Idiopathic Equinovarus Foot Deformity in an 8-year-old Girl

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History and Physical Examination

An 8-year-old girl presented to our clinic with a left foot deformity. According to the parents, the girl's foot was normal in appearance until they noticed a prominence on the dorsal aspect of her foot while trying on shoes approximately 1 year previously. The patient also had progressive difficulty with walking owing to frequent inversion injuries, and problems with shoe wear attributable to the development of dorsal foot callosities. Her only other medical history was vesicoureteral reflux, for which she underwent surgery when she was 8 years old.

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Each author certifies that his or her institution has approved the reporting of this case report, that all investigations were conducted in conformity with ethical principles of research, and that informed consent for participation in the study was obtained.

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Her physical examination revealed a normal-appearing right foot but an equinovarus deformity of the left foot (Fig. 1). Coleman block testing was normal, indicating a flexible hindfoot deformity. There was no clawing of the toes or intrinsic atrophy. There was a 1.5-cm decrease in left calf circumference measured 15 cm above the medial malleolus compared with the right. Her neurologic examination showed normal patellar tendon and Achilles tendon reflexes, full strength (5/5) of the posterior tibia, anterior tibia, and triceps surae musculature, and normal sensation. No skin stigmata or spinal asymmetry or deformity were observed.

We obtained radiographs (Fig. 2) and MR images (Fig. 3) of the foot; MR images of the brain and spine; radiographs (Fig. 4), CT scans (Fig. 5), and MR images (Fig. 6) of the pelvis; and a whole-body bone scan.

Based on the history, physical examination, and imaging studies, what is the differential diagnosis?

Imaging Interpretation

Radiographs of the left foot showed an equinovarus deformity and a mixed lytic and sclerotic lesion with dense lobular areas of calcification (Fig. 2). MRI revealed a multifocal stellate lesion in the calcaneus, which was low signal on T1-weighted (Fig. 3A) and T2-weighted (Fig. 3B) images and did not enhance with gadolinium (Fig. 3C). MR images of the brain and spine were normal.

The whole-body bone scan showed mild radiopharmaceutical uptake in the left ischium, left proximal femur, and left calcaneus. No other foci of abnormal uptake were evident.

Radiographs of the pelvis showed an area of sclerosis in the left ischium, and intrapelvic densities attributable to

Fig. 1A–C (A) Anterior, (B) posterior, and (C) lateral preoperative photographs of the left foot show the equinovarus deformity of the foot.

Fig. 2A–C (A) Anteroposterior, (B) oblique, and (C) lateral radiographs of the left foot show the equinovarus deformity of the foot and a mixed lytic and sclerotic lesion with dense lobular areas of calcification in the calcaneus. There is no associated periosteal reaction.

residual contrast (Fig. 4). CT scan of the pelvis revealed a densely sclerotic focus in the left ischium beginning in the cortical bone and extending into the medullary space (Fig. 5). MRI of the pelvis showed the left ischial lesion was low signal intensity on T1-weighted (Fig. 6) and T2-weighted images. No bony expansion, cortical destruction, periostitis, or associated soft tissue mass was observed.

Differential Diagnosis

- Hemangioma
- Chronic recurrent multifocal osteomyelitis
- Osteoma
- Enostosis
- Osteopoikilosis
- Melorheostosis

Neurosurgical consultation and evaluation did not reveal any evidence for central nervous system dysfunction or disease to include diastematomyelia, myelomeningocele, lipomeningocele, anterior horn cell disease, tight phylum, cerebellar disease, or intraspinal tumor. Subsequently, she underwent surgical correction for her foot deformity owing to progressive difficulties with ambulation. Tissue specimens from the flexor hallucis longus and posterior tibial tendon were sent for histologic analysis.

Based on the history, physical findings, imaging studies, and histology, what is the diagnosis?

Histology Interpretation

Grossly, both biopsy specimen (left flexor hallucis longus sheath, posterior tibial tendon/fascia) were composed of white-tan firm tissue with homogeneous white-tan cut surfaces. Microscopic examination of both specimen showed benign dense fibrocollagenous and intermixed adipose tissue, with focal synovial lining associated with the posterior tibial fascia and the flexor hallucis longus sheath (Fig. 7). Biopsy of the calcaneal lesion was not performed, as we believed it was a benign-appearing lesion and because we believed it would have been difficult to accurately localize the lesion for biopsy.

Diagnosis

Melorheostosis

Fig. 3A–C (A) Sagittal T1- and (B) T2-weighted MR images show a multifocal stellate lesion of the calcaneus, which is predominantly low signal on both images. (C) A sagittal T1-weighted fat-suppressed

MR image after intravenous gadolinium administration shows the lesion to be nonenhancing.

Fig. 4 An anteroposterior radiograph of the pelvis shows a sclerotic focus in the left ischium, and multiple intrapelvic densities from residual contrast material.

Fig. 5 A densely sclerotic lesion in the left ischium without any bony expansion or cortical disruption is seen on this axial CT image.

Fig. 6 An axial T1-weighted MR image shows a well-defined low-signal-intensity lesion in the left ischium, without any associated soft tissue component.

Discussion and Treatment

An 8-year-old girl presented with a progressive left foot deformity and multiple sclerotic bone lesions. The differential diagnosis included hemangioma, chronic recurrent multifocal osteomyelitis, osteoma, enostosis, osteopoikilosis, and melorheostosis.

We eliminated chronic recurrent multifocal osteomyelitis, as affected patients typically present with the triad of bone pain, fever, and multiple bone lesions. With the exception of the foot deformity, our patient was

Fig. 7 Benign dense fibrocollagen with intermixed adipose tissue (Stain, hematoxylin and eosin; original magnification, $\times 40$) is seen on this photomicrograph of the flexor hallucis longus biopsy specimen.

asymptomatic, never manifesting a fever or complaining of pain. Hemangioma of bone also was ruled out, as these vascular lesions show up as high signal intensity on T2-weighted images whereas our patient's MRI showed multiple low signal intensity lesions on T2 imaging. Osteoma, enostoses, and osteopoikilosis were ruled out based on the plain film imaging. Osteomas appear as smooth hemispheric densely corticated bone attached to the bone's cortex. Our patient's lesions were in the cancellous bone separated from the overlying cortical surface. Enostoses and osteopoikilosis appear as round or oval well-circumscribed lesions in the cancellous bone. This patient's lesions were neither round nor oval, appearing instead as irregular and densely sclerotic.

Melorheostosis is a rare, nonfamilial, and typically unilateral bone dysplasia leading to a peculiar hyperostosis of the cortical bone. Its etiology is unknown. Leri and Joanny [9] were the first to describe it in 1922. Affected bone displays wavy layers of hyperostosis alternating with osteopenic or normal bone in a pattern "resembling melted wax dripping down one side of a candle" [12]. It usually involves the periosteum and endosteum of long bones but also has been reported less commonly in the vertebrae, ribs, skull, and jaw [2, 5, 8, 11, 13, 16]. Although most patients have only one affected extremity, it is not unusual to have lesions in multiple extremities. Only approximately 400 known cases have been reported worldwide, with an overall prevalence estimated at one per million [6, 14].

The disease typically is discovered in childhood or adolescence, with a peak presentation between 5 and 20 years. Pain is a prominent initial complaint in adults but frequently is absent in children until late adolescence. Affected children usually present with soft tissue contractures and fibrosis, often leading to a substantial limb-length inequality [15]. Sometimes the joint contracture occurs before any radiographic evidence of bony changes. These contractures result from bone deformity, soft tissue fibrosis, and periarticular calcification.

Radiographically, children do not usually have the extracortical or subperiosteal hyperostosis that leads to the classic "melted wax" appearance seen in adults [15]. Instead, the hyperostosis is seen as either a patchy sclerosis or a linear, endosteal streaking. Bone scans show increased uptake with asymmetric cortical activity that may cross joints but are highly nonspecific. MRI shows areas of decreased signal intensity on all pulse sequences [7].

Fig. 8A–C (A) Anteroposterior, (B) oblique, and (C) lateral radiographs of the left foot 4 years after surgery show mild residual cavus and hindfoot varus. The calcaneal lesion is stable, remaining unchanged from preoperative imaging.

Histopathologic examination shows no pathognomonic features. The histologic analysis shows osteosclerosis with abnormal proliferation of compact Haversian bone distorting the periosteal and endosteal surfaces. The Haversian canals are surrounded by thick and irregular laminae. The bone can be woven, lamellar, or both. Periosseous fibrosis of the soft tissues and fibrosis of the skin and subcutaneous tissues are common, as was seen in our patient.

Treatment of this disorder is geared toward the relief of symptoms. Bone pain or pain resulting from soft tissue inflammation and synovitis is managed with NSAIDs, narcotics, steroids, or bisphosphonates. The goal of surgery is often correction of the deformity, as these contractures are fairly resistant to manipulation and serial casting. Surgical treatments have included extensive soft tissue releases, capsulotomies, and osteotomies, but recurrence is common, sometimes necessitating amputation [3, 15]. Some authors have reported successful results using the Ilizarov technique for limb lengthening, correction of joint contracture, and foot deformity correction [1, 4, 10].

The clinical course of this rare entity is slow but persistent progression into adulthood. The bony changes are benign and do not affect prognosis. Malignancy, such as osteosarcoma, has been reported but is exceedingly rare [15].

Our patient underwent surgical correction consisting of a radical plantar release, posteromedial release, and lengthening of the tendoachilles and posterior tibial tendons. Operative findings included abundant dense scar-like tissue extending from above the medial malleolus to the medial midfoot, and a thickened and nodular flexor hallucis longus tendon and contracted posterior tibialis tendon. These are interesting findings, as patients with melorheostosis frequently have extremity problems attributable to soft tissue contractures and fibrosis.

We believe this is the first documented case of an equinovarus deformity developing as a result of melorheostosis. The patient's recovery was uneventful and at her 4-year postoperative visit she was doing well. She does have residual cavus and hindfoot varus but is asymptomatic and active in ballet and soccer without any limitations. The left calcaneal lesion has remained unchanged on the most recent plain radiographs (Fig. 8).

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