibroma. The patient experienced complete resolution of mechanical symptoms and full range of motion at the four-week follow-up.

2. Introduction

Whilst benign soft tissue tumours of the hand and fingers (i.e., ganglion cysts or mucous cysts) are relatively common, primary neural tumours of the hand, both benign and malignant, are rare [1-5]. Neurilemoma, sometimes called Schwannoma, is the most common benign nerve tumour of the upper extremity, arising from the Schwann cells to produce a slow growing, well-circumscribed, eccentric lesion in the peripheral nerve. Though common on the flexor surface of the hand and forearm, occurrence in a proper digital nerve appears to be exceptional with only a handful of case reports documenting this phenomenon [1-3,5-10].Related, neurofibromas are similarly benign nerve tumours arising from Schwann cells within the nerve fasciculi but are less common and proving to be more difficult to excise than classic neurilemomas. They are most commonly related to neurofibromatosis type 1, also known as von Recklinghausen disease, usually presenting with multiple lesions, though solitary lesions have been reported. To our knowledge, no case report has described a patient presenting with a solitary lesion of a digital nerve neurofibroma [1,2,6].Clinical findings for both tumours are similar. Patients usually present with a painless mass, relatively uncommonly associated with a neurologic deficit. Palpation or compression of the lesion can produce sharp, radiating pain in a specific nerve distribution. The mass is often mobile in a transverse direction but not longitudinally and can frequently be misdiagnosed as a ganglion cyst. MRI can be useful to distinguish primary nerve tumours from other soft tissue tumours such as ganglion cysts or mucous cysts but at times it may not be possible to distinguish neurilemoma from a neurofibroma or malignant peripheral nerve sheath tumour (MPNST) [1-5].Specific treatment is not always necessary for either neurilemomas or neurofibromas due to their benign nature and slow growth. Given their location on the volar aspect of the hand and forearm, however, they are often painful during daily activities, justifying surgical treatment. The preferred procedure is intralesional excision under magnification. Postoperative neurologic deficit is reported between 5 and 15% for neurilemomas and is reported to be more common for neurofibromas though no precise values are reported in literature. Surgical excision almost always results in a complete removal of the lesion with improvement of the preoperative symptoms and recurrence is reported to be uncommon [1-6,8].

3. Case Report

A 21-year-old right-handed male with no medical history presented with a swelling of the fourth digit of the right hand. There was no notion of acute trauma, but the patient did report playing basketball as a hobby, suggesting some repetitive mechanical stress to the hands and fingers. He reported sudden onset radiating pain of the node and finger following a normal movement of the finger. There was intermittent focal pain ever since, especially after palpation of the swelling, during basketball and when shaking hands. He was not a smoker. Clinical examination showed a nodular subcutaneous swelling of the palmar ulnar side of the fourth digit of the right hand just proximal to the proximal interphalangeal joint (PIP) crease. This swelling was mobile in the axial plane. The patient reported no sensory loss in the finger. There was a positive local Tinel's sign but no radiating pain distal to the swelling. Mobility of the metacarpophalangeal (MCP), PIP and distal interphalangeal (DIP) joints was normal, and no vascular

abnormalities were noted. Radiograph of the hand and fingers revealed no abnormalities. Ultrasound showed a soft-tissue tumour of the ulnar proper digital nerve of the fourth digit just proximal to the PIP (Figure 1). Finally, MRI confirmed this nerve tumour with dimensions 4.7*4.5*20.0mm. The lesion was slightly hyperintense on a classic T1-weighted image and on proton density fat saturated (PD fat sat) image with strong and heterogenous contrast enhancement (Figure 2). This was most suggestive for a benign neurilemoma of the proper digital nerve. Given the mechanical symptoms during daily life, operative excision of the tumour was performed. Under 5.0x magnifying glasses, the epineurium was longitudinally incised, and the tumour was excised intralesional with achievement of complete en bloc macroscopic excision (Figure 3). Macroscopic analysis showed a lobulated yellowish tumour. Further microscopic analysis showed a circumscribed lesion, surrounded by perineurium (EMA+). There was a proliferation of bland, neuroid spindle cells in an alternating myxoid-collagenous matrix, with typical 'roapy collagen'. Some scattered mast cells were found interstitially (Figure 4). This proved the lesion to be a neurofibroma rather than a neurilemoma.At 4 weeks follow-up, the wound had properly healed, and the patient reported resolution of the mechanical symptoms with full range of motion the MCP, PIP and DIP joint. No sensory loss was reported, and the patient was satisfied with this result.

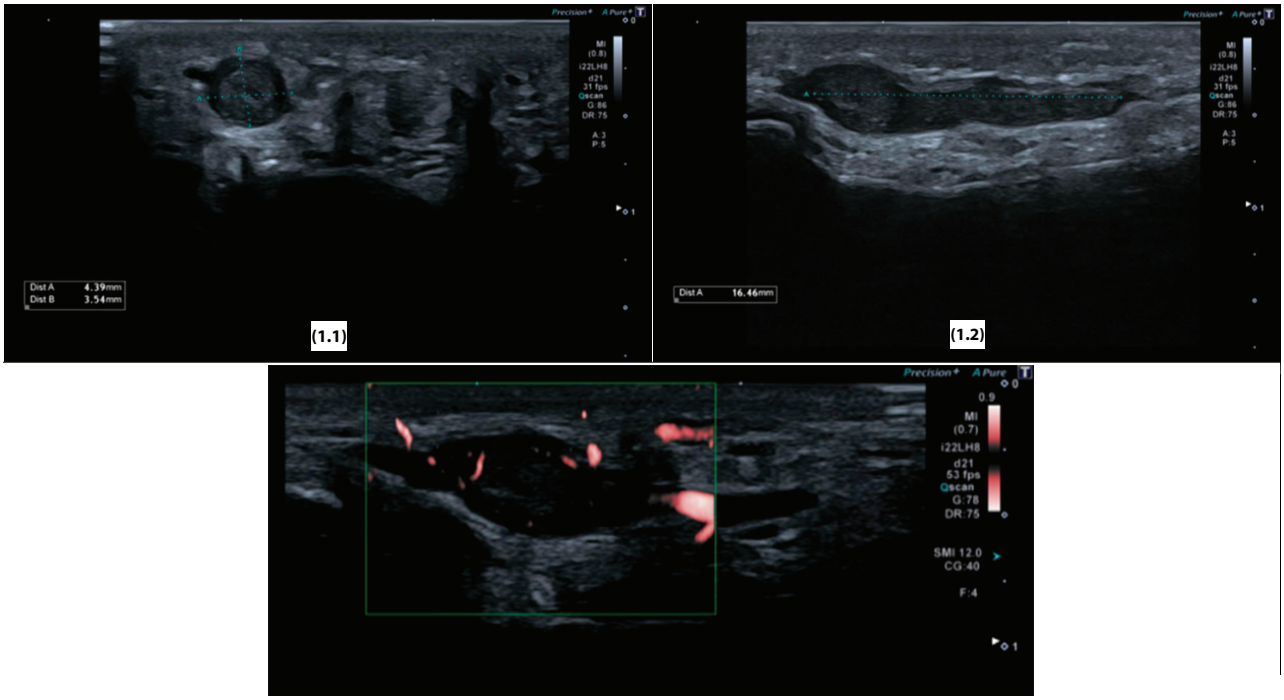

Figure 1: Ultrasound images showing a soft-tissue tumour of the ulnar proper digital nerve of the fourth digit just proximal to the PIP. Axial view (Figure 1.1) and longitudinal view (Figure 1.2) with duplex (Figure 1.3).

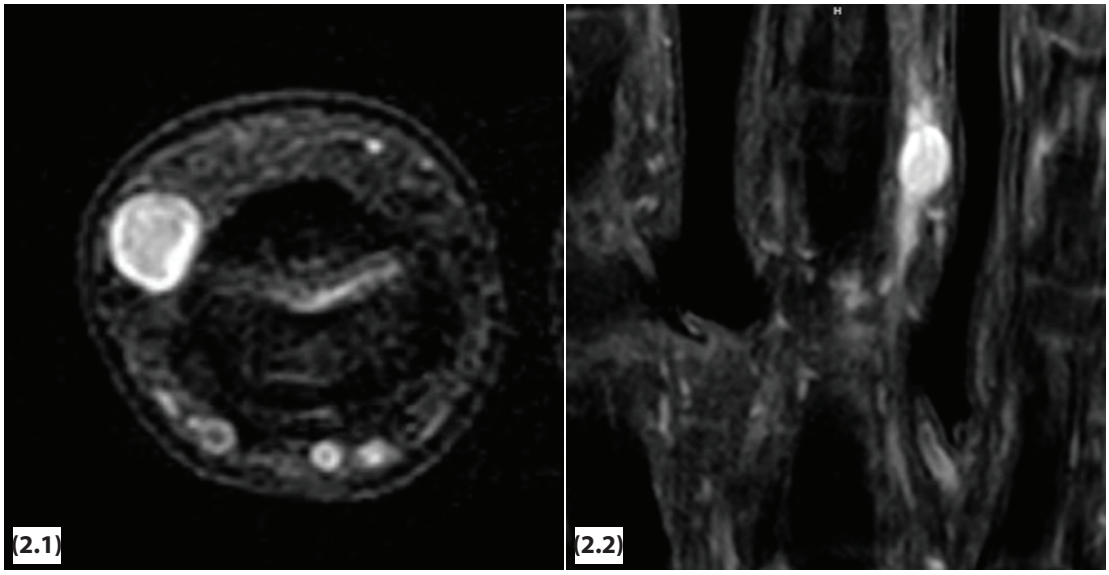

Figure 2: MRI images showing the nerve tumour with dimensions 4,7*4,5*20,0mm. The lesion was slightly hyperintense on proton density fat saturated (PD fat sat) image with strong and heterogenous contrast enhancement. Axial view (Figure 2.1) and coronal view (Figure 2.2).

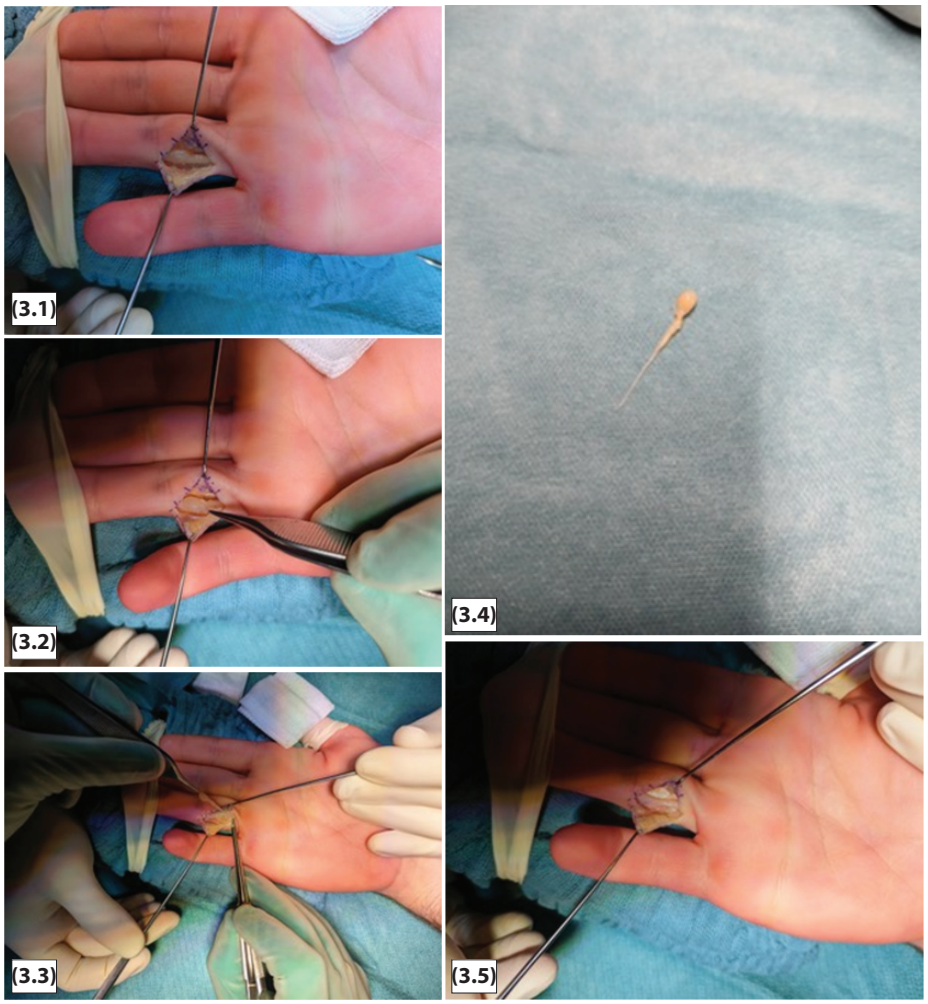

Figure 3: Clinical intra-operative photo's showing the lesion in situ (Figure 3.1 and 3.2), during dissection (Figure 3.3) and after removal on a surgical drape (Figure 3.4). Figure 3.5 shows the ulnar digital nerve after excision of the tumour.

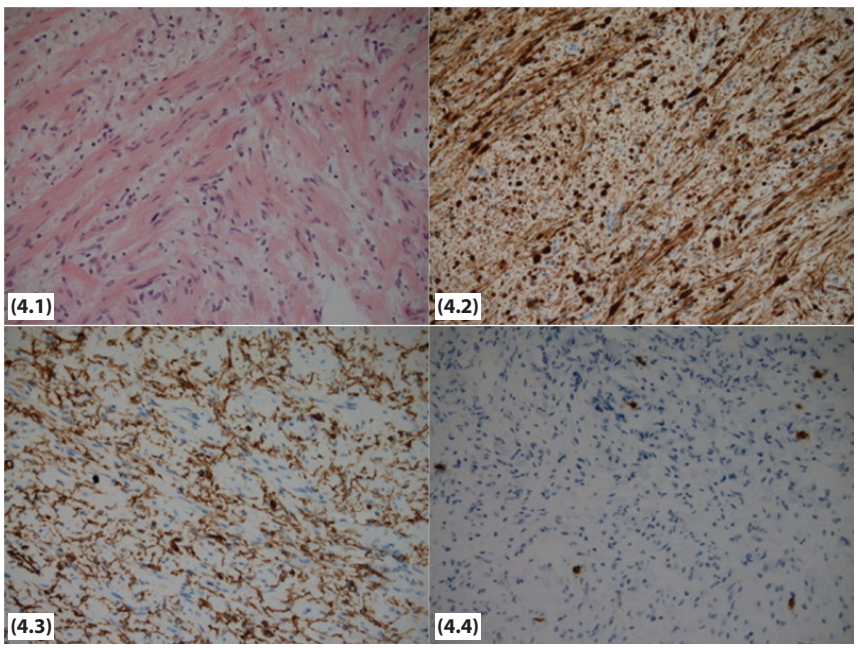

Figure 4: Figure 4.1 shows HE stain with bland spindle cell proliferation with alternating myxoid matrix and roapy collagen (original magnification x200). Figure 4.2 shows S100 strongly positive (nuclear and cytoplasmic) (original magnification x200). Figure 4.3 shows CD34 strongly positive (original magnification x200). Figure 4.4 shows CD117 showing some scattered interstitial mast cells (original magnification x200).

4. Discussion

Benign peripheral nerve sheath tumours (BPNST) are usually slow growing, well-circumscribed lesions in peripheral nerves and are more commonly found in the upper limb with the volar aspect of the forearm and hand as typical locations. These tumours should be distinguished from other benign soft tissue tumours (such as ganglion cysts, mucous cysts, giant cell tumours of the tendon sheath, fibromas of the tendon sheath, extraosseous chondromas…) and from MPNST. Primary neural tumours of the proper digital nerve are rare with only a handful of case reports describing neurilemomas of the proper digital nerve and to our knowledge no case report describing a neurofibroma of the proper digital nerve [1-6,8].A BPNST usually presents as a painless mass, relatively uncommonly associated with a neurologic deficit. Neurologic deficits in MPNST are usually due to the segmental demyelisation and not due to direct compression or lack of vascularity as shown by Das Gupta et al. [9]. Demyelisation is not found in neurilemomas nor neurofibromas. This in combination with their slow-growing nature is the most likely explanation for the lack of neurologic deficit in these tumours. The tumorous mass can, however, give local pain during daily activities, especially when they present on the volar aspect of the hand of digit. This can justify surgical treatment after discussing the risks and benefits with the patient [1-5].Surgical intralesional excision in line with the nerve fibres is the preferred method of treatment as reported in literature. Complication rates are low, but a postoperative neurologic deficit is reported between 5 and 15% for neurilemomas. This is reported to be higher for neurofibromas though no precise incidence is reported in literature. The deficit is often transient but exact values for this aspect are also lacking in literature [1-6,8]. In our case, the tumour was easily treated by surgical excision with good wound healing and no residual symptoms nor neurologic deficit at 4 weeks follow-up.

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