Soft-Tissue Benign Mesenchymoma in a Pediatric Patient

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Abstract

Soft tissue mesenchymoma is an exceptionally rare and benign neoplasm. Since its initial description in 1962, only 7 cases of cartilage predominant appendicular mesenchymoma have been reported. Of these, only 2 patients were skeletally immature. We report the clinical, pathologic, and radiologic features of a 13-year-old adolescent boy with a benign mesenchymoma in the distal leg.

Benign mesenchymomas are exceptionally rare tumors composed of 2 or more nonepithelial mesenchymal elements not usually found together in tumor tissue. We report the clinical, pathologic, and radiologic features of a 13-year-old adolescent with a chondrolipomatous mesenchymoma in the distal leg. Furthermore, we offer a literature review to summarize the current experience of this uncommon tumor as a means of facilitating diagnosis and guiding treatment.

The patient and the patient's guardian provided written and informed consent for print and electronic publication of this case report.

Case Report

A 13-year-old African American adolescent boy presented with a swollen, painful ankle after a recent twisting injury sustained while playing sports. Medical history did not contribute to the injury, and the patient and mother indicated no prior awareness of any abnormality or mass in the injured extremity. Physical examination revealed a large fixed painless mass on the anterior compartment of the distal leg measuring 10 cm in length by 6 cm in width. The mass was slightly tender to palpation, and the overlying and surrounding skin appeared normal. Range of motion at the ankle was unrestricted and symmetrical, compared with the contralateral side. The patient had strong dorsalis pedis and posterior tibial pulses, which were symmetrical. He had 5/5 motor strength in dorsiflexion and plantarflexion of the ankle with sensation intact in the first dorsal web space, dorsal, and plantar aspects of the foot.

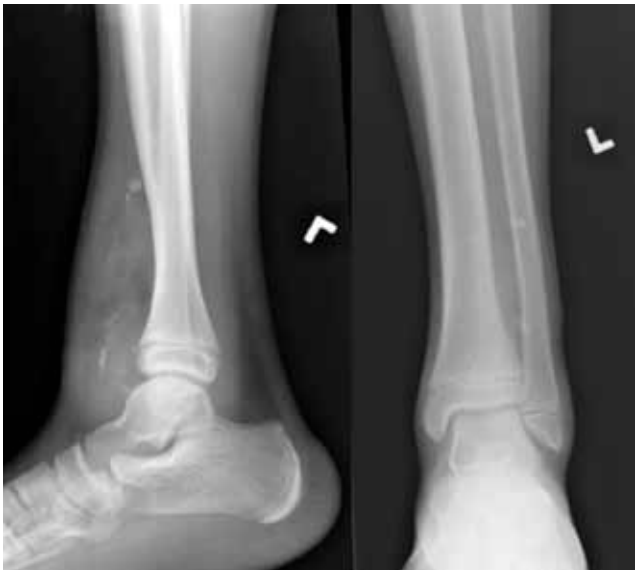

Figure 1. Radiographs demonstrated a fat containing soft tissue mass with multiple calcification and adjacent cortical scalloping of the underlying anterior cortex of the distal tibia.

Radiographs demonstrated a soft tissue mass that was hypodense compared to the surrounding muscle with adjacent cortical scalloping of the underlying, anterior cortex of the distal tibia (Figure 1). Multiple calcifications were also present in the mass. The calcifications did not demonstrate the typical appearance of phleboliths. The cortical scalloping of the anterior margin of the distal tibia that was present suggested a long-standing pathologic process.

Magnetic resonance imaging (MRI) evaluation with and without contrast revealed a heterogenous soft-tissue mass with approximate dimensions of 10 x 5.8 x 3 cm (Figure 2). Large portions of the lesion were composed of tissue identical to fat in signal intensity on the T1- and T2-weighted pulse sequences. These areas also demonstrated signal loss with frequency selective fat suppression. On the T1-weighted images, other portions

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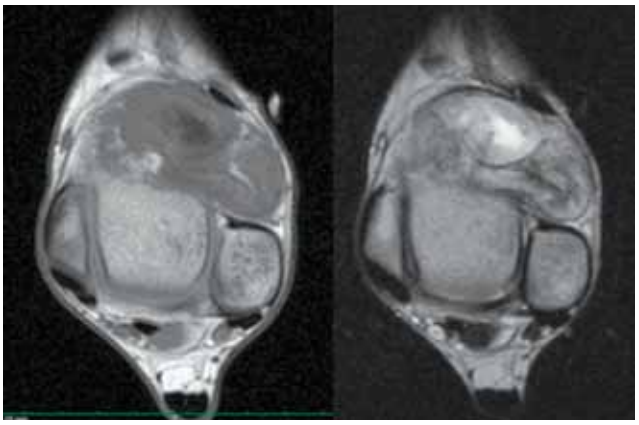

Figure 2. T1- and T2-weighted pulse MRI sequence demonstrating a heterogeneous post-contrast enhancement predominantly in a serpentine peripheral and septal fashion, with areas of nodular enhancement.

of the lesion were similar in signal intensity to skeletal muscle whereas some portions were lower in signal intensity than skeletal muscle. These very low signal intensity areas seen on the T1-weighted pulse sequences showed very high signal intensity on the T2-weighted pulse sequences both with and without fat suppression, indicating that the lesion contained fat. Other regions of the lesion were intermediate in signal intensity, higher than skeletal muscle but lower than fluid in signal intensity, on the non-fat suppressed T2 weighted pulse sequence. The mass demonstrated heterogeneous post-contrast enhancement predominantly in a serpentine peripheral and septal fashion, although some areas demonstrated nodular enhancement. The majority of the lesion did not demonstrate significant postcontrast enhancement. No periosteal reaction was identified and there was no surrounding peritumoral or intraosseous edema or bone marrow reaction.

A computed tomography (CT) scan deemed T was necessary to further define the mass and rule out osseous involvement. To minimize the number of interventions on this pediatric patient, and the risk of neurovascular injury due to possibly distorted anatomy, a CT guided biopsy was performed via an anterolateral approach. The biopsy was performed with the collaboration of the attending surgeon such that the biopsy tract was in the ultimate plane of resection. Multiple coaxial core biopsy specimens were obtained with a 14-gauge Temno Evolution soft tissue biopsy device (Carefusion, San Diego, California). The CT revealed a heterogenous intermuscular mass with multiple coarse calcifications. On CT, the lesion demonstrated large areas of fat attenuation, similar to skeletal muscle in attenuation as well as areas lower in attenuation than skeletal muscle. No osseous invasion was identified. Histological examination of the biopsy specimen demonstrated areas of chondroid, fibromyxoid, and adipose tissue. No sarcomatous elements were demonstrated.

Once final pathology was obtained and the diagnosis confirmed, the patient underwent definitive surgical treatment. Frozen section was not performed. This diagnostic tool would have been included if the mass in question was deeper seated or if the definitive diagnosis was less clear. A marginal surgical excision was performed as the mass was a benign lesion and was easily separated from underlying normal tissue (Figure 3). Tumor size was similar to the previously mentioned estimated measurements. Grossly, the tumor appeared to have a smooth, but irregular surface, and the mass was easily freed from its soft tissue bed with minimal blunt dissection. Once removed, the mass was transected, revealing islands of hyaline cartilage separated by yellow fatty tissue (Figure 4). Postoperative surgical pathology confirmed a benign mesenchymal neoplasm (10 x 3.5 x 3 cm) with cartilage predominate tissue (80 %) (Figure 5). Additionally, there were mixed fibroadipose, myxoid, hemangiomatous, and osseous elements. No sarcomatous elements were identified.

No postoperative complications developed. The patient began full immediate mobilization. Four weeks postoperatively, the patient was able to play in recreational athletics. After 14 months, there was no evidence of tumor recurrence.

Discussion

In 1948, Stout coined the term *mesenchymoma.*[1] Although this original description inappropriately described a collection of 8 malignant mixed mesenchymal tumors, this misnomer does reflect the confusion with regard to the accurate nomenclature of this pathologic entity. Possible elements include muscle, fat, blood vessels, cartilage and/or myxomatous tissue. This variability of tissue presence and predominance has further added to the challenge of consistent nomenclature.

True soft tissue benign mesenchymomas are rare occurring benign tumors. In actuality, benign mesenchymomas are an unrelated entity to malignant mesenchymomas. Benign mesenchymomas do not degenerate into malignant mesenchymomas. Malignant mesenchymomas are more commonly referred to as *sarcomas,* and this term denotes a specific mesenchymal de-differentiation of a specific mesodermal phenotype.

Controversy does exist regarding the true nature of benign mesenchymomas. Although currently neither recognized nor defined by the World Health Organization classification of soft-tissue tumors, benign mesenchymomas have been labeled a hamartoma due to the presence of abnormal mixtures of normally occurring tissue commonly present in the mass. However, some of the potential constituents of benign mesenchymomas include hyaline cartilage, a tissue not normally found in soft tissue. For this reason, as well as the possibility, although unlikely, of reoccurrence,[2] some clinicians instead classify benign mesenchymomas as a neoplasm. Regardless, agreement exists that benign mesenchymomas have no relation to malignant mesenchymomas.

In 1962, Le Ber and Stout reported 39 pediatric cases of what they considered to be true benign soft tissue mesenchymoma.[3] Interestingly, only 2 of these patients demonstrated chondroid predominance. Only 15 cases involved the upper or lower extremity, and only 5 cases involved muscle. Of the appendicular tumors described, none had chondroid tissue present.

Since the description by Le Ber and Stout,[3] only 7 additional cases of hyaline cartilage predominant appendicular mesenchymomas have been reported[4-8] and of these, only 2 patients were skeletally immature.[5,8] These case reports report anatomic distribution of benign mesenchymoma in the forearm,[4] palm,[5] hip,[6] knee,[6] heel,[6] thigh,[7] and sole of the foot.[8] After surgical excision, by either wide or marginal resection, tumor recurrence of chondroid predominant benign mesenchymoma never occurred.

Although early descriptions identified mesenchymomas as a pediatric tumor, in actuality, it may occur in patients of all ages. Mesenchymomas typically present as a painless mass. Morbidity occurs as a consequence of local infiltrative growth. Radiography may show calcifications or ossification within the mass. MRI will usually show a well-circumscribed heterogenous mass with a prominent pseudocapsule. Scalloping of abutting bony cortex may be observed.

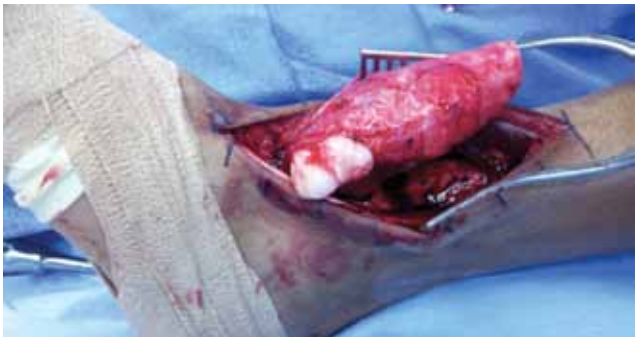

Figure 3. Gross specimen in-utero after surgical dissection.

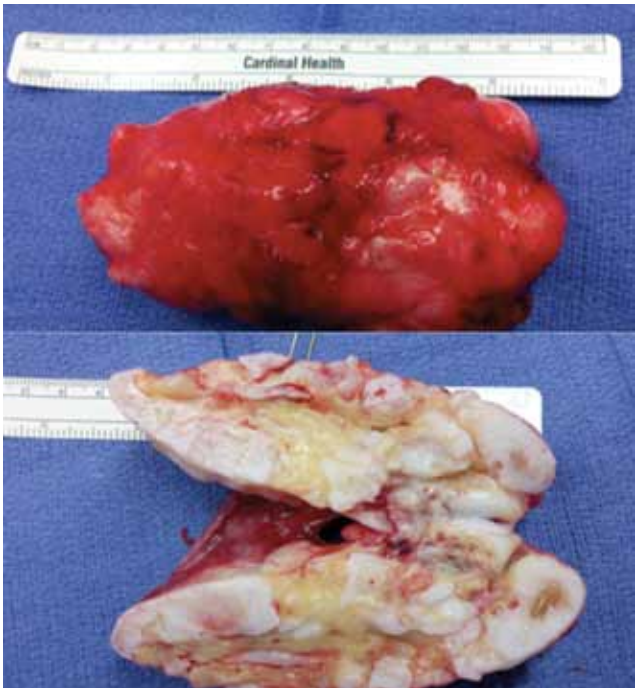

Figure 4. Gross specimen after transection.

The differential diagnosis for this soft-tissue mass includes extraskeletal chondroma, periosteal chondroma, synovial chondromatosis, chondroid lipoma, and skin adnexal mixed tumors/chondroid syringomas. Our histopathology demonstrated a lesion composed of intersecting bundles of spindle cells and collagen fibers. Well-defined areas of benign hyaline cartilage were prominent throughout. It was this mixture of mesenchymal elements that was the essential histologic feature of this entity that identified it as a benign soft-tissue mesenchymoma.

The diagnosis of extraskeletal chondroma with metaplastic components was considered. These lesions have been described with histological fat components.[9] Atypical to these lesions, we demonstrated the presence of several mesenchymal components, including hemangiomatous areas. Furthermore, extraskeletal chondromas typically have osseous components demonstrating enchondral ossification, however, the osseous component encountered in our lesion demonstrated showed a haphazard distribution. For these reasons, we believe the diagnosis of a benign mesenchymoma is more appropriate for this unusual mesenchymal neoplasm. Our patient's presentation is typical for a hyaline cartilage containing appendicular mesenchymoma. Initial radiographic investigation revealed a large soft tissue mass containing several large calcifications. The imaging characteristics of the lesion initially raised the concern for a malignant lesion such as myxoid liposarcoma. Although these tumors are very rare in the adolescent age group they have been reported.[10] The radiologic consideration of synovial cell sarcoma, which may show calcifications, was excluded from the differential diagnosis due to the presence of adipose tissue within the mass.

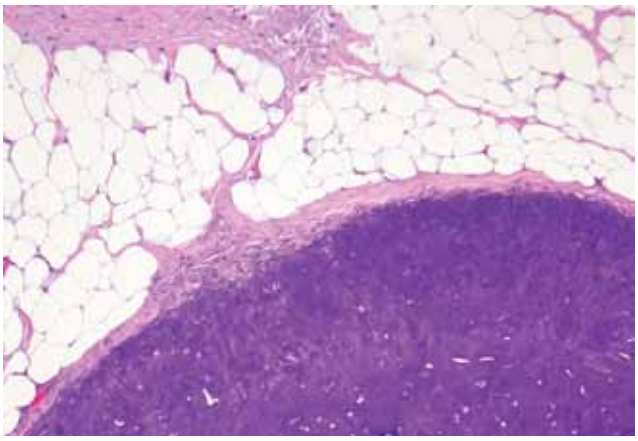

Figure 5. Histologic specimen demonstrating cartilage predominate tissue, with limited mixed fibroadipose, myxoid, and osseous elements.

The tibial scalloping reflected the presence of a slow growing mass, a characteristic also shared with synovial cell sarcoma. Certainly, the ramifications of this misdiagnosis could have been profound. Definitive tissue diagnosis was obtained prior to operative intervention, and thus marginal excision was deemed appropriate and has been proven curative in this case.

Benign mesenchymoma are slow growing but are potentially locally infiltrative tumors that have no risk of malignant degeneration. Due to the variability in tissue components, clinical and radiographic examination may erroneously suggest underlying malignancy. We describe the clinical, radiologic, and pathologic findings of mesenchymoma, and offer this review as a reminder to clinicians of this rare but treatable pathologic entity.

Authors' Disclosure Statement

The authors report no actual or potential conflict of interest related to this article.

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This paper will be judged for the Resident Writer's Award.