The Journal of Foot & Ankle Surgery 53 (2014) 493–496

Contents lists available at ScienceDirect

The Journal of Foot & Ankle Surgery

journal homepage: www.jfas.org

Liposarcoma of the Great Toe: A Case Report

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ARTICLE INFO

Level of Clinical Evidence: 4

Keywords:
diagnosis
ingrown toenail
malignancy
neoadjuvant radiotherapy
pleomorphic
tumor

ABSTRACT

Soft tissue sarcomas are rare malignancies, of which liposarcomas are the most common. Pleomorphic liposarcoma accounts for 5% of liposarcoma diagnoses and most commonly presents in the thigh. A 57-year-old female presented with a 5-year history of a persistent, painless swelling of the left great toe that had been previously diagnosed and treated as an in-grown toenail. After magnetic resonance imaging and core biopsy, a grade 2 pleomorphic liposarcoma was diagnosed. Treatment consisted of neoadjuvant radiotherapy and amputation of the great toe and proximal half of the first metatarsal with primary closure. The patient had no evidence of local recurrence or metastatic disease after 1 year of follow-up and, with the use of a prosthesis, had a good functional outcome. This is the first documented presentation of a high-grade pleomorphic liposarcoma of the great toe to our knowledge. Although soft tissue sarcomas are rare, a high index of suspicion is required by clinicians when presented with a soft tissue mass to promptly diagnose and treat these potentially fatal lesions.

Soft tissue sarcoma (STS) is a type of malignancy derived from the connective tissue that accounts for approximately 1% of all adult malignancies. Liposarcoma is the most common type of STS, accounting for 20% of all adult STS diagnoses and typically presents in the sixth decade of life (1,2). Three subtypes of liposarcoma have been recognized: well-differentiated/dedifferentiated, myxoid/round cell, and pleomorphic. Pleomorphic liposarcoma (PLS) is the rarest subtype, constituting 5% of all liposarcomas, and is characterized by the presence of lipoblasts and nuclear pleomorphism. Patients will typically present with a slow-growing painless lump deep within the limb musculature or retroperitoneum (1). The 5-year survival of patients diagnosed with liposarcoma has been 82%. The disease-specific 5-year survival of patients with PLS has been 53% (2), because patients diagnosed with this type of sarcoma will be more likely to develop metastatic disease than those with other types of liposarcoma (3). PLS has rarely been seen within the foot (4–6), with only 2 cases reported in published studies and no known documented cases of PLS affecting the great toe. We present a rare case of a high-grade PLS that presented as a persistent, ingrown toenail of the left great toe.

Financial Disclosure: None reported.
Conflict of Interest: None reported.

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Case Report

A 57-year-old female had first presented to her general practitioner with a slow-growing mass of the left great toe 6 years previously. An ingrown toenail was diagnosed, and the patient underwent surgical treatment. However, the mass continued to grow slowly for the next 4 years, with more rapid growth within the most recent 6 months, prompting referral to an orthopedic surgeon.

The physical examination revealed a deformed toenail with an associated painless multilobulated mass that had encapsulated the entire distal phalanx to the left first web space (Fig. 1). Movement was uncompromised at the metatarsophalangeal joint but restricted at the interphalangeal joint. The mass was firm, nonmobile, and nonpulsatile and did not transilluminate. The overlying skin was normal in color, with no evidence of peripheral necrosis. The findings from the systemic examination were normal. Magnetic resonance imaging showed a heterogeneous soft-tissue mass measuring 5.7 × 4.8 × 6.8 cm that had not infiltrated the surrounding structures but had caused divergence of the first and second digits (Fig. 2). No cortical bony destruction or marrow edema was noted. Contrast enhancement indicated a sarcomatous change within a lipomatous mass. Therefore, the patient was referred to a specialist soft-tissue sarcoma center for additional management as recommended by the national guidelines (7). Subsequently, a core biopsy obtained by a senior radiologist revealed white lesional tissue consisting of mature adipocytes and pleomorphic lipoblasts. In other areas, spindle cells were seen within a myxoid

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http://dx.doi.org/10.1053/j.jfas.2014.02.009

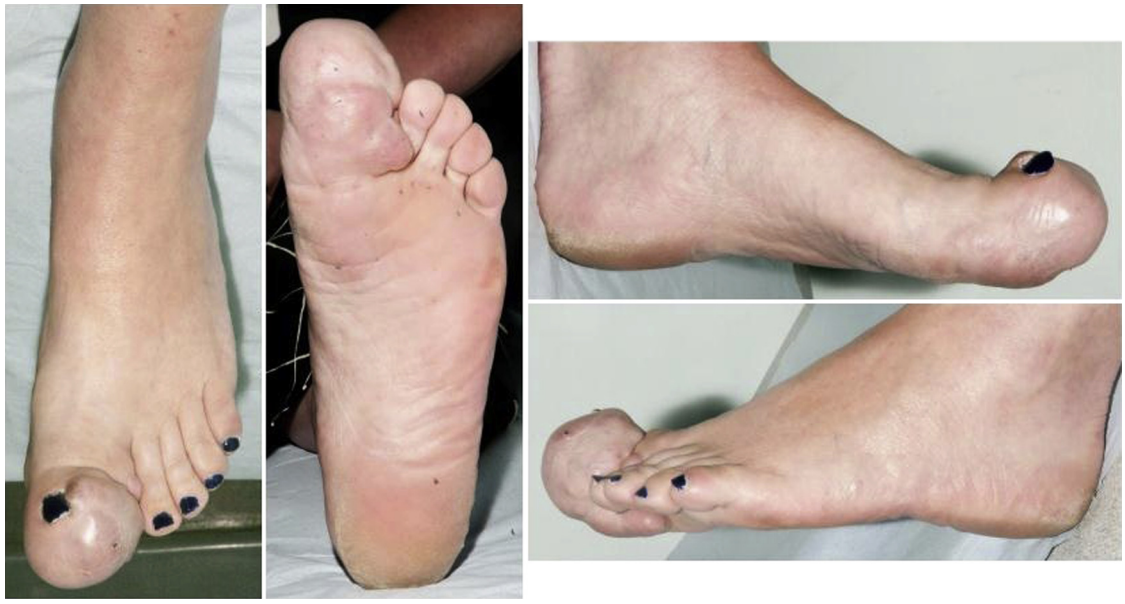

Fig. 1. Clinical photographs taken at the initial presentation.

medium that also contained lipoblasts. Elsewhere, spindle cell proliferation was noted but with minimal pleomorphism (Table and Fig. 3). The histologic impression was that of a Trojani grade 2, PLS (8). Thoracic computed tomography showed 2 small soft nodules, 2 and 3 mm, in the left upper lobe. A subsequent computed tomography scan performed 3 months later excluded metastatic disease.

The imaging and histologic studies were reviewed by the multidisciplinary team, after which the patient underwent neoadjuvant radiotherapy to a dose of 50 Gy, with the aim of reducing the level of amputation, in accordance with the patient's wishes. However, a repeat magnetic resonance imaging scan of the foot showed no reduction in the tumor mass. Surgery was performed approximately 6 weeks after radiotherapy completion. The great toe, metatarsophalangeal joint, and distal one half of the first metatarsal were amputated, and primary closure was achieved. Histologic examination of the resected specimen showed that marginal excision had been achieved; therefore, wider excision was performed 3 weeks later with primary closure. The follow-up examination 1 year after surgery showed no evidence of local recurrence or metastatic disease. The patient was using a first ray orthosis and had had a satisfactory functional outcome (Fig. 4).

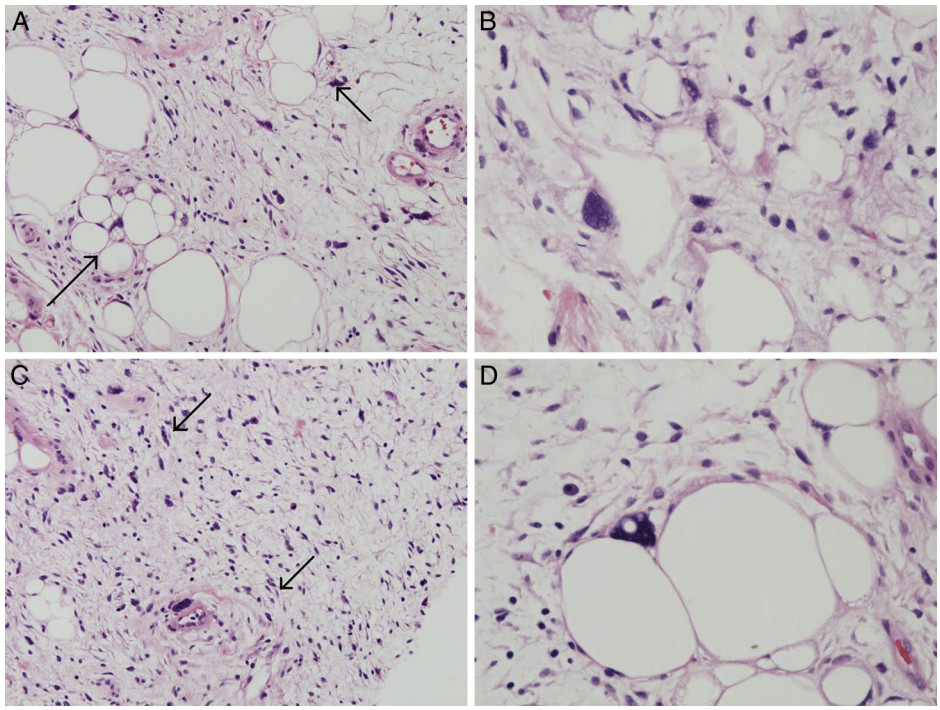

Fig. 2. Histologic photographs of core biopsy specimens. (*A*) Fat cells (*arrows*) of variable shape and size, many with hyperchromatic nuclei. Some fat cells with multivacuolated cytoplasm are present. A background of atypical spindle cells can also seen (100×). (*B*) A closer view of a malignant lipoblast (400×). (*C*) Population of atypical spindle cells (*arrows*) (100×). (*D*) One very large atypical cell can be seen adjacent to a blood vessel (400×). Views show the overall appearance of a pleomorphic liposarcoma.