

Periungual Myxoid Neurofibroma

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A 34-year-old woman presented with a slow-growing nontender nodule on her left index finger that had been present for 2 years. The tumor was excised and was histologically determined to be a myxoid neurofibroma. We report this case because of the rarity of both the tumor and its periungual location and to provide a review of the literature.

Myxoid neurofibroma is a benign tumor of perineurial cell origin.^{1,2} Myxoid neurofibroma has a higher incidence in teenagers and young adults^{3,4} and usually presents as a solitary dermal nodule that is either nontender or mildly tender. The most common locations described are the face, shoulders, and arms.^{1,4} Although these lesions also have been reported on the feet, they show a predilection for the upper extremities.⁵ Lesions vary in size from 0.3 to 2.0 cm in diameter and are usually described as pink or bluish. Electron microscopic examination shows a partial or total basal lamina around each cell that supports a perineurial cell origin. In addition, tumors demonstrate positive immunohistochemical staining for S-100 protein, which further supports a neural crest origin.⁴

Case Report

A 34-year-old woman presented with a slow-growing nodule on her left index finger that had been present for 2 years (Figure 1). The patient reported that other than sensitivity to pressure, the lesion was nontender. She sought medical attention for cosmetic improvement.

The patient denied any past medical problems and was on no medications. She reported prior allergic reactions to sulfonamides, cefaclor, and amoxicillin. Examination revealed a 1- to 2-cm



Figure 1. Periungual nodule of the left index finger.

flesh-colored nodule involving the radial portion of the left index finger. The nodule was elevating and bowing the fingernail. Sensation in the area was intact and immersion of the finger in ice water for several minutes caused no discomfort compared with the contralateral index finger. An x-ray revealed a cortical defect from compression of the radial aspect of the distal phalanx (Figure 2).

Excision of the tumor was performed under digital block. An incision was made through the nail wall perpendicular to the axis at its radial proximal corner, exposing the full extent of the nail bed up to the germinal matrix. An incision was then made through the radial aspect of the nail bed. There was

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Figure 2. An x-ray of the left index finger distal phalanx illustrating a cortical defect.



Figure 3. Gross presentation of the tumor intraoperatively.

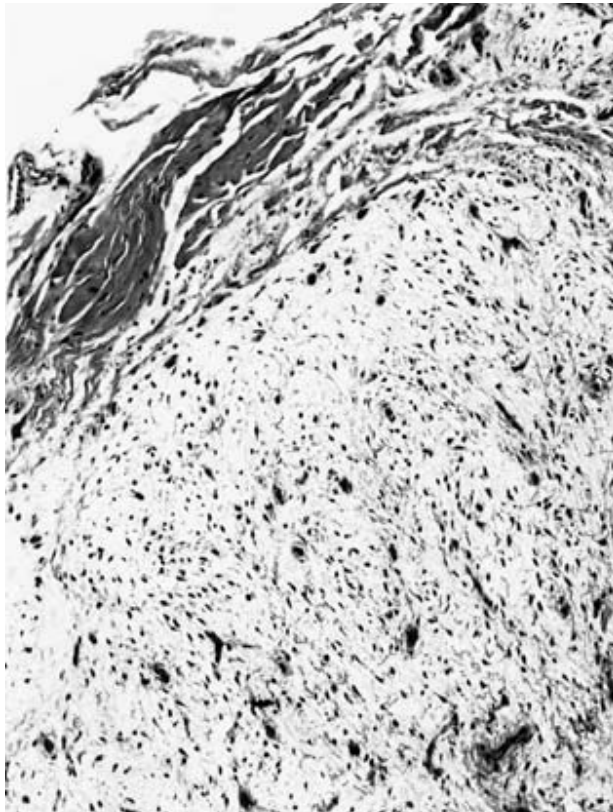


Figure 4. Histologically, the tumor shows a proliferation of wavy spindle-shaped cells embedded in a mucinous stroma (H&E, original magnification $\times 150$).

a smooth white tumor growing up into the nail bed, from which separation was tedious and had to be done completely by sharp dissection. The nail bed was separated from the tumor without perforation and every attempt was made to remove the tumor in its entirety. It was observed that the tumor extended deeply and appeared to be compressing the bone but not invading it.

The tumor extended into the soft tissues on the volar aspect of the digit and seemed to originate from the neurovascular bundle. Grossly, it was lobulated and translucent (Figure 3). Histopathology results revealed a well-circumscribed nodule composed of spindle-shaped cells with wavy nuclei. Mast cells were present and there was abundant mucin (Figure 4). The microscopic diagnosis was myxoid neurofibroma.

Comment

Myxoid neurofibromas have been described under many different terms including nerve sheath myxoma, pacinian neurofibroma, neurothekeoma, bizarre cutaneous neurofibroma, and lobular neuromyxoma. The tumor is most notorious for being a component of the NAME syndrome (nevi, atrial myxoma, myxoid neurofibroma, and ephelides).⁶ Myxoid neurofibroma is usually a solitary lesion, as in our patient; however, they can be numerous and may recur following an incomplete initial excision.

Early reports mention the danger of these tumors masquerading as malignancies. Histologic examination of myxoid neurofibroma is paramount to exclude malignant tumors such as neurofibrosarcoma and epithelioid sarcoma.

Our patient underwent surgical resection without complication. She has had no recurrence to date. To our knowledge, this is the second report of a myxoid neurofibroma in the periungual region.⁴ Myxoid neurofibroma should be included in the differential diagnosis of periungual tumors.

REFERENCES

1. Angervall L, Kindblom LG, Haglid K. Dermal nerve sheath myxoma. *Cancer*. 1984;53:1752-1759.
2. Erlandson RA, Woodruff JM. Peripheral nerve sheath tumors: an electromicrographic study of 43 cases. *Cancer*. 1982;49:273-287.
3. Rosati L, Fratamico FC, Eusebi V. Cellular neurothekeoma. *Appl Pathol*. 1986;4:186-191.
4. Donofrio V, Passeretti U, Russo S, et al. Neurothekeoma of the thumb: a case report. *Tumori*. 1988;74:751-754.
5. King DT, Barr RJ. Bizarre cutaneous neurofibromas. *J Cutan Pathol*. 1980;7:21-31.
6. Atherton DJ, Pitcher DW, Wells RS, et al. A syndrome of various cutaneous pigmented lesions, myxoid neurofibromata and atrial myxoma: the NAME syndrome. *Br J Dermatol*. 1980;103:421-429.