CASE REPOSITORY

Reticular Perineurioma of the Hand: Diagnosis and Treatment of a Rare Case of Hand Mass

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Reticular perineurioma is a rare and recently delineated morphologic variant of benign perineurioma of skin and soft tissues. Because of its nonspecific gross appearance, varying histologic patterns, and potential range of cellularity, perineurioma of the hand is likely to be confused with more commonly encountered tumor or tumor-like conditions such as schwannoma, neurofibroma, fibromyxoid tumors, and giant tumor of tendon sheath. We report the case of a 20-year-old woman who presented with a slowly growing mass of the hand, which was eventually identified as a reticular perineurioma. *(J Hand Surg Am. 2016;■(■):■−■. Copyright © 2016 by the American Society for Surgery of the Hand. All rights reserved.)*

Key words Hand, imaging, perineurioma, tumor.

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Superficial soft tissue masses of the hand are common in clinical practice. Considering the multitude of subcutaneous masses described in the hand, the diagnostic approach should be systematic to narrow the differential diagnosis. Perineuriomas are rare peripheral nerve sheath neoplasms and an uncommon entity among mesenchymal tumors.[1,2] Reticular perineurioma represents a recently characterized subset of perineurioma rarely encountered in the hand.[3] It is likely to be confused with more commonly encountered tumor or tumor-like conditions such as schwannoma, neurofibroma, fibromyxoid tumors, and giant tumor of tendon sheath.[4] We present a combined imaging and pathological approach, which was essential to identify the diagnosis of perineurioma and thereby avoid inappropriately aggressive therapy.

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Received for publication July 28, 2016; accepted in revised form November 4, 2016.

No benefits in any form have been received or will be received related directly or indirectly to the subject of this article.

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0363-5023/16/■ ■-0001$36.00/0
http://dx.doi.org/10.1016/j.jhsa.2016.11.009

CASE REPORT

A 20-year-old woman presented with a 2-year history of a persistent nonulcerated nodule located between the third and fourth distal intermetacarpal spaces of the right hand. The lesion was situated on the palmar side of the hand, easily palpable in the immediate subcutaneous tissue and mobile (Fig. 1). No skin invasion was noticed and no neurovascular deficit was associated with the mass. The mass was painful under pressure although not sensitive to cold.

The patient had no relevant medical history other than juvenile osteoporosis that was treated with calcium and vitamin D. After standard radiological imaging that did not display abnormalities, magnetic resonance imaging (MRI) showed a round well-delineated mass 1.3 cm in diameter, which was hyperintense in T2 and intermediate density in T1 (Figs. 2, 3). There was no vascular or nerve

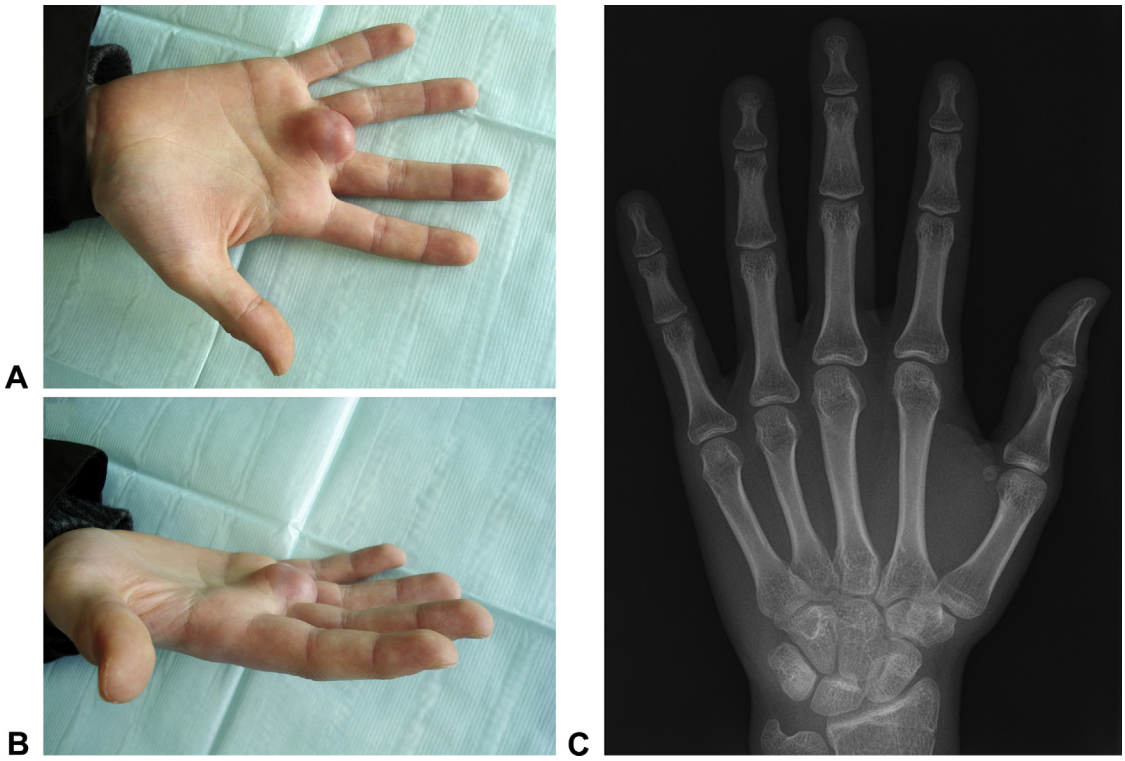

FIGURE 1: A, B Clinical appearance of the subcutaneous mass. **C** Conventional x-ray.

compression, no invasion of soft tissue, and no continuity or contact with tendon, bone, or adjacent joints. The presence of this slow-growing hypervascularized mass was thought to be consistent with a benign or unspecified nerve tumor.

After radiological investigations, the mass was removed without perioperative complications and confirmed the absence of nerve or tendon infiltration. The wound healed without complication. Histopathology completed with immunohistochemical profile was positive for epithelial membrane antigen (EMA) and vimentin, negative for S-100 protein, and mostly negative for cytokeratin, revealing the features of a reticular perineurioma and establishing the diagnosis. There were no signs of recurrence 48 months after treatment and the patient reported no symptoms.

DISCUSSION

The three most frequent types of benign mesenchymal tumors in the hand are synovial ganglion cysts (52%), giant cell tumor of the tendon sheath (11%), and epidermoid cysts (6%).[5]

In the current patient, the lesion was not located over a tendon or joint, and thus excluded a synovial cyst. Giant cell tumor could be considered, but MRI findings did not show uniform low signal intensity in T1 or high signal intensity in T2-weighted sequences (Fig. 2), and angio-MRI showed aspecific mass vascularization (Fig. 3). Mucoid cysts can be transilluminated and are normally located in the distal interphalangeal joint. Lipomas (about 3% of discovered hand masses) can arise anywhere in the hand and share similarities with our case. However, they have a less solid consistency and can be clearly recognized on fat saturation MRI sequences.[6] We considered peripheral nerve sheath tumors such as schwannoma or neurofibroma to be diagnostic possibilities, because they globally account for about 3% of all benign soft tissue neoplasms of the hand. However, they are generally associated with a Tinel sign and are more commonly located distally over the digital nerves. No irradiating pain and intolerance to temperature variations was present, which clinically excluded glomus tumors. Among benign mesenchymal tumors, myxoma is the most prevalent and could not be excluded by either the clinical or MRI findings.[8] Malignant tumors should be always considered in the differential diagnosis.[5,7] However, these are rare and not tender, and show a rapid evolution, with radiological evidence of infiltration. None of these features were seen in our patient.

Perineuriomas are rare peripheral nerve sheath neoplasms and a distinct entity among mesenchymal tumors.[1,2] The perineurium is made of layers of flattened and elongated cells surrounded by collagen