



Test yourself: question: “painless right leg swelling”

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Diagnosis

Ossifying subperiosteal hematoma of the right tibia.

Discussion

Subperiosteal hematomas are a unique, but infrequently seen, entity associated with type 1 neurofibromatosis. Although 50% of patients with type 1 neurofibromatosis exhibit musculoskeletal manifestations, patients more commonly present with progressive scoliosis or the classic tibial psuedarthrosis [1]. The less common subperiosteal hematoma causes anxiety at clinical presentation, as it can be mistaken for an enlarging neurofibroma concerning for malignant transformation [2]. Fortunately, subperiosteal hematomas have characteristic radiologic findings that can separate it from the more concerning differential considerations.

Due to the underlying mesodermal dysplasia associated with type 1 neurofibromatosis, the periosteal tissue is weakened, allowing it to separate from the underlying bone with an associated subperiosteal hematoma [2–5]. This can be identified on plain radiography, as shown in

Fig. 1, with periosteal new bone formation along the affected bone diaphysis, which typically appears within 10–14 days upon onset of the mass [4]. In addition to plain radiography, ultrasonography is a relatively inexpensive and widely accessible modality that can be used in further characterization. Ultrasound characteristically demonstrates a sharply marginated, elliptical mass beneath the elevated periosteum tightly apposed to the cortical surface with internal heterogeneous lace-like echogenicity typical of an organizing hematoma (Fig. 2). The addition of color Doppler can be used to further evaluate the vascularity of the mass to differentiate subperiosteal hematomas, which classically lack intrasubstance flow, from neurofibromas undergoing malignant transformation, which would display varying degrees of intrasubstance neovascularity.

Although other imaging modalities have been described in the evaluation of subperiosteal hematomas, magnetic resonance imaging (MRI) has emerged as the imaging study of choice in the evaluation of a musculoskeletal mass [1]. The signal characteristics of a hematoma vary depending on the age and stage of evolution of the hematoma at time of MR evaluation. As seen in this case, the MR reveals the classic presentation with the subperiosteal mass demonstrating signal iso-intense to hyper-intense as compared to adjacent skeletal muscle on the T1 weighted images (Fig. 3). The T2 images demonstrate the sharp margination of the stripped periosteum with the lamellated mixed signal of the hematoma (Fig. 4). Gradient echo sequences often add additional information demonstrating blooming artifact related to hemosiderin deposition within the mass. Reactive edema surrounding the hematoma may be robust, as seen in the case shown. If contrast is administered, periosteal and subperiosteal new bone enhancement would be expected in the healing response, but the hematoma itself would not enhance.

Once appropriately diagnosed, treatment for subperiosteal hematomas is largely conservative with

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observation and activity modification. It is important to counsel patients and families that the areas of swelling representing the subperiosteal hematoma will largely remain unchanged in appearance as the lesion undergoes interval ossification but may remodel over time as the child grows.

Compliance with ethical standards

Conflict of interest Each author certifies that he or she has no commercial associations that might pose a conflict of interest in connection with the submitted article.

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