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Solitary Subungual Myxoid Neurofibroma of the Thumb: A Case Report

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Purpose: Subungual tumors are a common cause of nail plate deformity, and may be caused by fibrokeratoma, Koene's tumor and glomus tumors. Neurofibromas, either as part of neurofibromatosis or as a solitary tumor are exceptionally rare in the digits.

Methods: A 44-year-old man presented with painless onychodystrophy and nail plate elevation of the right thumb due to a small subungual mass that had started growing 3 years ago. Sensory evaluation of the distal phalanx was normal, and no discoloration nor infection signs were seen. The nail plate was extracted under local anesthesia, and the mass was delicately removed without injury to the nail bed. The nail matrix was repaired with primary closure.

Results: Histopathology shows a well circumscribed, cellular tumor with myxoid stroma. Tumor cells were S-100 protein positive, and the patient was diagnosed with myxoid neurofibroma. There has been no sign of recurrence to date, 14 months after the operation.

Conclusion: Presentation of cutaneous neurofibromas in the digits is an uncommon finding. They may occur as a manifestation of neurofibromatosis or as a solitary tumor. Subungual neurofibromas are exceptionally rare. To our knowledge, there are only ten reports of solitary subungual neurofibroma unrelated to neurofibromatosis to date. We report a rare case of solitary subungual myxoid neurofibroma of the thumb, that was treated through total excision, with preservation of the nail matrix.

Key Words: Neurofibroma, Nail diseases, Onychodystrophy

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I. INTRODUCTION

Subungual tumors are a common cause of nail plate deformity and are often misdiagnosed as an inflammatory or fungal disease. The most common subungual nail tumors include fibrokeratoma, Koene's tumor and glomus tumors.[1] However, neurofibromas, either as part of neurofibromatosis or as a solitary tumor are exceptionally rare in the digits. We report a case of solitary subungual myxoid neurofibroma of the thumb.

II. MATERIALS AND METHODS

A 44-year-old man presented with painless onychodystrophy and nail plate elevation of his right thumb due to a subungual mass that had started growing 3 years ago (Fig. 1). The lunula was distorted, and a slit of sterile matrix was exposed at the ulnar perionychium. Sensory evaluation of the distal phalanx was normal, and there was no associated inflammation.

The nail plate was extracted under local anesthesia. A bulging mass in the proximal ulnar aspect of subungual tissue was revealed. We designed an elliptical incision on the nail matrix that extended onto the germinal matrix. A solid, encapsulated, white mass was

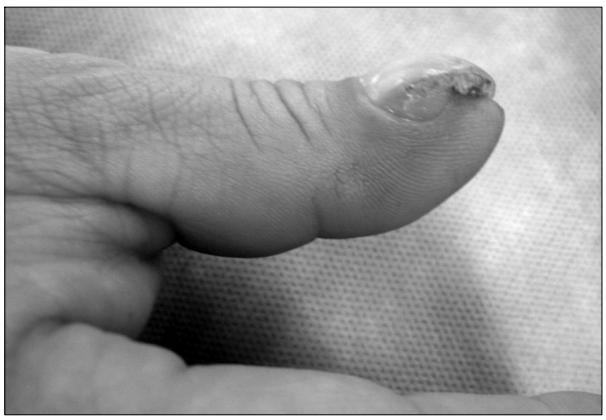

Fig. 1. Preoperative view of the patient's thumb viewed from the ulnar side.

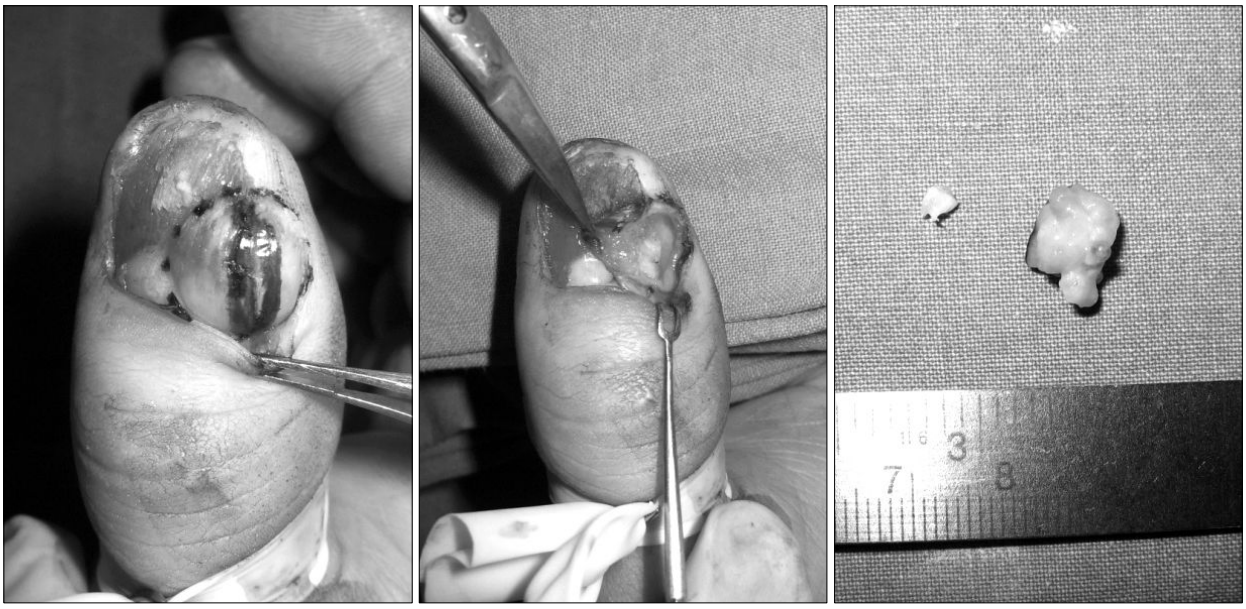

Fig. 2. Preoperative (Left) and postoperative (Center) views of the right thumb, with the excised specimen (Right). The margin of the mass extended under the nail fold. Total excision was completed with preservation of the overlying nail matrix, and coverage was possible with primary closure. The specimen was an encapsulated white, solid rubbery mass measuring 1.0 × 0.8 centimeters.

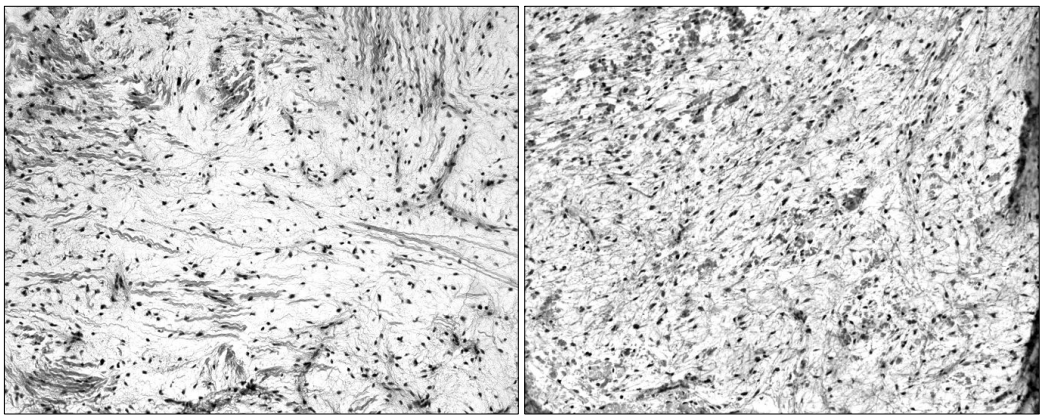

Fig. 3. Histopathology findings. Irregularly arranged spindle cells with elongated and wavy nuclei in a myxoid stroma with thin collagen fibers scattered in between seen in hematoxylin and eosin staining (× 200, Left). Protein S-100 stain was positive (× 200, Right).

removed by delicate dissection, and the matrix was repaired with primary closure then covered with the original nail (Fig. 2).

III. RESULTS

Histopathological findings revealed a well circumscribed, cellular tumor with myxoid stroma, whose tumor cells were S-100 protein positive, corresponding with the diagnosis of myxoid neurofibroma (Fig. 3). There has been no sign of recurrence to date, 14 months after the operation (Fig. 4).

IV. DISCUSSION

Cutaneous neurofibromas occur as a manifestation of neurofibromatosis or as a separate, solitary tumor. Presentation in the digits is rare in both cases, and subungual neurofibromas are exceptional. There are only ten reports of solitary subungual neurofibroma unrelated to neurofibromatosis in the literature.[2-6] The first such mass was reported by Runne and Orfanos

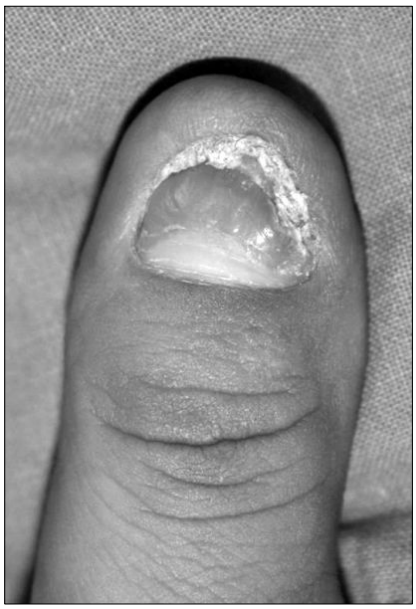

Fig. 4. The patient's thumb at 14 months follow-up. Nail tissue from the germinal matrix is seen growing from underneath the eponychium with no major distortions.

in 1981. Though solitary neurofibromas are known to develop between the third and fourth decades without sexual predominance,[3] recent reports in the literature were usually of females, ages ranging from 13 years to 60.[2-5] The masses arise from small nerves, grow slowly, and present as painless nodules. Definitive diagnosis is made through pathological studies. Surgical findings consist of a well circumscribed tumor, and excision is considered adequate treatment. Care should be taken to minimize trauma to the nail matrix. We report a rare case of solitary subungual myxoid neurofibroma of the thumb, which was treated through total excision with preservation of the nail matrix.

V. CONCLUSION

Cutaneous neurofibromas of the digits are rare, and subungual neurofibromas are exceptional. This is a documentation of our experience with a case of solitary subungual myxoid neurofibroma of the thumb.

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