Pleomorphic hyalinized angiectatic tumor of soft parts is a recently described neoplasm that most commonly affects the lower extremities. It is locally aggressive but has not been known to metastasize. This article presents a case of a softball-sized tumor on the dorsum of the foot that was identified as pleomorphic hyalinized angiectatic tumor of soft parts. Previously, it would have been misdiagnosed and treated as either a neurilemoma or a malignant fibrous histiocytoma. (J Am Podiatr Med Assoc 89(6): 307-311, 1999)
The release of vasoactive mast cell products may be the mechanism that causes an increased vascular permeability. This release of mast cell products may occur as a response to tissue injury induced by tumor infiltration.

Three of the 14 tumors reported by Smith et al did not possess the irregular infiltrative margins characteristic of the other tumors, but had well-defined borders. Because those three tumors possessed all of the other characteristics of the tumors in the study, they were included in the same group. These three tumors were composed of perivascular hyalinized material with relatively few neoplastic cells. The neoplastic cells of these three tumors were possibly less destructively invasive than those of the other tumors, which may have allowed the accumulation of such large quantities of hyalinized material that further tumor cell invasion was retarded or even halted, resulting in the formation of the well-circumscribed masses.

The clinical behavior of pleomorphic hyalinized angiectatic tumor of soft parts is characterized by local recurrence, which can be aggressive to the point of necessitating the amputation of an affected extremity. Such locally aggressive growth, even with extensive surgery and radiation, could suggest that this tumor is a low-grade sarcoma. However, the absence of documented metastases, even with the pleomorphic appearance of the cells, led Smith et al to avoid the word “sarcoma” in their description of these lesions.

Case Study

On December 1, 1996, a 31-year-old woman presented to the Medical Foot Center in Spokane, Washington, with a large mass on the dorsal surface of her right foot. She reported that the mass had been present for 6 years.

Following the first appearance of the mass in 1990, a magnetic resonance imaging study was read by the radiologist as a diffuse soft-tissue swelling over the dorsum of the foot involving an area 5 cm in diameter. There were no significant abnormalities of the osseous structures, and there was no involvement of the dorsal interosseous muscles or extensor tendons. Moreover, no definite focal mass or well-circumscribed fluid collection was found in the area. The patient was told to monitor the mass and report any changes that occurred.

For the next 4 years, the mass expanded slowly. The patient reported that the rate of enlargement increased rapidly during the 2 years prior to examination, and that in the past 3 months she had begun to experience pain in the mass and could feel “bumps” in it.

Clinical examination revealed a nontransilluminating tissue mass 5 to 8 cm in height and extending from the proximal phalanges to within 2 cm of the anterior ankle. The mass was 10 cm wide and extended from the medial to lateral borders of the foot. The patient’s primary-care physician described it as “a softball sitting on the top of her foot.” Multiple nodules were noted on palpation, which the patient reported as painful. Radiographic examination showed diffuse soft-tissue swelling over the entire dorsum of the foot. Surgical excision was discussed with and agreed to by the patient.

Surgical Methods and Findings

The patient underwent surgical excision of the soft-tissue tumor under intravenous and local sedation on December 13, 1996. A linear incision was made on the lateral aspect of the dorsum of the right foot, followed by sharp and blunt dissection. Two firm, nodular masses were noted in the subcutaneous layer of the foot; these masses were well demarcated from the skin as well as from the tendon sheath overriding the extensor digitorum tendons. The mass was followed proximally and distally, and continued to be well demarcated. The first group of nodules encountered measured approximately 6 cm in diameter; all of these nodules were removed.

Surgical exploration medially over the dorsum of the foot revealed another nodular mass in the same area. This well-demarcated mass was approximately 10 cm in diameter. Because of the width of the mass, a second incision was made on the medial aspect of the foot, dorsal to the first metatarsal. Sharp and

Figure 1. Angiectatic vessels showing subendothelial fibrin deposits (left), and formation of a lamellar collagenous cuff (right) (H&E, ×400).
blunt dissection was continued until the entire mass was removed intact. Some subcutaneous neurologic involvement was noted, and the nerve fibers were resected as necessary. The dorsalis artery of the foot was not involved and the mass did not appear to penetrate into the muscular or fascial layers of the foot. On gross examination, the lesion varied from a grayish-tan to a yellow color, and had a firm, multilobulated, rubbery feel on palpation.

**Pathology Findings**

Gross examination of the mass revealed a 7.5 × 5 × 1.5-cm grayish-tan to yellow-gold portion of tissue. Examination of a cross-section revealed a grayish-white to yellow-tan tissue with apparent gel-filled cystic spaces. Two other grayish-tan similar portions of tissue aggregated to 4.5 × 3 × 1.5 cm. A representative section from each portion was submitted for histologic examination.

Further examination of the tissue after staining revealed that approximately 80% of the gross section was myxoid and pale yellow in color. The remainder of the section was more firm and fibrous in consistency and grayish-white in color. Multiple additional representative sections were submitted for histologic examination.

Microscopic sections of the neoplasm showed that portions of adipose tissue had been replaced by a multinodular lesion with both myxoid and fibrous areas. The fibrous areas corresponded to the firmer portions of the lesion described grossly. These were composed of markedly pleomorphic spindle-shaped cells and occasional bizarrely shaped multinucleated giant cells (Fig. 2). Chromatin was coarse and irregular, and many cells had huge central nucleoli. Cytoplasmic and nuclear vacuoles were also found in the fibrous areas. These areas assumed a vaguely storiform pattern (Fig. 3). Occasional giant cells had a Touton pattern with xanthomatous cytoplasm. A few pigmented cells were also found, and they stained positive for iron. The remainder of the neoplasm was myxoid in character and somewhat less cellular. Individual cells had spindled-to-stellate configurations deep within a basophilic mucoid background (Fig. 4). Focal areas of tissue necrosis were identified. Many of these focal areas were surrounded by palisaded histiocytes that produced well-defined granulomas.

**Figure 2.** Pleomorphic spindle-shaped tumor cells with prominent intranuclear inclusions (H&E, ×400).

**Figure 3.** Low-power microscopic view showing a well-defined focus of myxoid change (left) within the tumor (H&E, ×100).

**Figure 4.** The focus of myxoid change consisting of delicate spindle-shaped cells within a fibromyxoid extracellular matrix. Note the presence of intranuclear inclusions similar to those shown in Figure 2 (H&E, ×400).
Scattered moderate-sized vascular channels were present throughout the lesion, some of which were partially occluded by fibrin thrombus (Figs. 5 and 6). Patchy infiltrates of lymphocytes and plasma cells were also present. Despite the degree of cytologic atypia, mitotic figures were rare (Fig. 7). The lesion became decreasingly cellular at its periphery, but did extend to the surgical margins. Special stains for fungal and acid-fast organisms were negative.

**Case Results**

Because pleomorphic hyalinized angiectatic tumor of soft parts has been only recently described, long-term behavior of the tumor is difficult to predict. The lesion of the patient in this report was considered a low-grade lesion that probably would not metastasize but might be locally aggressive. It was therefore determined, after surgery had been performed, that wider excision might be necessary to eradicate the tumor.

At 2 months after surgery, the patient reported a return to near-normal range of motion of the foot, with slight stiffness and numbness, following a regimen of prescribed physical therapy. The recommendation for wider excision of tissue into the extensor tendons in conjunction with creation of a muscular flap from a donor site to prevent regrowth and possible metastasis of the lesion was discussed. The patient expressed concern about the functional loss of the foot with such a radical resection. Further consideration by the physicians and the patient resulted in a decision to delay further surgery until there was evidence of regrowth.

**Conclusion**

A case of pleomorphic hyalinized angiectatic tumor of soft parts has been described. This tumor is a locally aggressive lesion with a low probability of metastasis. The differential diagnosis should include neurilemoma and malignant fibrous histiocytoma. Treatment consists of wide excision, but the tumor often recurs.

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**References**

21, 1996.


