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# Finger Nerve Sheath Myxoma: A Rare Case Report

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**Abstract**

**Introduction:** Nerve sheath myxoma (NSM) is a rare benign neoplasm of nerve sheath origin with Schwann cell differentiation. NSM and neurothekeoma were considered one single phenomenon in the past.
**Case Presentation:** A 28-year-old woman presented to our hospital clinic with pain and a fixed firm mass in the proximal phalanx of the right second finger. Excisional biopsy was performed and histological examination revealed NSM.
**Conclusions:** Based on the findings, NSM should be considered in the differential diagnosis of firm and tender masses in the upper extremities of young adults.

*Keywords:* Nerve Sheath Myxoma, Finger, Neurothekeoma

## 1. Introduction

Nerve sheath myxoma (NSM) is a benign and rare soft tissue tumor, which can involve the face and extremities, most commonly the fingers and the hands. So far, rare cases involving mucosal surfaces have been recorded. Also, NSM can present in the spinal cord and intracranial space (1, 2), although it is usually found as dermal lesions in young adults (3).

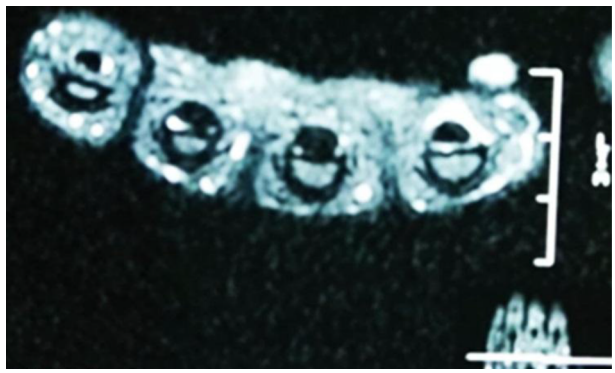
## 2. Case Presentation

A 28-year-old woman presented to our clinic with subcutaneous nodule in the proximal phalanx of the right second finger in the palmar aspect (5 × 5 mm). She had experienced pain in the finger over the past year and her finger had grown in size during the past nine months. The patient described mild vague pain on palpation, while she experienced no sensorial or motor disturbances and her pain did not exacerbate at low temperatures.

The patient had no history of trauma and no malignancies or other medical disorders were reported in her family. Her laboratory tests indicated normal results. In addition, plain radiography was found to be normal. Nevertheless, magnetic resonance imaging (MRI) showed a well-defined, heterogeneous, solid tumor in the radial side of the right second finger (Figure 1).

After surgical excision, a well circumscribed, creamy, rubbery tissue (5 × 5 mm), surrounded by a prominent fibrous border, was detected in the specimen. In microscopic examination, a myxoid nodule, containing spindle and epithelioid cells with no significant nuclear atypia, was found (Figure 2). The cells showed S-100 protein immunoreactivity.

**Figure 1.** MRI of the Hand



There is a slightly heterogeneous hyperintense lesion on the radial side of the proximal phalanx of the second finger.

The patient's symptoms disappeared after the operation and several follow-ups during 24 months showed no evidence of recurrent lesion. Her status returned to normal and she had no limitations in her daily activities.

The present study was approved by the ethics board of the hospital clinic, and a written consent was obtained from the patient for the case report.

## 3. Discussion

NSM is a rare benign soft tissue tumor with Schwann cell differentiation, which commonly involves the skin of

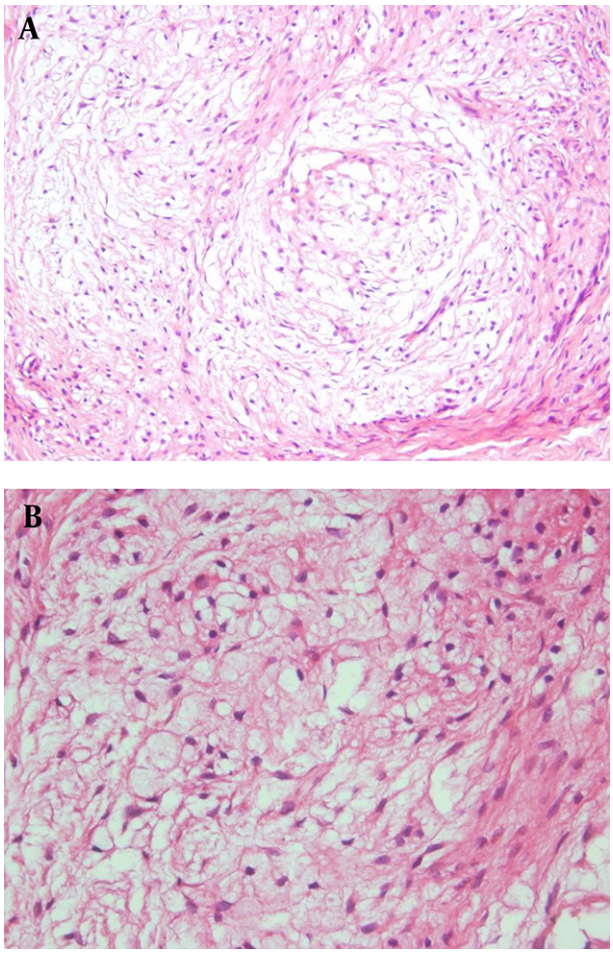



**Figure 2.** A, Tumor cells arranged in nodules separated by fibrous septa (H&E, × 20); B, spindle-shaped cells with no nuclear atypia (H&E, × 40)

fingers and hands in young patients (4). NSM was first described in 1969 by Harkin and Reed, according to histological and electron microscopic studies (3). They believed that sensory nerve endings were the origin of NSM, whereas other investigators presumed a perineural sheath or Schwann cell origin (5).

NSM is commonly a dermal tumor, although other rare sites, such as the oral cavity, intracranial, intraspinal, and paravertebral spaces, trunk, foot, and ankle, have been also described (6). NSM has been reported in different age groups, ranging from five-month-old infants to elderly patients; nevertheless, it commonly presents in the third and fourth decades of life (1, 2). These tumors can involve the knee and pretibial regions, commonly in the third and fourth decades of life (2). NSM is slow growing, and in the majority of cases, the lesions remain painless and subepidermal for many years before indications for surgical resection appear (6, 7).

Microscopically, NSM is composed of multiple lobules of myxoid tissues, separated by dense fibrous septa, and is usually limited to dermal and subcutaneous tissues. It is paucicellular and contains spindle cells, occasionally forming cords, networks, and syncytial nests. The cells are always positive for S-100 protein. Also, glial fibrillary acidic protein (GFAP) is another useful marker for NSM, which is positive in most cases. Nevertheless, NSMs are negative for smooth muscle actin (SMA), desmin, CD68, HMB-45, synaptophysin, and chromogranin (2).

Diagnosis of NSM is usually simple when the tumor is in its usual location on finger. However, in rare sites, such as the paravertebral space, diagnosis may be challenging (3, 7). Microscopic differential diagnosis of NSM includes different myxoid tumors, e.g., neurothekeoma, superficial angiomyxoma, myxoid neurofibroma, low-grade fibromyxoid sarcoma, superficial acral myxoma, and juxtaarticular myxoma (3); however, the main differential diagnosis is neurothekeoma. In fact, NSM and neurothekeoma were considered a single phenomenon in the past, while after the advent of immunohistochemistry, they were separately defined. These tumors have different gene expression profiles (8), and neurothekeoma has been suggested to belong to the category of fibrohistiocytic tumors.

Neurothekeoma mostly involves the upper extremities or the head and the neck and occurs in the second or third decade of life. The male-to-female ratio is almost 1: 1 in NSM, whereas in neurothekoma, it is 1: 1.8 (7). In fact, neurothekeoma is an entirely different benign tumor with a less myxoid matrix and a less defined border, commonly showing a whorling architecture. It also lacks a specific immunohistochemical profile and is S-100-negative. This tumor expresses a variety of non-specific markers, including NKI/C3, NSE, and SMA antibodies and occurs in children or young adults (frequently found on the face) (7).

NSM arises from sensory nerve endings (9). Positive reaction of the antibodies to S-100 protein shows that the progenitors of this tumor are Schwann cells (5). The present case was a classic myxoid type with low cellularity and a large myxoid matrix. The appropriate treatment of NSM is surgical excision with a safe margin. Myxomas must be completely excised considering the 50% recurrence rate (4). However, in our case, no recurrence was observed in the two-year follow-up.

Based on the findings, NSM should be considered in the differential diagnosis of all patients with a soft tissue mass in the finger. Surgical excision with a safe margin should be performed for these patients, regarding the high recurrence rate in simple local excision. Diagnosis may be difficult in an uncommon presentation of NSM; also, differential diagnosis includes malignant tumors. Histopatholog-

ical and immunohistochemical studies should be carried out, and close follow-up should be performed after complete surgical excision.

## Footnote

**Authors' Contribution:** The study design, data acquisition, drafting, and revision of the manuscript: Tina Shooshtarizadeh and Mehdi Mohammadpour.

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