Case Study: A Large Solitary Neurofibroma of the Toe

Differentiating between masses of the foot may be critical.

BY ALBERT RAMINFARD, DO, STANLEY R. KALISH, DPM, MARC A. BRENNER, DPM

Introduction

The majority of benign soft tissue masses are non-neoplastic and of little clinical consequence. However, some locally aggressive, large masses including neurofibromas can lead to morbidities including pain, paresthesias, numbness, weakness and even atrophy due to mass effect and compromised innervation. Neurofibromas are typically benign nerve sheath tumors in the peripheral nervous system. They evolve anywhere along a nerve from the dorsal root ganglion to the terminal nerve branches.[1] Neurofibromas are commonly, but not always, associated with Neurofibromatosis Type 1 (NF-1). The solitary form occurs in those who have an isolated neurofibroma and do not have the full syndrome of neurofibromatosis with other masses and/or symptoms such as cafe au lait spots.[2] However, solitary neurofibromas may be the first manifestation of the generalized syndrome of NF-1 and persistent follow-up and monitoring is crucial in these patients.[3]

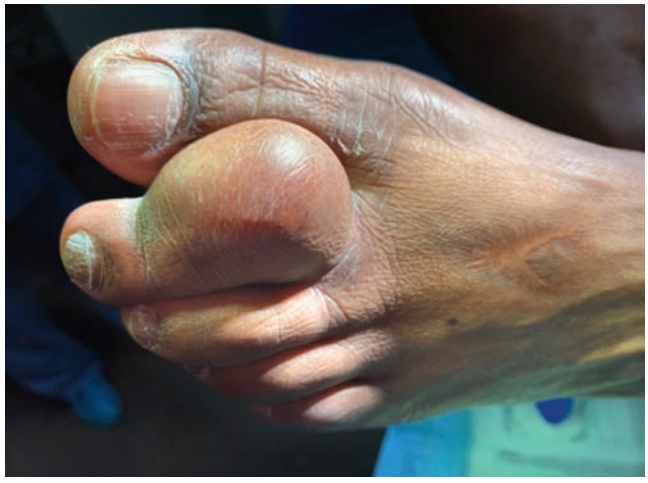

Figure 1: Patient's left foot at presentation. The mass at the 2nd digit had become so large the patient needed to augment his shoes just to wear them and ambulate without pain.

Cutaneous neurofibromas often occur as sporadic, solitary lesions in healthy adults. They typically present as asymptomatic, soft, skin-colored or hyperpigmented papules or nodules that are less than two centimeters (cm) in diameter. Neurofibromas are more common in young adults, without a gender preference. They have been known to exhibit a predilection for the trunk and head compared to other body surfaces.[4] However, solitary neurofibromas have also been reported in the following locations: the spine, retroperitoneal space, mandible, cheek mucosa, nose, bladder, abdominal wall, lower lip and scrotum.[1]

In our case, a patient presented with a large mass in the region of his 2nd toe (Figure 1). Based on our review, there has not been a previously reported case of a neurofibroma presenting in toes and any previous reports of neurofibromas in the region of the feet or plantar surfaces in any capacity have been rare.[5] This patient's presentation was atypical for a neurofibroma and led to concerns of the mass actually being more malignant with infiltration into the foot. Beyond the possibility of a neurofibroma, we developed a large set of differentials for what this mass may have been prior to diagnostic work-up including osteosarcoma, fibrosarcoma and giant cell tumor. The possibility of the mass being a more benign soft tissue mass such as a schwannoma, ganglion cyst or lipoma was also considered.

Neurofibromas are commonly, but not always, associated with Neurofibromatosis Type 1 (NF-1).

Case Report

A fifty-eight year-old male with no past medical history presented with a large mass on his left second toe. He reported that the mass had progressively increased in size over the last 5 years. When it initially appeared, he was not concerned as it was painless, but it had become painful over time due to mass ef-

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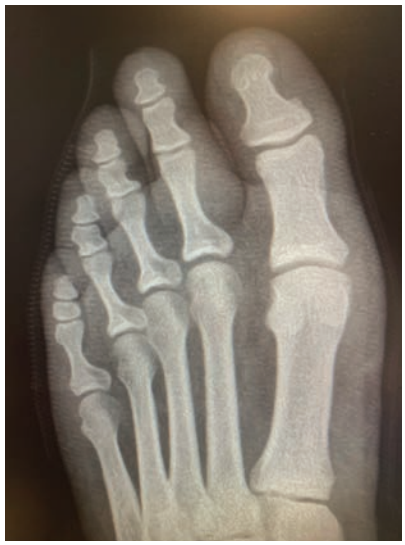

Figure 2: A dorsoplantar radiograph of the patient's left foot exhibiting soft tissue mass without bony erosions or overlying edema.

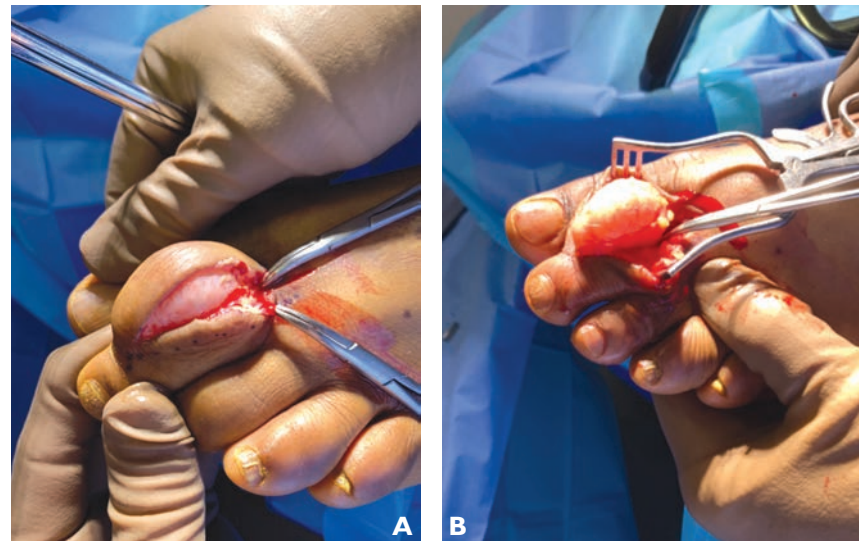

Figure 3a, 3b: Perioperative images of the excision of the soft tissue mass. Initial incision exhibited an encapsulated mass (Figure 3a). Through further retraction and resections around the surrounding tissue, the mass was able to be excised without complications (Figure 3b).

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fect, causing him to cut open his shoes or wear open-toed shoes to simply make room for his feet. His foot began to look as if it had a bony deformity and even his ambulation began to alter. Upon presentation (Figure 1), the mass was not tender but based on history and size was concerning for a potentially malignant mass. To be able to differentiate the level of infiltration of the mass and due to concern for bony involvement, multiple view X-rays were ordered (Figure 2). The ultimate goal of care became excision and biopsy of the mass with minimal residual damage to the healthy tissue of the toe.

Perioperative excision of the mass revealed a 4.3 x 3.8 x 1.7 cm soft tissue mass with no bony infiltration (Figure 3 a,b). Samples were sent for biopsy to ensure that the mass was well-differentiated and not concerning for underlying malignancy (Figure 4). Excision of the mass was ultimately successful, leading to optimal results for cosmesis and functionality (Figure 5). Through histological analysis, biopsies of the mass yielded spindled cells with varying nuclei but without pleomorphism and occasional background mast cells within a fibrillary collagenous matrix. This is known to be histologically compatible to a neurofibroma, rather than a more malignant or poorly differentiated mass. Upon follow-up three months later, the patient still has not had any recurrence or appearance of new neurofibromas, masses, or cafe au lait spots. He remains asymptomatic now and is very happy with the results of his excision. He will continue to follow up and be monitored to ensure he has not developed NF-1 syndrome.

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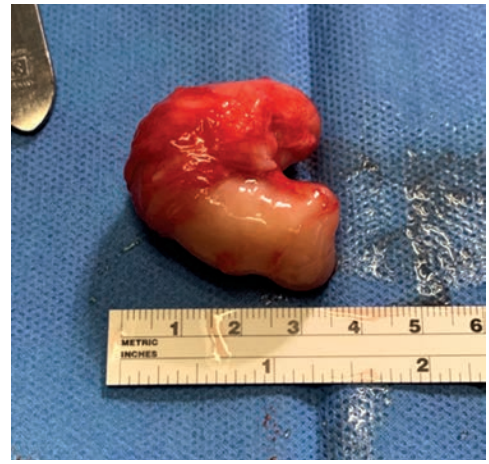

Figure 4: Excised mass, measuring approximately 3.8cm in width. Samples from this mass were sent for biopsy and histological analysis due to concern for malignancy of the mass.

Discussion

Differentiating between masses of the foot may be critical—there must always be suspicion for malignancy and concern for seeding prior to considering surgical intervention. The lower extremities are not a typical location for masses to present. In such situations, one must always consider the risk of bony tumors such as osteosarcoma or giant cell tumors due to the high risk of malignancy and metastasis, but proper examination can help guide differentials. Soft tissue masses may still be malignant, but are much more likely to be benign. They will be softer upon palpation on exam and may be mobile to palpation as well. These tumors may still lead to morbidities, however, as mass effects can lead to pain, paresthesias and weakness pending nerve or vascu-

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lar infiltration or compression. Neurofibromas represent this subset of possible soft tissue masses that can cause mass effect symptomatology.

This case highlights the importance of proper diagnostic workup both pre- and post-operatively. Neurofibromas are typically not painful, not located in the foot or toe, nor are typically this large (greater than 4cm). However, it was important to ensure in this case that the mass was encapsulated and was not infiltrating on any aspect of the toe. There was neither bony involvement nor nerve infiltration. With proper imaging, history and physical examination, we were able to determine that surgical excision with associated biopsy would be the most beneficial method of both diagnosing and treating the patient. If there was involvement of bony tissue or nerve-related symptoms such as numbness, weakness, or paresthesias present there would be a greater concern for a malignant mass. That situation would make surgical intervention more difficult to consider due to obstacles with defining margins for excision and risks of seeding malignant cells.

Post-operatively, histological analysis of our biopsy samples revealed that despite this soft tissue mass being substantially large (much greater than the typical 2 cm or less size) it was a benign solitary neurofibroma. Though patients commonly present with neurofibromas that are smaller than 2cm in size, this patient's hesitancy in pursuing medical care allowed the slow-growing mass to enlarge to a point greater than what we usually observe or expect. This is an important consideration to keep in mind as growing masses may be overlooked by many patients pending location either due to a lack of symptoms, lack of awareness, or fear of a diagnosis revealing malignancy. Through surgical excision, this patient was able to return to his baseline lifestyle and has significant satisfaction with the cosmesis after his procedure. He is now more likely to follow up with his physicians to ensure that he is monitored for any signs or symptoms concerning for NF-1 Syndrome. Although the toe is typically an unusual and unreported location for a solitary neurofibroma, it is important to consider within the differential of soft tissue masses that can present in the podiatric landscape. **PM**

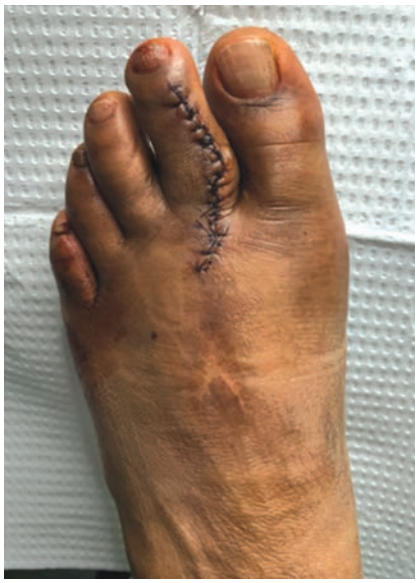

Figure 5: Left foot directly after surgical excision and suturing. Mild edema around the area of the previous mass can be seen here, which resolved over time. The patient is now able to ambulate comfortably with only a small scar as minor cosmetic damage.

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References

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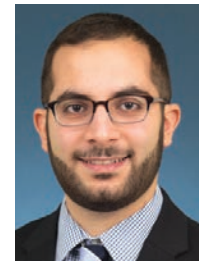

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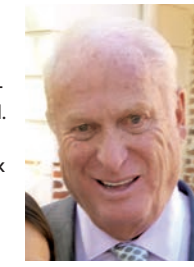
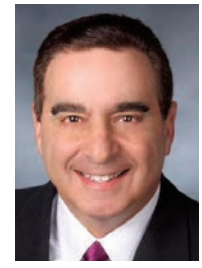


Stanley R. Kalish, DPM, FACFS is the surgeon of record for this case. He is a multi-award-winning podiatrist skilled in treating various foot and ankle conditions for over 40 years including Podiatry Management's Lifetime Achievement Award. He is a well-known author, and inventor of both the TLS drain and the Kalish osteotomy for hallux valgus correction. He specializes particularly in foot and ankle surgery, limb salvage, limb lengthening, bunions, and wound care as a senior staff member of Emory University and in his offices in Jonesboro and Sandy Springs, GA. Contact at srkalish@bellsouth.net



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