Nodular fasciitis involving the palm

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ABSTRACT

Nodular fasciitis (NF) is a self-limiting fibrous neoplasm that can be mistaken for a soft tissue sarcoma. It is characterised by rapid growth, slight pain and local tenderness. Although it is frequently found in the forearm, a lesion distal to the wrist is quite rare. We present two unusual cases of NF involving the palm, supported by detecting ubiquitin specific protease 6 gene rearrangement. The first patient had non-intraneural NF presenting as peripheral neuropathy affecting the digital nerve while the second patient suffered from painless, non-tender NF in the palm, which had not regressed spontaneously during the five months prior to surgery.

KEYWORDS

Palm – Nodular fasciitis – Neuropathy – Gene rearrangement

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Nodular fasciitis (NF) is a self-limiting reactive process composed of fibroblasts and myofibroblasts. Although NF may occur virtually anywhere on the body, the lesion is commonly found in the forearm. Two large series of NF patients showed only 0–2% of cases occurred in the hand.^{1,2} In about half of the cases, there is associated soreness, tenderness or slight pain.^{1,5} The features of NF on magnetic resonance imaging (MRI) are non-specific. As a result, NF is a difficult diagnosis in tumours of the hand. Rearrangement of the ubiquitin specific protease 6 (*USP6*) gene has recently been reported as a recurrent and specific finding in NF.⁴ We describe two cases of NF involving the palm, confirming this finding of *USP6* rearrangement.

Case 1

A 55-year-old man presented with a mass in the palm with paraesthesia in the radial aspect of the right middle finger. He had no history of trauma and his medical history was unremarkable. Four weeks after noticing the paraesthesia, he became aware of the mass involving the tissue in palmar side of the index and middle metacarpophalangeal joints. On MRI, the lesion measured 18mm × 16mm × 12mm; it was isointense on T1 weighted imaging and isointense to hyperintense on T2 weighted imaging (Fig 1). The digital nerve (visible as a small round area of hypointensity on MRI) was trapped within the tumour.

Figure 1 Preoperative magnetic resonance imaging: T1 weighted axial sequence (A), T2 weighted axial sequence (B) and T1 weighted axial sequence with gadolinium enhancement (C). A small round area of hypointensity (the digital nerve) is visible

A giant cell tumour of the tendon sheath (or schwannoma) was suspected and an excisional biopsy performed. The tumour was easily resected with preservation of the digital nerve (Fig 2). It was composed of spindle shaped fibroblasts lacking nuclear hyperchromasia, and extravasated erythrocytes and inflammatory cells were identified (Fig 3). Immunohistochemical analysis revealed that the tumour was positive for α -smooth muscle actin and vimentin but negative for S100 protein, CD34, desmin and epithelial membrane antigen. A diagnosis of NF was made. This was supported by fluorescence in situ hybridisation (FISH), which was positive for *USP6* gene rearrangement (Fig 4). The patient's paraesthesia disappeared three months after surgery.

Case 2

A 50-year-old woman noticed a mass in the palm 5 months prior to visiting our clinic. She had no history of trauma and her medical history was unremarkable. On physical examination at our hospital, a non-painful and non-tender mass measuring 1cm in diameter was observed between the proximal palmar crease and thenar crease. On MRI, the lesion measured 10mm $\times$ 8mm $\times$ 7mm, and was isointense on T1 and isointense to hyperintense on T2 weighted imaging (Fig 5). A fibroma was suspected and an excisional biopsy performed. The tumour was attached to the palmar aponeurosis. It was composed of spindle shaped cells in a myxoid matrix with scattered inflammatory cells (Fig 6). The tumour was diagnosed as NF. This diagnosis was supported by FISH demonstrating *USP6* gene rearrangement (Fig 7). There was no local recurrence at the eleven-month follow-up visit.

Figure 3 Haematoxylin and eosin staining of the tumour in case 1 (200 $\times$ magnification)

Discussion

Peripheral neuropathy is an unusual symptom in NF.^{5,6} Furthermore, in our patients, the tumour arose in the palm and lesions distal to the wrist are rare. For this reason, NF was not considered initially as a differential diagnosis. The characteristic radiological feature of case 1 was a small round area of hypointensity observed on MRI, which was the peripheral nerve. This has also been observed in other cases of intraneural NF.^{7,8} The digital nerve could be easily

Figure 2 Intraoperative photographs showing the tumour (A) and resection with preservation of the digital nerve (B)

Figure 4 Fluorescence in situ hybridisation for *USP6* gene rearrangement revealing 18% split signal

Figure 6 Haematoxylin and eosin staining of the tumour in case 2 (200× magnification)

separated from the tumour and the neuropathy disappeared three months after surgery.

NF usually has a history of rapid growth and a preoperative duration of two months or less.⁹ Our second patient, however, had noticed the growth five months before presentation and its size remained stable during that time. About half of patients with NF experience associated soreness, tenderness or slight pain.^{1,3} NF occurring in the palm is rare (except for intraneural NF). Only 18 cases have been reported in the English literature.^{3,6,9–19} A third of these patients ($n=6$, 33%) suffered from pain or tenderness. In NF involving the palm, such symptoms appear to be less frequent than in other locations.

NF may be confused with numerous benign and malignant mesenchymal lesions. Although some cases of NF may resemble palmar fibromatosis (Dupuytren's disease), palmar fibromatosis is a poorly circumscribed lesion that typically infiltrates the surrounding soft tissue, in contrast to the circumscription of NF. Histologically, it is characterised by spindle shaped fibroblasts arranged in long, sweeping fascicles and separated by abundant collagen, which is different to NF.

Most cases of NF exhibit rearrangement of *USP6* (located at 17p13).⁴ This discovery has challenged the traditional view of NF, which saw the growth as reactive and

Figure 5 Preoperative magnetic resonance imaging: T1 weighted axial sequence (A) and T2 weighted axial sequence (B)

Figure 7 Fluorescence in situ hybridisation for *USP6* gene rearrangement revealing 26% split signal

introduced the concept of transient neoplasia. FISH for *USP6* gene rearrangement is a useful ancillary test in cases where the location of the lesion is unusual and NF is a potential diagnostic consideration.²⁰

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