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Synovial Sarcoma of the Digits: A Case Report of an Unplanned Excision

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ABSTRACT

Synovial sarcoma is a rare occurrence in the lower extremity, although the presenting symptoms can mimic those of other more common and benign musculoskeletal pathologies. We present the case of a patient who was originally thought to have a Morton's neuroma or ganglionic cyst. The correct diagnosis, synovial sarcoma, was determined only after an unplanned excision. Despite the patient presenting with symptoms similar to those of a compressive neuropathy, a high index of suspicion should be present when a patient presents with any soft tissue mass, especially if it has an unusual clinical appearance to avoid an unplanned excision.

Malignant soft tissue tumors of the foot are extremely rare, although synovial sarcoma is most commonly seen in the extremity (80%) (1) and is the most typical malignant tumor of the foot (2–4). Soft tissue tumors of the foot are usually histologically benign (2), with ganglions accounting for about one third (5). The exact prevalence of malignant soft tissue tumors of the foot has been difficult to ascertain because of their rarity. In a study of 401 cases of soft tissue tumors of the foot, 259 were identified as malignant (6). A more recent study from Syria considered the relative prevalence of malignant soft tissue tumors of the extremities. Of the 308 cases of soft tissue sarcomas, 10% were synovial sarcoma and 7% were found in the foot and ankle (7). The incidence of soft tissue sarcoma of the extremities between genders is similar (8). Often, the lesion has been present for months or years and might have had recent rapid growth.

The term "synovial sarcoma" has been described as a misnomer (9). In contrast to its name, synovial sarcoma is a carcinosarcoma-like tumor with true epithelial differentiation (1,10). The cause of synovial sarcoma remains unclear (11). Trauma might be an incidental, rather than a true predisposing, factor (12). The clinician must not be influenced by the misconception that slow-growing tumors are often considered benign (13).

Clinical differentiation of soft tissue sarcomas and benign tumors in the foot is difficult (14). Clinical features include a palpable mass, swelling, local warmth, and discomfort, although they are often painless until they encroach on other anatomic structures (15). In 1 report, several clinical and radiographic features of a mycetoma were shown to mimic those of synovial sarcoma (16). In contrast, primary synovial sarcoma in the foot can mimic infection or traumatic edema (17). Synovial sarcoma can, therefore, be misdiagnosed as a benign lesion (18). It can also be confused with arthritis, bursitis, synovitis, and posterior tibial tendon dysfunction, in particular, given its slow-growing nature (10). Other misdiagnoses have included ganglia, plantar fasciitis, acute gouty arthritis, Morton's neuroma, tarsal tunnel syndrome, fibroma, and complex regional pain syndrome (19–22). Dorsal sarcomas of the foot can be neglected until they are of considerable size, although those occurring on the plantar surface of the foot are often detected earlier because of discomfort or pain when walking (15).

Unplanned surgical excision refers to situations in which surgery is undertaken without preoperative imaging studies or consideration of the possibility of malignancy. Some evidence has suggested that patients who underwent unplanned excision of a soft tissue sarcoma had poorer oncologic outcomes than those who underwent primary planned excision (15,23). We describe a case of an unplanned excision. Unplanned excision can result in inadequate resection of the surgical margins, requiring further wide excision, and residual disease, which can result in limb amputation or loss of life (15,24).

Case Report

A 67-year-old female presented to the first author's (J.T.) private practice in February 2013. She had been referred by a general podiatrist for a complaint of pain in her right foot. The pain had been present for 3 years, with no recollection of trauma before the onset of symptoms. A previous ultrasound scan had shown a large neuroma/

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bursal complex between the second and third metatarsal heads and extending over the plantar surface superficial to the metatarsophalangeal joints. The patient had been given some functional foot orthoses and digital props, which had provided her with some relief. The patient was fit and enjoyed exercising on a regular basis. The patient described burning, numbness, tingling, and pain beneath the right forefoot and between the right second and third toes, which would radiate to the tips of the toes. The burning and shooting pain was also present between the right third and fourth toes, but to a lesser extent. She reported that the pain was worse when rising onto the toes, and she would avoid this motion of her foot. Pain had become worse over time since the first onset of the symptoms. Her general medical history and family history were noncontributory. She reported taking no medications. Her surgical history included multiple benign cysts removed from her right leg approximately 25 years earlier. She had had an uneventful postoperative course and recovery after these procedures. On examination, marked separation of the right second and third toes was present, with swelling noted, especially in the web space (Fig. 1). Mild pre-existing webbing of the second and third toes was present. Pain was noted on plantar palpation of the right second interspace and to a lesser degree in the third interspace. A large palpable plantar mass was found that extended into the second interspace, with the swelling also extending dorsally. Reproduction of the burning and tingling symptoms was elicited with palpation overlying this region and with lateral forefoot compression.

The patient was referred for radiographic and further ultrasound imaging. The radiograph indicated the presence of a soft tissue mass between the second and third toes at the level of the proximal phalanges. It measured approximately 25×17 mm. A focus of calcification measuring 4×2 mm was seen in the distal aspect of this lesion. The ultrasound scan showed the presence of "a large neuroma in the second and third web spaces, measuring $38 \times 21 \times 21$ mm, which had appeared to have coalesced to form a large lesion and had grown through the web space to the dorsum of the foot, disrupting the dorsal tissue planes." Her presenting symptoms were consistent with intermetatarsophalangeal joint bursitis and secondary compressive neurofibrosis of the plantar digital nerve (Morton's neuroma complex) of the right second interspace, with the suspicion of an additional soft tissue lesion, such as a ganglionic cyst or bursal complex, or damage to the metatarsophalangeal joint area. We decided to proceed to surgery to obtain a more definitive diagnosis by histopathologic examination.

The patient underwent surgical investigation and excision of the suspected neuroma in both interspaces. The patient was placed in a supine position, under general anesthesia, and a tourniquet was not used. The surgical approach was a single dorsal incision over the right second intermetatarsal space that extended in a linear fashion from just proximally to the second metatarsophalangeal joint and distally into the second web space. A large mass was identified just below the subcutaneous tissues and seen to disrupt multiple tissue planes. Extensive dissection was undertaken in a distal to proximal direction, and the mass was excised (Fig. 2). The third interspace was then investigated, and a second specimen with a visual appearance consistent with a neuroma was excised. Histopathologic examination of the second interspace mass showed this to be a synovial sarcoma. Detailed examination revealed an encapsulated nodular solid-cystic mass composed of tumor cells with biphasic spindle and epithelioid differentiation, including areas of true glandular growth (Fig. 3). An unusual aspect of this sarcoma was the extent of glandular differentiation present (Fig. 4). Foci of dystrophic calcification were also seen. Immunochemistry showed a staining pattern typical of a biphasic mesenchymal spindle and epithelioid cell malignancy with a mixed coexpression pattern of epithelial (epithelial membrane antigen and broad-spectrum cytokeratin) and other stromal markers (s100 and Bcl-2; Fig. 5). Excision was narrowly incomplete. Fluorescent in situ hybridization confirmed the presence of the signature translocation X;18, causing SSX1-SS18 gene fusion of uncertain functional significance. The specimen from the third interspace was typical of compressive neurofibrosis (Morton's type) of advanced degree.

Referral to a specialist orthopedic cancer surgeon for further management was recommended, and the patient underwent tarsometatarsal (Lisfranc) amputation. At the patient's request and after a joint discussion between the patient, oncologist, and orthopedic cancer surgeon, it was decided not to proceed to chemotherapy or radiotherapy.

After the Lisfranc amputation (Fig. 6), the patient was fitted with a prosthetic type of shoe filler for the right foot. The patient wore this for approximately 12 months but reported that the dorsal flap of this filler, which was quite rigid, was limiting her amount of right ankle dorsiflexion during gait. This was contributing to left knee symptoms,

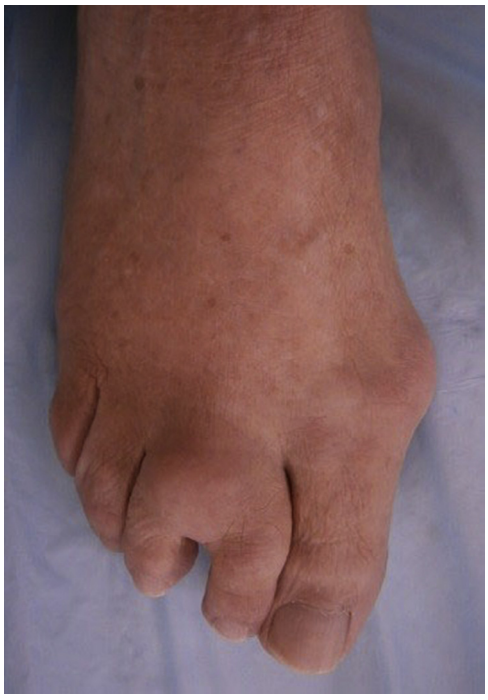

Fig. 1. Clinical photograph of the foot showing separation of the right second and third toes, with swelling noted, especially in the web space.

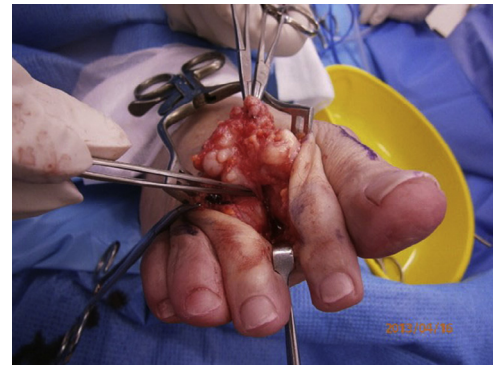

Fig. 2. Intraoperative photograph showing extent of soft tissue lesion resected from the right second web space distally that appeared to be a part of the common plantar digital nerve in the second intermetatarsal space proximally.