

# A Rare Soft Tissue Perineurioma of the Hallux

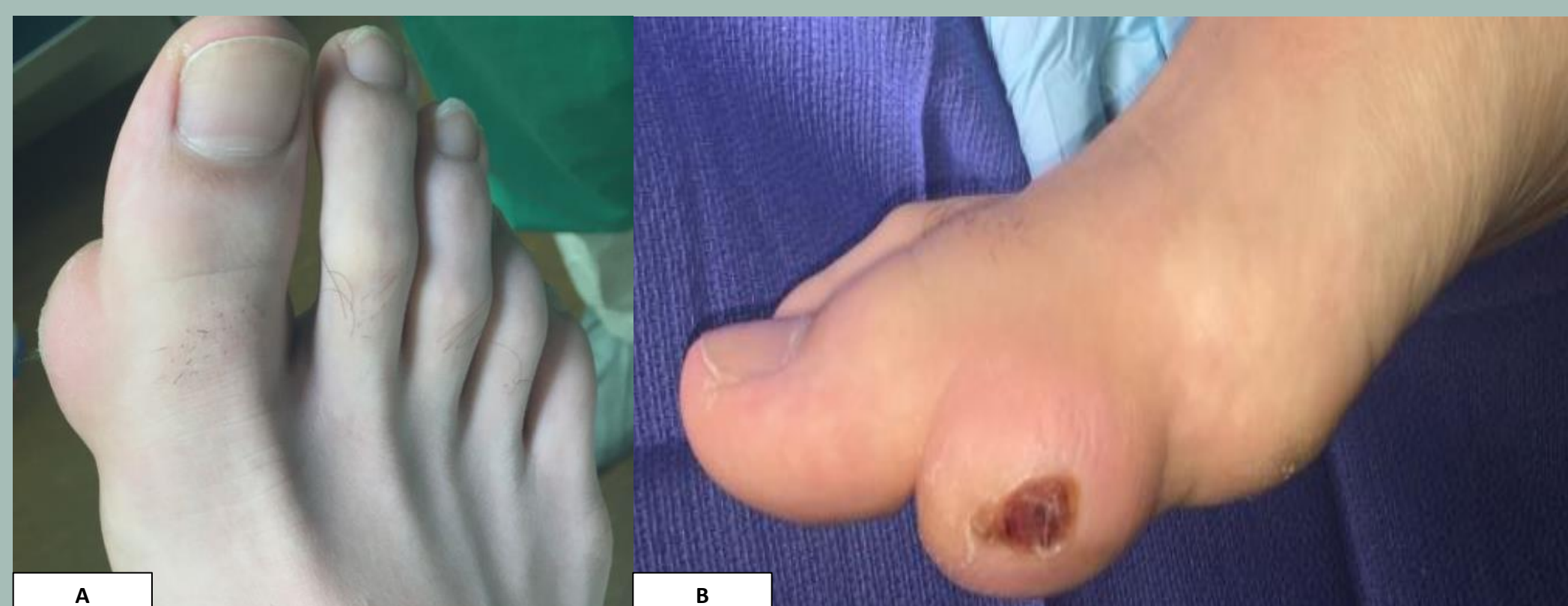
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## Introduction

A 18 year old male presented to the office for a painful mass to his right big toe for about 2 years. The mass started out to be small and grew in size over the years. He was referred to a dermatologist who took a biopsy which showed the mass to be benign. The patient was active in school with sports so his family and he opted to remove the mass as it was interfering with his daily activities.



Figures A and B: Preoperative clinical image of soft tissue mass and ulceration

## Background

Perineuriomas are rare tumors of the peripheral nervous system. These tumors present as well-circumscribed, firm, non-encapsulated soft tissue masses found in the dermis (1-2). They usually present as non-painful soft tissue masses in the extremities, which are often misdiagnosed as more common tumors such as neurofibroma, fibromyxoid tumors and giant cell tumors of tendon sheaths(4). Perineuriomas are usually benign and rarely reoccur, however several cases of malignant perineuriomas have been reported in the literature (2-3). First described in 1978 by Lazarus and Trombetta (1), they are now classified into two types: intraneural which arise directly from the nerve trunk, and extra neural which arise from adjacent soft tissue. Formation of intraneural perineurioma occur when neoplastic perineural cells incorporate a part of the endoneurium (1). Histologically these tumors display elongated spindled cells with loose storiform short fascicular and whorled growth patterns (4). While perineuriomas can display many atypical histologic features, they seem to have no clinical significance (2).

## Methods

Procedure 1: Excision of the skin ulceration

- 2 semi-elliptical converging incisions made on the medial aspect of the hallux and around the ulceration, underlying the proximal phalanx
- Ulceration ellipsed down to the level of the soft tissue mass
- Specimen passed this off in a sterile cup within formalin Pathology for histological analysis

Procedure 2: Excision of soft tissue mass

- Foot exsanguinated and tourniquet inflated to 250 mmHg
- Utilized loupe magnification at 2.5X
- Metzenbaum scissor used to bluntly dissect around the soft tissue mass
- Multiple loculations visualized, mass was firm and nodular
- Did not appear to be intertwined within the flexor tendon or within the capsule
- Small arterial blood flow within the mass visualized
- Cautery used to maintain hemostasis
- Neurovascular bundles, both dorsally and plantarly protected
- Mass excised and sent to Pathology, measured over 4 cm
- Irrigated incision with 1 L of normal saline utilizing mechanical lavage
- Tourniquet released and hemostasis achieved
- Remodeled the medial side to allow for improved closure and less redundant skin, to help prevent hematoma plantarly

Post-operative: weight bearing as tolerated in post-op shoe

- Seen in clinic for suture removal at 2 weeks
- Incision healed without incident



Figure C: Soft tissue mass excised measuring approximately 4 x 3 cm

Figures D and E: Intraoperative images of closure without tension and good perfusion to right hallux



| Antibody/Reagent | Block | Slide      | Result (staining pattern)                                     |
|------------------|-------|------------|---|
| S100             | A1    | Individual | Negative  |
| Melan A          | A1    | Individual | Negative  |
| Beta catenin     | A1    | Individual | Weak membranous and cytoplasmic staining; no nuclear staining |
| AE1/AE3          | A1    | Individual | Negative  |
| Actin            | A1    | Individual | Negative  |
| Desmin           | A1    | Individual | Negative  |
| CD34             | A1    | Individual | Positive  |
| EMA              | A1    | Individual | Negative  |

Figure F: Pathology histological analysis from Brigham and Women's Hospital in Boston, MA

## Histological Analysis

Specimen was sent for an extradepartmental pathology consultation to Dr. Christopher Fletcher at Brigham and Women's Hospital in Boston, MA. He notes in his read: "This is a seemingly circumscribed spindle cell neoplasm with neural-like cytomorphology and a prominent collagenous stroma. The lesional cells have tapering nuclei and indistinct cytoplasm, and are arranged in a predominantly whorled but focally more fascicular fashion. There is no atypia or pleomorphism. There is no evident Antoni A/B zonation. Immunostains show multifocal weak positivity of EMA along with more diffuse positivity for Claudin-1, while S100 protein and SMA are negative. The appearance fit very well with a soft tissue perineurioma."

In conclusion, the soft tissue mass is not malignant and there is little to no tendency for local recurrence. Immunohistochemical stains were performed to classify the neoplasm (Figure F).

## Discussion

The goal of our treatment was to remove the tumor and allow the patient to return to previous level of function. The literature for perineuriomas describes a very distinctive histological pattern that has a low incidence rate. The rarity of the tumor makes it unique and one that should be studied in further detail in the foot and ankle community to prevent misdiagnosis for malignancy.

## Works Cited

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