Giant Bone Island of the Tibia in a Child

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Abstract

Giant bone islands have been previously diagnosed only in adults. A symptomatic giant bone island of the tibial diaphysis in a 10-year-old boy is presented in this case report. Imaging investigation included plain radiographs, scintigraphy, computed tomography, and magnetic resonance imaging. Although the findings suggested a giant bone island, the appearance of several atypical imaging features necessitated an open biopsy. Histology confirmed the diagnosis of a giant bone island. Its length, which exceeded 10.8 cm, is the longest reported in the literature. No additional surgical intervention was required. The clinical symptoms and signs, as well as the radiographic appearance of the lesion, were not changed after 15-year follow-up.

A bone island is a focus of normal cortical bone located within the medullary cavity. The vast majority of bone islands are small, measuring from 1 mm to 2 cm in size. They are found more frequently in adults than in children. The lesion can be virtually diagnosed on the basis of its characteristic clinical and imaging features. Differential diagnosis may be difficult when the lesion manifests itself uncharacteristically by being symptomatic, very large, and hot on bone scan.[1-4]

The term *giant bone island* has been used to describe a large lesion[1] that measures more than 2 cm in any dimension.[5] Giant bone islands have been described only in adults,[1,5-15] and the longest bone island length reported is 10.5 cm.[10] They are usually symptomatic and associated with increased radionuclide uptake on bone scintigraphy.[14]

The history and the clinical and imaging presentation of an even longer, symptomatic, and scintigraphically hot lesion in the tibial diaphysis of a 10-year-old boy is reported. The lesion further exhibited several atypical imaging features necessitating an open biopsy, which confirmed the diagnosis of a giant bone island. The pertinent differential diagnosis and the clinical and radiographic findings after 15-year follow-up are also presented and discussed. The patient provided written informed consent for print and electronic publication of this case report.

Case Report

A 10-year-old boy was admitted for surgical repair of an inguinal hernia. Physical examination revealed a painless but tender anterior bowing of the right tibial diaphysis. The patient was a healthy-appearing white male with normal vital signs, gait, and posture. His parents noticed a slight protuberance of the tibia at age 2.5 years. No medical advice was asked for the bone swelling after that time. After recovery from the inguinal hernia repair 3 weeks later, the bone lesion was thoroughly examined. Radiographs showed an oblong, homogenous region of dense sclerosis in the diaphysis of the right tibia. The lesion had relatively well-defined margins and was located in the medullary cavity. Speculations were not obvious in the periphery of the lesion, which exhibited a sharp circumscription (**Figures 1A, 1B**). A well-defined lytic area was evident at the distal part of the lesion (Figure 1B). There was no periosteal reaction. Blood and serum chemistries were within normal limits, including serum calcium, phosphorus, and alkaline phosphatase. A conventional 3-phase bone scintigraphy (300 MBq) with technetium-99m HDP (hydroxydiphosphonate) indicated increased uptake in the area of the lesion but no other skeletal abnormality (**Figure 2**). Computed tomography (CT) showed that the lesion was purely intramedullary and densely blastic. The lesion originated from the medial cortex, which was thickened (**Figure 3A**). The lesion extended to the anterolateral cortex, which was thinned and includ-

Figure 1. (A) Anteroposterior and (B) lateral radiographs of the right tibia show a large intramedullary region of sclerosis with sharp circumscription.

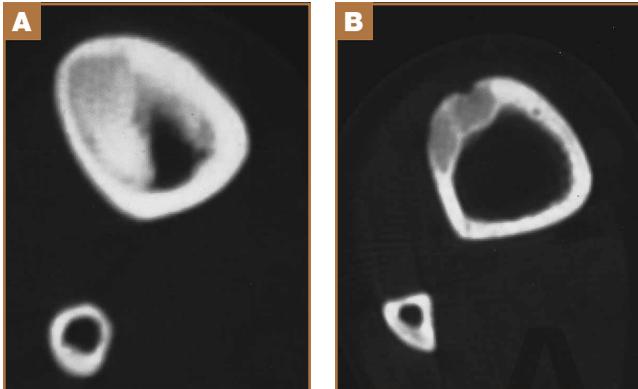Figure 2. Bone scintigraphy shows increased uptake of the lesion.

Figure 3. Axial computed tomography scan shows a hyperdense intramedullary lesion. (A) The lesion originated from the medial cortex, which was thickened, and extended to the anterolateral cortex, which was thinned. (B) At the distal part of the lesion, the anterior cortex was thickened, included lytic areas, and showed an anterior area of destruction.

ed a lytic area. In the distal part of the lesion, the anterolateral cortex was thickened, included lytic areas, and exhibited an anterior portion of cortical destruction (**Figure 3B**). The fatty marrow adjacent to the region of sclerosis appeared normal. There was no evidence of extraosseous soft-tissue changes. On both T1- and T2-weighted magnetic resonance imaging (MRI), the lesion exhibited low-signal intensity. The lesion measured 10.8×2.2×1 cm. It originated from the medial cortical bone of the tibia, blended into the medullary cavity, and extended anteriorly towards and through the anterior cortex. The area of cortical destruction was clearly evident on the axial MRI. The periosteum

Figure 4. T1-weighted sagittal magnetic resonance imaging shows an intermediate-signal intensity focus at the distal part of the lesion.
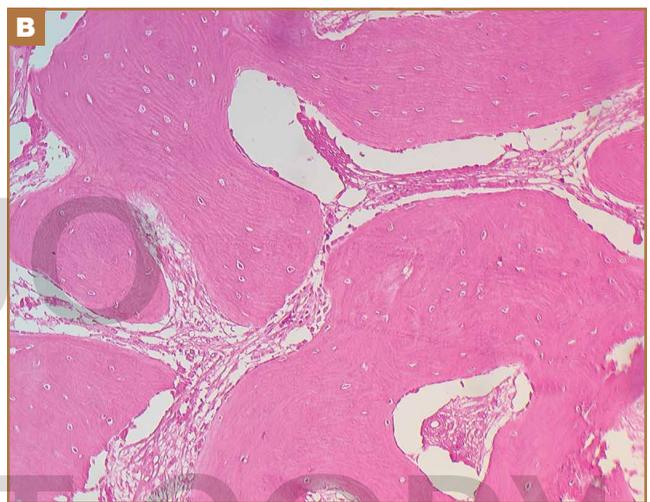

Figure 5. Microscopic features. (A) Low-power photomicrograph, at the level of soft-tissue attenuation on computed tomography imaging, indicates dense, compact woven bone with long, projecting surface processes within the fat of adjacent cancellous bone (magnification ×2.5). (B) Higher magnification of photomicrograph in Figure 5A shows woven bone containing interstices of fibrous vascular tissue with a linear rimming of osteoblasts (magnification ×10).

was displaced and eroded anteriorly by focal radiating bony streaks. No enhancement was seen after the intravenous administration of gadolinium-diethylenetriamine pentaacetic acid (Gd-DTPA) as a contrast medium. There were no extraosseous soft-tissue changes. In the distal part of the lesion, sagittal and axial MRI showed a 1.2×0.8×0.7-cm well-defined ovoid focus, with characteristics of cystic degeneration that exhibited intermediate-signal intensity on T1-weighted MRI (**Figure 4**) and high-signal intensity on T2-weighted MRI.

An open biopsy was performed. Macroscopically, a wedge of compact bone measuring 3×1.7×0.6 cm was taken. Microscopic examination showed a thinned periphery of lamellar (mature) bone with haversian canals and, beneath it, woven (immature) bone with long-surface processes projecting within adjacent cancellous bone (**Figure 5A**). The woven bone

Figure 6. (A) Clinical and (B, C) radiographic presentation at 15-year follow-up.

contained loose vascular fibrous tissue. No osteoclasts were noted, and the very few osteoblasts lining the bone trabeculae were small, single-layered, and flat (**Figure 5B**). There was no evidence of neoplastic cells. There was no abnormality of the periosteum and the surrounding soft tissues.

The histology was pathognomonic of a giant bone island. No additional surgical intervention was recommended.

The postoperative course was uncomplicated, and the patient was discharged 2 weeks later. An above-the-knee plaster was recommended for 3 months and a below-the-knee splint for an additional 2-month period. Full weight-bearing was allowed only after the postsurgical sixth month to prevent an impending fracture. The tibial bowing was tender to pressure or palpation, and the patient reported mild spontaneous pain during follow-up. Radiographs 1 year after surgery indicated that the bone area removed for biopsy was replaced by compact bone. MRI performed 4 years after surgery showed that the volume of the lesion in relation to the host bone was not changed.

At the last follow-up 15 years after surgery, the anterior tibial bowing was not changed (**Figure 6A**), but the patient additionally complained of skin irritation after intense training wearing boots during military service. The radiographic appearance of the lesion was also not changed, while the periphery of the lesion exhibited scarce radiating bony streaks with rounded contours (**Figures 6B, 6C**). The clinical symptoms and signs from wearing military boots completely subsided after a couple of weeks' rest from daily army activities, but the mild spontaneous pain and the local tenderness over the tibial bowing persisted.

Discussion

Giant bone islands are more likely to be associated with clinical symptoms than the usual small-sized bone island. Some degree of pain was detected in 8 of 10 patients with a giant bone island presented in the literature, but it was induced by trauma in 3 of them.[14]

Radiographic appearance is among the distinguishing diagnostic features of a giant bone island. It appears as an ovoid,

round, or oblong, homogenously dense, single or multiple focus of sclerosis within the medullary cavity; it is oriented along the long axis of the host bone, and it exhibits peripheral pseudopodia or radiating spicules producing the typical "thorny" or "paintbrush" appearance.[8,16,17] It does not exhibit cortical penetration and it is not associated with periosteal reaction.[10]

The CT findings include a sclerotic and hyperdense focus with spiculated margins extending into the adjacent cancellous bone. The lack of bone destruction and soft-tissue mass are also diagnostic.[3,7] MRI findings will reflect the low-signal intensity characteristics of cortical bone on all pulse sequences.[18]

Enostoses usually exhibit no activity on skeletal scintigraphy, while giant lesions generally show increased radiotracer uptake.[5,9-11,14,19-27] The latter may result from the increased amount of bone turnover, which is seen more often with larger lesions because of active bone deposition and remodeling.[20,21,23,28]

Histopathology of a giant bone island appears identical to the well-described pathologic appearance of smaller bone islands. The lesion is composed of compact lamellar bone and haversian systems, which blend with the adjacent spongiosa. The surrounding cancellous bone forms thorn-like trabeculae radiating from the lesion and merging with the cancellous bone.[1,4,5,8,28]

The presumptive diagnosis of a bone island is based on the clinical findings, imaging features, and follow-up examinations. An asymptomatic, isolated, sclerotic bone lesion showing the typical features of a bone island on plain radiography, CT, and MRI, whatever its size, that is nonactive on bone scan may be easily diagnosed. However, a symptomatic patient with a hot lesion on scintigraphy should be carefully observed. In addition, larger lesions may raise the suspicion of a neoplasm, such as a sclerotic variant of osteosarcoma. In such cases, an open biopsy may be undertaken. No specific treatment is required after the diagnosis has been confirmed. There is no literature to suggest that, after adequate biopsy confirmation, excision or resection is necessary. Follow-up radiographic examination of the lesion should be suggested to monitor for any potential growth.[2,10,23]

The first giant bone island appearing in a child is presented in this report. The lack of a causative factor leading to the anterior tibial bowing indicated that the bone deformity was caused primarily by the lesion. The present case is unusual for the appearance of several atypical features, some of which have not been previously described. Peripheral radiating spiculated margin was absent on the patient's initial radiographs and CT imaging. MRI indicated only the presence of radiating bony streaks that displaced and eroded the periosteum on the anterior border of the lesion. The CT findings that the lesion likely originated or was in close proximity with the medial cortex of the tibia were also atypical. It has been previously reported that spinal lesions located immediately below the cortex tend to fuse with the endosteal surface, while similar features may also be seen in the appendicular enostoses.[4,29] Other CT findings, such as the thinning of the overlying anterolateral cortical bone, as well as the cortical thickening at the periphery of the lesion associated with areas of soft-tissue attenuation and anterior cortical destruction, have not been described even in the atypical features of a giant bone island. The lytic area resembling a nidus that was evident

at the distal part of the lesion was more likely consistent with an area of resorption, which, although rare, has been described on giant lesions.[2,9,29] The substantial amount of woven bone transforming to lamellar bone that was evident in the present patient's microscopic features is also an atypical finding, although it may be expected to some degree in scintigraphically hot, large lesions.[28] The clinical and imaging progress of the lesion supported the diagnosis of a giant bone island. The degree of the anterior tibial bowing and the volume of the lesion in relation to the host bone were not changed throughout the follow-up period, indicating that the growth of the lesion followed the growth of the normal bone.

The differential diagnosis of a giant bone island includes a variety of benign tumors and tumor-like lesions, as well as malignant bone lesions.[2,4,23,28,30,31] In the patient presented in this report, the diagnosis of an atypical sclerotic presentation of a nonossifying fibroma or healing stage of this lesion could be consistent with some of the CT findings, including the eccentric origin from the cortex associated with medial cortical thickening, the anterolateral cortical thinning, and the soft-tissue attenuation of cortical areas. In addition, unifocal osteofibrous dysplasia may also present with a long intracortical diaphyseal lucency within an area of marked cortical sclerosis and cause a bowing deformity. Both diagnoses were excluded, since no fibrous stroma was evident on the histologic examination of the lesion. A large or giant long-bone osteoma would be associated with the outer cortical margin of bone but would not involve the intramedullary space. The scintigraphically increased uptake of radioisotope, as well as the CT and MRI findings, were not consistent with the diagnosis of osteoid osteoma, osteoblastoma, or osteomyelitis. Although most imaging findings were consistent with a benign lesion, and contrast-enhanced MRI showed no increased vascularity, anterior cortical disruption necessitated a bone biopsy to rule out any potential malignancy.

The histopathology in association with the clinical and imaging findings indicated the diagnosis of a giant bone island. The increased proportion of maturing woven bone over lamellar bone indicated an active remodeling lesion that could be related to the patient's age, since the clinical and radiographic features of the lesion were not changed after 15-year follow-up.

Conclusion

This is the first giant bone island diagnosed in a patient before puberty. Its greatest length was 10.8 cm, which is the longest reported in the literature. The imaging appearance included several atypical features that are very rare or have not been reported. Microscopic features indicated less mature lamellar bone and a prominent proportion of maturing woven bone. The clinical and the radiographic appearance of the lesion were not changed after 15-year follow-up.

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