CHAPTER 19

BENIGN OSSEOUS TUMOR OF SOFT TISSUE

Luke D. Cicchinelli, D.P.M.
Manuel González San Juan, Podólogo
Javier Aycart Testa, Podólogo

Heterotopic bone formation is a well-documented entity that classically occurs in large muscle groups such as the thigh or shoulder. The occasional presentation of a psuedomalignant or non-neoplastic osseous lesion in the soft tissues of the foot is often referred to as myositis ossificans or its variants. Two recent podiatric articles thoroughly discuss the pedal manifestations of this localized disorder.[1,2] After reviewing these articles and other references, several points needed to be emphasized.

The authors will present a case in which the clinical, radiographic and surgical evidence is consistent with non-traumatic myositis ossificans. As an example of the paradoxes of this disorder, the histological diagnosis by one pathologist was chronic myositis ossificans with additional characteristics of an osteochondroma. A second pathologist reported a diagnosis of osteochondroma with additional post-traumatic changes.

Myositis ossificans is a form of heterotopic ossification and must be differentiated from dystrophic and metastatic calcification. Dystrophic calcification is the most common type. It is localized and associated with decreased carbon dioxide tension as seen in tissue degeneration and areas of decreased blood supply. Common examples include arteriosclerosis, phleboliths, calcification of heart valves, tendons, skin and subcutaneous tissues as seen in scleroderma. Metastatic calcification involves the deposition of calcium salts in tissue that has not been previously traumatized. This is a disease process secondary to disturbed calcium metabolism in systemic conditions such as hyperparathyroidism or chronic renal insufficiency. Common examples include renal calculi and calcified basal ganglia. Foci of abnormal calcification can stimulate local transformation of fibroblasts to osteoblasts giving rise to heterotopic bone formation.[3]

CLASSIFICATION

Noble described the first system of classification for myositis ossificans which was later redefined by Paterson.[3,4] *Myositis ossificans progressiva* is a metabolic disorder that appears shortly after birth and results in heterotopic ossification of muscles. The disease often spreads to multiple muscles and is usually fatal in childhood. The lesions begin as soft, tough masses that eventually become ossified. Biopsy and surgical trauma may aggravate the condition and hasten the patient's demise. The disorder is also known as fibrodysplasia ossificans progressiva or Munchmeyer's disorder.

Myositis ossificans circumscripta is non-neoplastic heterotopic bone formation without an associated history of trauma. It is associated with neuromuscular disorders, chronic infections, severe burns, and poliomyelitis. In a sense, these etiologies can be considered a form of soft tissue trauma, although they are distinct from a direct-impact traumatic event. This condition can be seen in paraplegics without a history of trauma or any histological evidence of hematoma formation, and is also known as "paraosteoarthropathy."[1]

Traumatic myositis ossificans circumscripta is a non-neoplastic heterotopic bone formation associated with a single major episode of trauma, or repetitive episodes of micro-trauma. This is the most common form of myositis ossificans and accounts for approximately 60% of all cases. Other soft tissue structures may be involved including fascia, subcutaneous fat, aponeuroses, tendons or periosteum. This form is characterized by hemorrhage followed by fibrosis and granulation tissue formation. Cartilage may subsequently form and lead to endochondral ossification, with osteoblasts being formed by surrounding pluripotential mesenchymal cells or from adjacent periosteum.

An additional classification system proposed by Gilmer and Anderson focuses on sites of bone

formation rather than histogenesis.[5] Lesions are classified as *extra-osseous* (within muscle and without direct connection to bone), *periosteal* (discrete bone attached to the underlying bone), and *pariosteal* (most often found within muscle against the shaft of a long bone).

CASE REPORT

A 66-year-old female presented with vague medial arch discomfort in the left foot, as well as hallux valgus and hammertoe deformities. The duration of the medial arch symptomatology was two years, and bothered the patient when shoes were worn, or direct pressure applied.

Physical examination revealed mild fullness of the medial arch and a firm palpable mass plantar to the first metatarsal shaft. The mass was not attached to the skin and seemed embedded in the intrinsic musculature. Standard radiographs revealed an ossified, well-organized mass with lacy calcifications throughout its entire dimension. It was not possible to determine from the plain films whether the mass was attached to the first metatarsal (Figs. 1A, 1B). A computed tomography (CT) scan depicted a clear gap between the tumor and the adjacent metatarsal. A zone of mature bone was present at the periphery of the lesion. The central portion demonstrated some linear areas of ossification, but was largely free of mature bone (Fig. 1C).

The patient denied any history of antecedent trauma to the affected area. A provisional diagnosis of myositis ossificans secondary to chronic, insidious micro-trauma was made, and surgical excision was recommended. The mass was easily removed through a medial longitudinal incision since it was freely mobile and not attached to any surrounding structures. The patient healed uneventfully and was without any symptoms or signs of recurrence at one year postoperative.

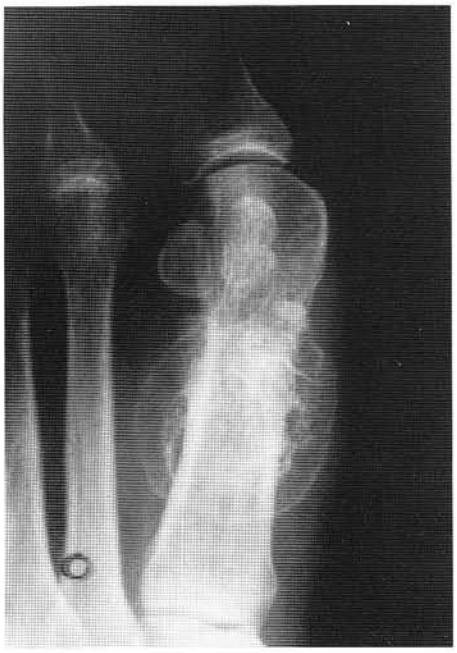

Figure 1A. Dorsoplantar view of a well-delineated ossific mass inferior to the first metatarsal. It is not clear if the mass originates from the metatarsal on this plain film.

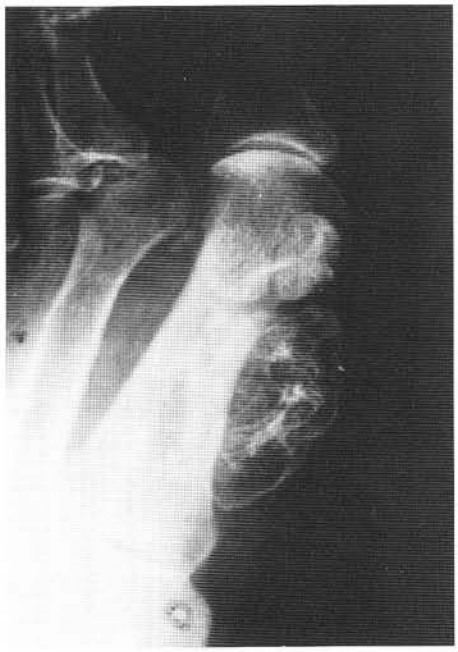

Figure 1B. Oblique view of the mass.

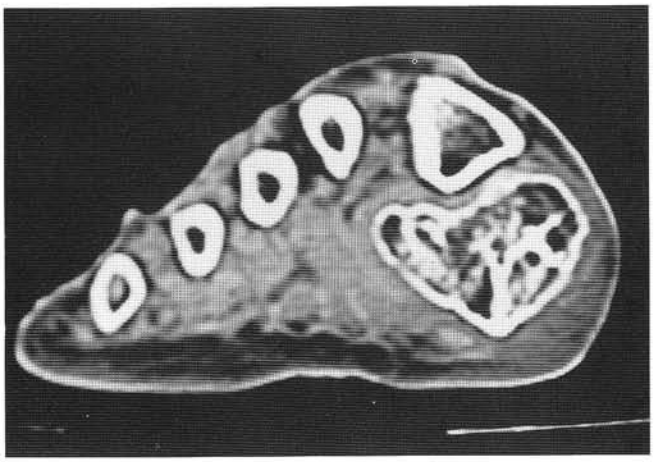

Figure 1C. The CT scan shows the mass to be separate from the metatarsal and posses a well-formed peripheral cortex.

PATHOLOGY

Macroscopically, the mass was a grayish-white, irregular ovoid shape measuring 3.7 x 2.7 x 2.2 centimeters in its largest dimensions (Fig. 1D). Upon sectioning, it consisted of a peripheral rim of cartilaginous and osseous tissue, and a soft, yellowish center. Microscopically, the tumor exhibited an external surface made-up of well-formed osseous and connective tissue. In some areas this exterior was covered with a hyaline cartilaginous hood that had osseous trabeculations advancing toward the interior. A transitional zone of diminishing maturity was present between this periphery and the center of the lesion. The central portion of the lesion consisted of mature adipose tissue and vessels (Fig. 1E).

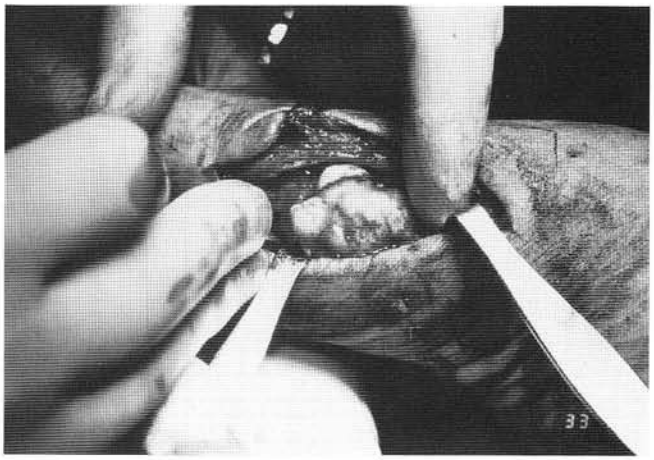

Figure 1D. Gross appearance of the mass.

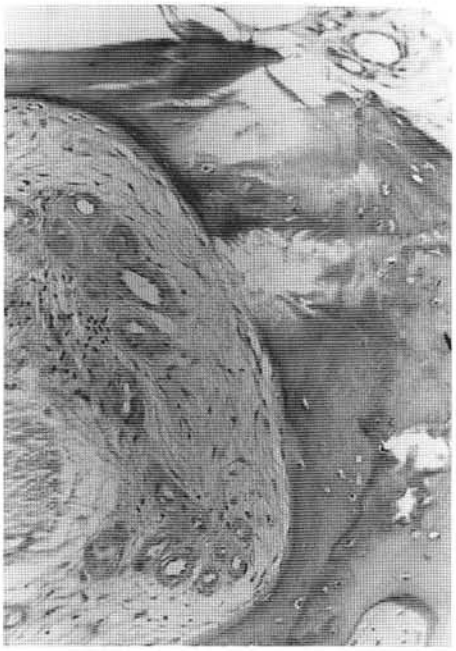

Figure 1E. (40X) Microscopic specimen showing well-differentiated peripheral bone and an immature central zone. (Courtesy of Dr. Jose Montans Araujo)

DISCUSSION

The most important point to emphasize concerning mysotitis ossificans is that it may mimic a malignancy. Depending on the stage of the disease process, there are clinical, radiographic and histological similarities to neoplasms such as osteosarcoma. In acute cases, the aggressive onset and rapid development may resemble a malignant tumor.[6] Furthermore, a history of trauma is not necessary for the formation of myositis ossificans nor is location in muscle. Wakely et al. reported that up to one third of cases are located in subcutaneous tissue rather than deep skeletal muscle.[7] According to various authors, between one-third and one-half of cases of non-neoplastic heterotopic bone formation within soft tissue develop without a history of trauma.[8,9]

The plain-film characteristics of myositis ossificans may also make it difficult to distinguish it from a malignancy. Initial radiographic changes are seen in approximately 2 to 3 weeks as increased soft tissue density. Flocculent densities of calcification and ossification occur in 6 to 8 weeks and then encyst and mature in 5 to 6 months. The lesions often then decrease in size leaving a radiolucent area between the tumor and the underlying normal cortex.[10,11] The most common differential diagnoses include extra-skeletal sarcoma, parosteal or surface osteosarcoma, and fibrosarcoma because the radiographic changes are a progression and may represent any or all of these conditions at some point in time.[11-14] Soft tissue osteosarcoma should be considered until myositis ossificans is fully confirmed.[12]

Two distinct radiographic signs of non-neoplastic heterotopic bone formation are the presence of a lucent line between the lesion and the cortex of adjacent bone, and peripheral calcification. The clear band of separation between neoplastic and normal bone is very suggestive of a benign lesion, but may be difficult to see on plain

films. It is also important to remember that aggressive malignant osseous tumors commonly ossify centrally rather than peripherally.[6,9]

CT scans aid in the identification of the zonal phenomenon and improve the accuracy of diagnosis of myositis ossificans.[8,15] A lucent line between the mass and normal adjacent bone is readily seen on CT scan. In the case presented, the CT scan clearly showed the separation of the osseous mass from the first metatarsal and confirmed the presence of the peripheral rim of calcification. This is in contrast to osteochondromas which routinely show a stalk of attachment to the originating bone (Figs. 2A-2C).

Other cases of heterotopic bone formation will be less clear with plain films or CT scans, and the diagnosis is made only by biopsy (Figs. 3A, 3B). Bone scans are a further radiological study that may detect myositis ossificans prior to plain films but they suffer from poor specificity. A recent article describes the benefit of serial bone scans in determining the maturity of osseous tumors located in the soft tissues.[10]

Confusion in the microscopic diagnosis occurs in the interpretation of early-stage biopsies or pieces of tissue in which complete maturation has not yet occurred.[15] If only the center of the lesion is taken in a biopsy, diagnostic problems arise for the pathologist. The cellularity, mitotic activity and infiltrative spread in early stages, also termed "reactive mesenchymal proliferation," may easily mimic malignancy.[9,11] Wakely et al. state "it is well recognized that if only the central core of myositis ossificans is sampled, the distinction between it and sarcoma is nearly impossible." This potential diagnostic dilemma exists for aspiration cytology as well as open biopsy, which makes fine needle aspiration somewhat controversial due to the chance of misdiagnosis.[7,8]

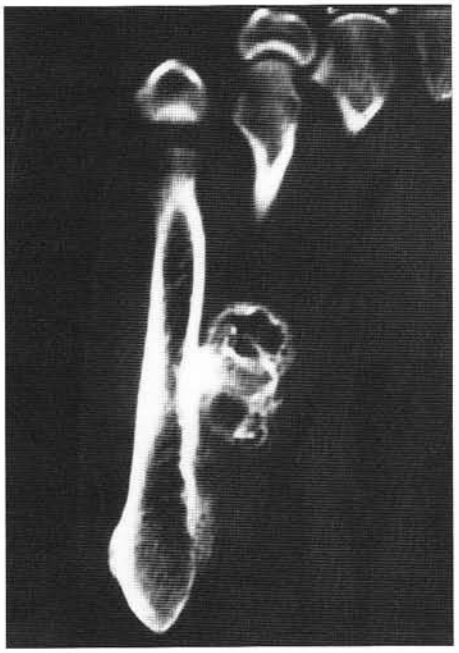

Figure 2B. The CT scan clearly shows a "stalk" of attachment, highly suggestive of an osteochondroma. Excisional biopsy confirmed the diagnosis.

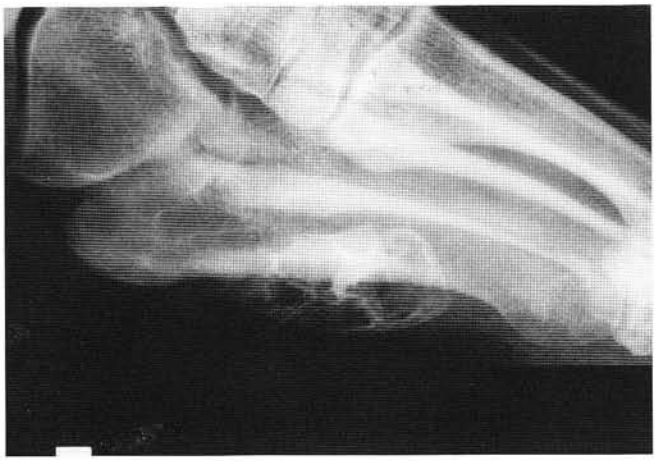

Figure 2A. A similar ossific mass plantar to the fifth metatarsal shaft. This had been slow growing for over a year, with a gradual increase in plantar pain upon ambulation. Plain films do not show whether the mass originates from the metatarsal.

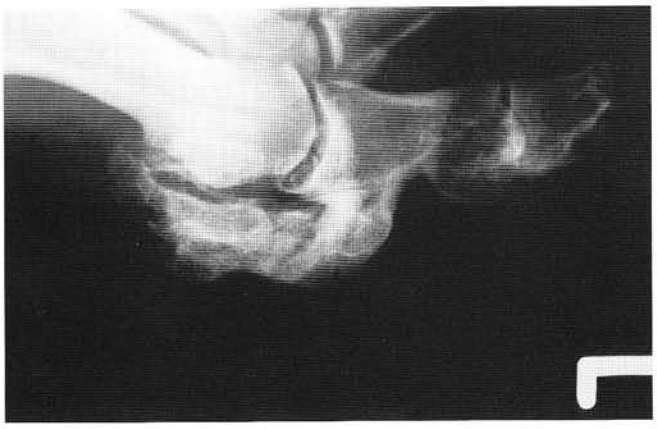

Figure 2C. This is an osteochondroma clearly originating from the plantar aspect of the base of the proximal phalanx.

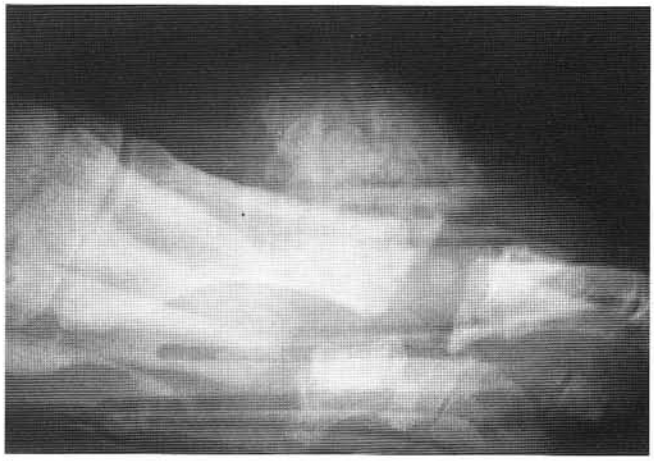

Figure 3A. Exuberant bone formation in a diabetic female that formed between six and twelve weeks after a panmetatarsal head resection.

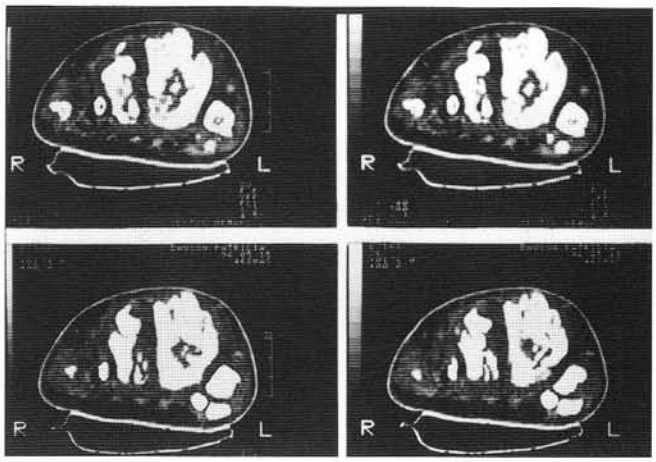

Figure 3B. The CT scan shows the osseous mass circumferentially around the second and third metatarsals. Surgical exploration and removal confirmed "exuberant, reactive bone formation" with a foci of fractures through the second and third metatarsal shafts. The autonomic neuropathy and hypervascular state accounted for this benign bone formation in the soft tissues.

The classic histological finding in myositis ossificans is the zonal phenomenon originally described by Ackerman.[17] The central zone consists of undifferentiated mesenchyma with high-grade mitotic activity, and an overall sarcomatous picture can exist. The intermediate zone is composed of immature osteoid in a fibromuscular background and in the peripheral zone, mature lamellar bone is noted with a more mature fibrous stroma.[15,17] Clearly, the correct diagnosis of myositis ossificans requires an adequate and representative sample of histological material from the entire lesion.

Arteriography has occasionally been discussed as having a potential use in the evaluation of heterotopic bone formation, particularly when plain film radiographs are atypical for myositis ossificans. However, even arteriography exhibits confusing characteristics such as hypervascularity that closely resembles a malignant neoplasm. Recently, Tamura et al. described the finding of arteriovenous shunting in a case of myositis ossificans, a finding formerly thought to be seen solely in malignant processes.[14]

Recent treatment recommendations stress the importance of understanding that although myositis ossificans may have an aggressive onset, it is benign and self-limiting. This non-neoplastic process requires a conservative approach and cautious observation.[6,11] Spontaneous regression and rapid subsiding of symptoms has been observed on occasion, prompting some authors to avoid surgery and evaluate the condition with time.[7,8] Conversely, since it is a non-neoplastic condition, the likelihood of cure if it is excised at a mature stage is great.[11] General recommendations are to observe the condition and watch for the zoning phenomenon or shrinkage.[12] If surgical excision is contemplated, it should be performed when the lesion is mature. CT scans will help plan the timing and performance of the procedure.[17] Resection at an immature stage may lead to confusion with malignancy or reoccurrence. Overly aggressive approaches and false diagnoses have led to radiotherapy, radical resection and amputation in the treatment of myositis ossificans.

Occasional reports exist of malignant transformation of myositis ossificans. However, there is substantial speculation regarding whether the initial diagnosis of myositis ossificans in such cases was accurate. Also there are questions about the role of previous treatments such as radiation therapy in the formation of the malignancy. A recent case report of malignant myositis ossificans exemplifies this.[18] This case actually represents an ossifying soft tissue metastasis from an occult gastric adenocarcinoma primary and not the transformation of benign myositis ossificans to malignancy. Nuovo et al. summarize by saying "the rarity of so-called malignant transformation of myositis ossificans, plus tenuous verification under close scrutiny in some cases, emphasizes its isolated occurrence, if it exists at all."[11]

CONCLUSION

The benign nature of this non-neoplastic process requiring a conservative approach cannot be sufficiently stressed. In those clinical, radiologic, and/or histologic presentations in which the diagnosis of myositis ossificans may be entertained but is questionable, a reasonable period of observation is mandatory. It is also stipulated that, if the mass does not manifest zoning maturation and shrinkage as would be anticipated for myositis ossificans, other diagnoses should then be considered.[11] Surgical excision of a mature lesion may be curative.

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