A Case Report

Clear Cell Sarcoma Presenting as an Interdigital Neuroma

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ABSTRACT

The incidence of malignant soft-tissue sarcomas in the general population is approximately 1.4 per 100,000. Approximately 2% of all cancer deaths are attributable to these tumors. Presented is a case history of a soft-tissue malignant neoplasm that was originally thought to be an interdigital neuroma and that eventually required a modified Chopart's amputation. A review of the literature is presented on other soft-tissue malignant tumors that have an affinity for the foot and ankle. The intention of presenting this case is to caution physicians that malignant lesions can simulate a benign process and should always be considered in the differential diagnosis of any foot mass.

CASE REPORT

A 16-year-old high school cheerleader noted an insidious onset of pain in her left foot. Although very active in sports, she could not recall any trauma. After approximately 2 years of mild symptoms, she presented to an orthopaedic surgeon with sharp shooting pains in the sole of her foot. She noted these pains especially when standing. Plantar palpation between the second and third metatarsal heads and conventional roentgenograms (Figure 1) suggested a small, tender mass.

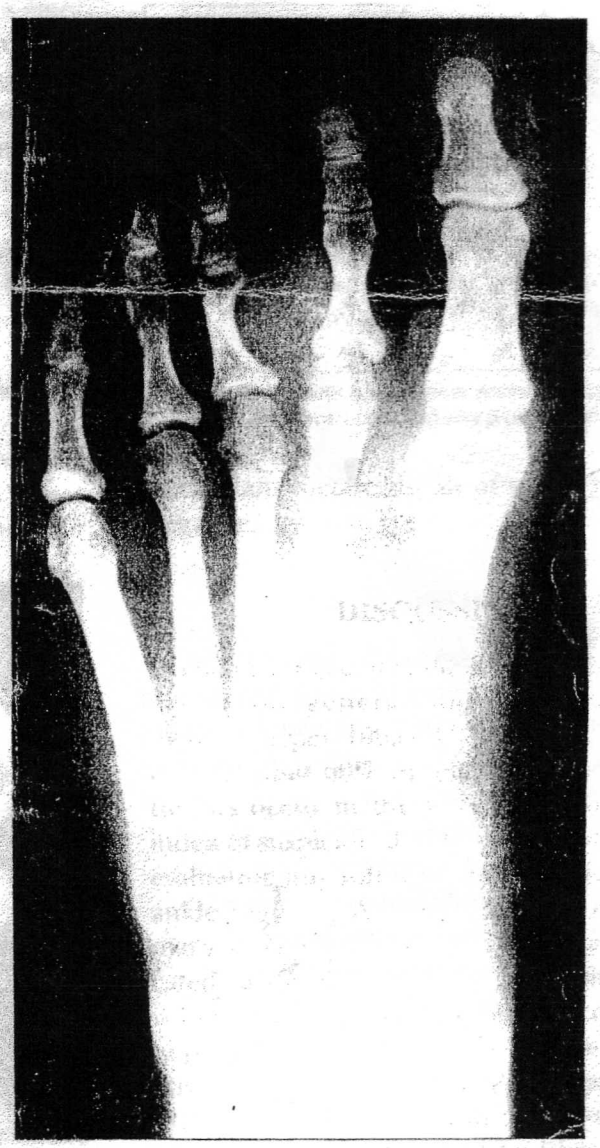

Figure 1. *A soft-tissue density is noted between the second and third metatarsal heads, causing slight divergence.*

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Tinel's sign over the area revealed nerve irritation. A mistaken diagnosis of an interdigital neuroma was made by the original treating orthopaedist. A computed tomographic (CT) scan and later magnetic resonance imaging (MRI) confirmed a mass in the second and third metatarsal interspace (Figure 2). A dorsal approach was then performed by the same orthopaedist to excise the neuroma. Pathologic evaluation revealed the mass to be a clear cell sarcoma (melanoma of soft parts). This diagnosis was later confirmed by combined light microscopic, immunocytochemical, and ultrastructure observations, as well as by electron microscopy. Photomicrographs of the biopsy specimen are shown in Figures 3 and 4.

After the patient was referred to our care, a chest roentgenogram and CT scan of the chest were performed and were negative for metastasis. We performed a modified Chopart's amputation to ensure adequate margins around the tumor. The patient is currently pain free at 2 years postsurgery and is able to run and walk on rough terrain with her prosthesis. Ankle and subtalar motion are equal to her opposite side (Figure 5), and she has returned to high school cheerleading. Follow-up examinations at 6-month intervals have included chest roentgenograms and body bone scans to rule out distant metastasis, all of which have been negative.

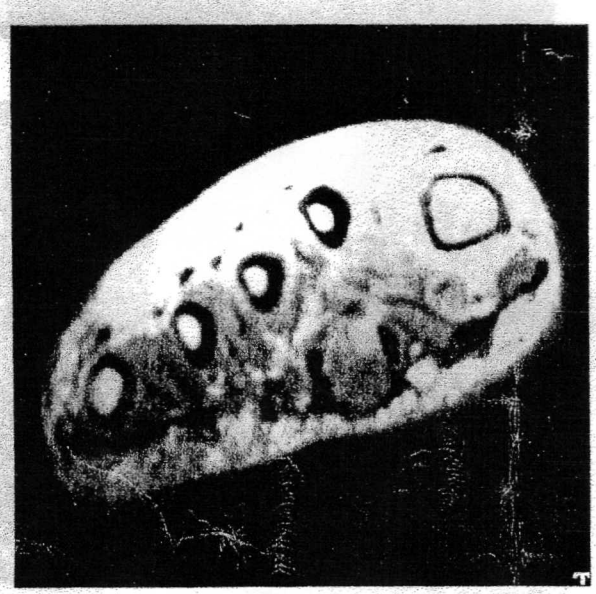

Figure 2. *T-2 weighted MRI images reveal a 1-cm mass deep to the plantar fascia.*

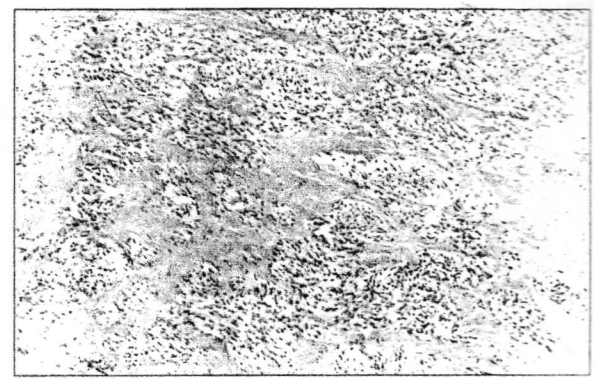

Figure 3. *Within the fibrous stroma are nests and fascicles of round or spindle-shaped cells with clear or faintly eosinophilic cytoplasm (80×).*

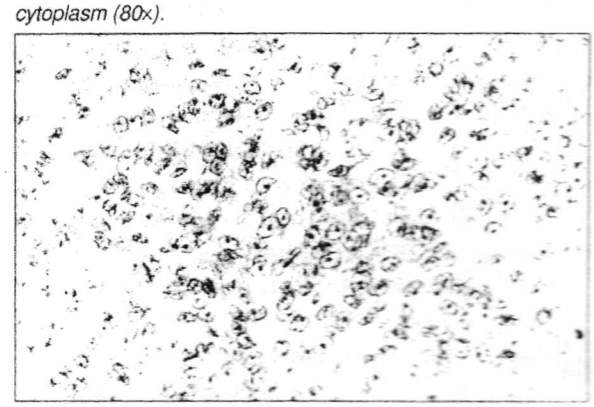

Figure 4. *Tumor cells have clear or eosinophilic cytoplasm and characteristically oval nuclei with very prominent nucleoli (625×).*

DISCUSSION

The incidence of malignant soft-tissue sarcomas in the general population is approximately 1.4 per 100,000.[1] Since some sources estimate that 60% of all malignant soft-tissue tumors occur in the lower extremity, a high index of suspicion should be maintained when evaluating any soft-tissue mass in the foot and ankle region.[2] Although only 8% of the primary sarcomas discovered every year are located in the foot,[3] any physician with a large volume of foot and ankle cases is likely to have exposure to this problem. By performing Bayes-Rule analysis, Kirby et al reported the most common sarcoma in the foot to be synovial sarcoma, followed by fibrosarcoma, malignant fibrous histiocytoma, rhabdomyo-

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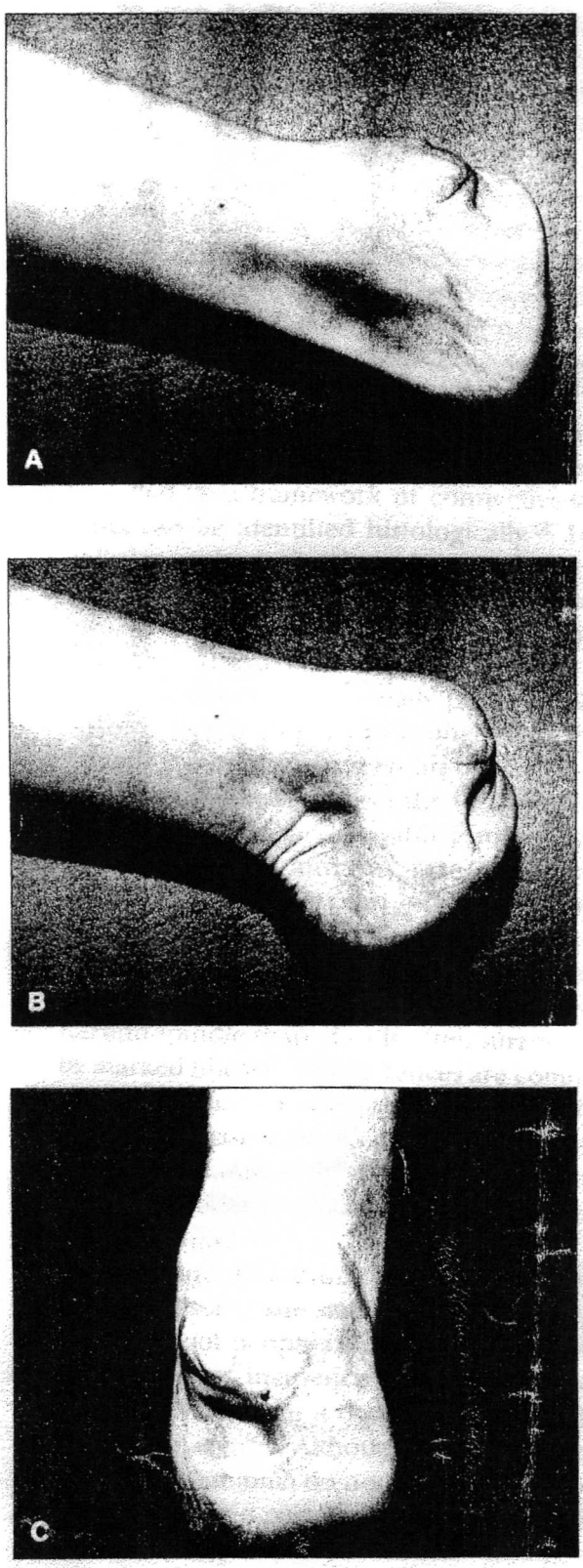

Figure 5. *Excellent range of motion is noted with the modified Chopart's amputation. Maximal dorsiflexion (A) and plantar flexor (B) are shown. No valgus deformity is apparent (C).*

sarcoma, and liposarcoma.[4] An abbreviated list of primary soft-tissue sarcomas that have an affinity for the foot appears in the Table.

Synovial cell sarcoma presents as a slow-growing deep mass, and only slightly over half are painful.[5] Ninety percent occur in patients under 50 years of age. They tend to occur in para-articular regions, close to tendon sheaths, bursae, and joint capsules.[6] They are often misdiagnosed as synovitis, bursitis, or arthritis. The Armed Forces Institute of Pathology reported 345 cases from 1970 to 1979, 22.5% of which occurred in the foot, ankle, and lower leg. They appear as round or oval masses on roentgenograms, and less than a third of the cases have focal calcifications.[7]

The classic type of synovial sarcoma is biphasic, composed of both epithelial cells and spindle cells.[2] A monophasic form composed only of fibrous-like spindle cells is an important histologic subgroup, although it has a similar clinical behavior to the classic form. The overall 5-year survival rate is 40%.[8]

Fibrosarcoma usually presents as a slow-growing solitary palpable mass. It usually remains painless until its size encroaches on other structures. Forty percent of the cases occur in the lower extremity, and the ages of patients usually range from 30 to 55 years.[9,10] Fibrosarcoma appears histologically with a fasciculated growth pattern and fusiform or spindle-shaped cells, often in a herringbone pattern.[2] The survival rate of patients is 41% to 54%.[11]

Malignant fibrous histiocytoma is the most common soft-tissue sarcoma of adults aged 50

Table. Soft-Tissue Sarcomas With an Affinity for the Foot

Synovial sarcoma
Fibrosarcoma
Malignant fibrous histiocytoma
Rhabdomyosarcoma
Liposarcoma
Clear cell sarcoma
Epithelioid sarcoma
Malignant schwannoma

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to 70 years.[7] This usually also presents as a painless mass that the patient has noticed for several months. This tumor shows great histologic variation. Most are cellular neoplasms composed of plump spindled and pleomorphic cells, often arranged in a storiform pattern.[2] The 5-year patient survival rate is 36%.[12]

Clear cell sarcoma (malignant melanoma of soft parts) presents as a slow-growing, deep-rooted mass intimately positioned next to tendons or aponeuroses. Pain and tenderness are noted in approximately half of the cases. This tumor is usually seen in patients 20 to 40 years of age. Compact fascicles of fusiform cells surrounded by a framework of connective-tissue septa can be identified histologically.[13] Intracellular melanin is often detected with the Fontana stain or by ultrastructural examination.[2,14] Tumors have recurred or metastasized as long as 10 years after radical excision.[15]

Epithelioid sarcoma usually presents insidiously as a painless mass in young adults.[16] Although it is most often seen on the hands and fingers, it can be found on the feet 9% of the time. Epithelioid sarcoma also tends to invade tendon sheaths and migrate proximally. Speckled calcifications can sometimes be seen on roentgenograms.[16,17] If it is situated on the dermis, epithelioid sarcoma can be confused with an "infected wart." It appears as polygonal and spindle-shaped cells often surrounded by marked fibrosis.[2] Recurrences are common if adequate margins are not maintained. Wide or radical excision of this malignant neoplasm gives acceptable control. Regional lymph-node dissection is indicated in the presence of palpable nodes.[17,18]

Malignant schwannoma comprises about 10% of all soft-tissue sarcomas. It is a primary malignancy of peripheral nerves and therefore has its highest incidence about major nerve plexuses. Most often it occurs in patients 20 to 50 years of age.[19-25] Although patients with malignant schwannoma do not always present with pain, beware of a painful mass in a patient with von Recklinghausen's disease. Malignant schwannoma is associated with neurofibromatosis, presenting in 50% of cases.[23,25,26] This tumor has a remarkable ability to spread through nerve sheaths; one must take frozen sections of the nerve to ensure clean margins. Histologically these tumors resemble fibrosarcomas, but they also maintain features of the normal Schwann cell.[2,25] For a solitary mass, the 5-year patient survival rate is 50%.[24,25]

Kirby et al stated that factors such as the gender of the patient, a history of trauma, the duration of symptoms, the presence of pain or of neurological symptoms, and the size of the lesion are not useful discriminators between malignant and benign lesions.[4] Probably the best approach to diagnosing any mass in the foot, in addition to conventional roentgenography, is either a CT scan or MRI. If the lesion appears to be primarily bony in nature, CT is the most appropriate study. Magnetic resonance imaging is helpful in soft-tissue sarcomas not only to help determine the extent of the lesion but also to help differentiate benign from malignant lesions.[27] If these studies are suggestive of a sarcoma, a total-body bone scan, chest roentgenograms, and a CT scan of the chest are indicated because of the high probability of the lesions metastasizing to the lungs. As always, a careful assessment must be made for metastasis of regional lymph nodes, the liver, and other bony sites.

The histopathologic classification of soft-tissue tumors can be difficult, especially in a group of lesions that have spindle cells as a common component. Knowledge of exact anatomic location, age of patient, and adequate sampling of the tumor is essential. At times, a precise pathologic diagnosis may require additional studies, such as immunohistochemistry or electron microscopy, that require special handling or fixation of the tissue. For this reason, consultation with a pathologist before or at the time of surgery can be invaluable in ensuring that all necessary procedures can be performed on the biopsy material.

Paramount in cancer surgery is close attention to the location and technique of the biopsy.[28] In the case presented, the mistaken preliminary diagnosis of an interdigital neuroma could have been disastrous if the sur-

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geon had made a different approach. Some surgeons have advocated a plantar approach for a primary interdigital neuroma, because they believe this affords them better visualization. In this case, however, a plantar approach might have violated the plantar compartments, prohibiting amputation at Chopart's joint. Malignant lesions must be treated by wide excision or amputation, and contamination of other compartments must be avoided.[8,15,17,29-31]

The surgeon performing the definitive procedure should, whenever possible, also perform the biopsy. Neoadjuvant and adjuvant chemotherapy, as well as postoperative radiotherapy, may be indicated, depending upon the grade and type of tumor.[2,32,33] Follow-up examinations must include physical examination and chest roentgenograms at 2- to 3-month intervals for 2 years, then every 6 months for 1 year, and then annually.

CONCLUSION

Although rare, soft-tissue malignancy must be considered for any mass in the foot and ankle region. This is particularly worrisome, since most of these lesions have no characteristic findings on conventional roentgenography. Clearly some tumors have an affinity for the foot and ankle. In the case presented, the appearance in a teenage patient of an interdigital neuroma between the second and third metatarsal interspace is very unlikely and should have raised suspicion. Atypical masses should be approached carefully to avoid discovering these sarcomas after a biopsy of a presumed benign mass.

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