We report here a case of Ribbing disease, the diagnosis of which became possible due to bone scan.

CASE REPORT

A 31-year-old female presented to an orthopedician with bilateral leg pain since 1 year. Pain started in the right leg 6 months before the left leg, and at the time of presentation she had persistent pain on both sides, more on the right. She had no other site of pain. There was no history of fever, trauma or undue stress on the bones. There was no history of autoimmune diseases in her family. On examination, the patient did not have signs suggestive of vascular or muscular etiology of leg pain. There was no bony tenderness. She underwent a radiograph of both legs which showed cortical thickening with bone marrow edema in bilateral tibial diaphyses with minimal adjacent soft tissue edema. Bone biopsy revealed predominantly dense lamellar bone with irregular sized and spaced haversian systems. Serum and urine markers of bone metabolism were within normal limits. The patient was treated with analgesics, and had partial relief from pain. Medullary rimming is the next treatment option in case pain progresses. This report emphasizes the role of bone scan in the diagnosis of this rare condition.

Keywords: Bone scan, dysplasia, leg pain, ribbing disease
resonance imaging (MRI) which revealed cortical thickening with bone marrow edema in bilateral tibial diaphysis along with minimal adjacent soft tissue edema [Figure 3]. The patient's hematological parameters were within normal limits. There was no leukocytosis. Serum and urine markers of bone metabolism were also nondiagnostic. The patient then underwent a tibial biopsy which revealed predominantly dense lamellar bone with irregular sized and spaced haversian systems [Figure 4]. The bony lamellae were thick, sclerotic with few osteocytes. Photomicrographs at higher magnification (40×) demonstrate dense lamellar bone with fragments of normal cortical bone. The haversian canals are variable in caliber, irregular spaced and are increased in number.

It is important to differentiate this entity from the more common causes of bony leg pain like stress fractures, shin splints, osteomyelitis, fibrous dysplasia, osteoid osteoma, osteosarcoma, and other rarer causes like adamantinoma, melorheostosis, hyperphosphatasia, histiocytosis, lymphoma, intramedullary sclerosis, endosteal hyperostosis and sclerosteosis.

Bone dysplasias like Engelmann disease are a close differential of Ribbing disease. The distinction between Ribbing disease and Camurati-Engelmann disease (progressive diaphyseal dysplasia) has been unclear. Engelmann disease has been more frequently reported in literature. It is a progressive disorder associated with pain, muscle weakness, fatigue, waddling gait, and anemia. Though Engelmann disease and Ribbing disease may appear to be identical radiographically, Seegei et al. pointed to certain clinical and histologic differences. Engelmann disease presents during childhood, while Ribbing disease usually...
Indian Journal of Nuclear Medicine | Vol. 26: Issue 1 | January-March, 2011

Reinus WR, Fischer KC, Ritter JH. Painful transient tibial edema. Radiology 1962;167:689-94.

Bettini G, Bonvi V. Etiopathogenetic, clinical, and radiographic considerations on Ribbing's disease: Report of 2 cases of familial nature. Ann Intern Med 2001;134:541-9.

Chanchairajira K, Chung CB, Lai YM, Haghhighi P, Resnick D. Intramedullary osteosclerosis: Imaging features in nine patients. Radiology 2001;220:225-30.

The treatment of ribbing disease is mainly symptomatic and is done with nonsteroidal anti-inflammatory drugs in increasing dose as required. However, bisphosphonates have been used in some cases of Ribbing and Camurati-Engelmann diseases, with discordant outcome; however, the low bone turnover that seems to characterize this disease and the prevalence of osteoblastic activity over the osteoclastic action are probably the reasons why osteoclast inhibitors may have limited success. Medullary rimming is a treatment option where medical management fails.

Our case illustrates that a three-phase 99mTc-MDP bone scan has a very important role to play in the diagnosis of Ribbing disease. Firstly, it can rule out shin splints and stress fracture as the etiology of pain. Increased uptake restricted to the tibiae/femora can rule out other bone dysplasias like Camurati-Engelmann disease, which have a more extensive involvement. Also, the nature and pattern of uptake can exclude primary neoplasms and metastases. A close differential on bone scan seems to be chronic osteomyelitis, especially since the onset of Ribbing disease may be unilateral and both diseases exhibit increased tracer activity on all three phases of the bone scan. Clinical history and examination may play a crucial role in such cases. Since the uptake of MDP is dependent on the osteoblastic response, it may be worthwhile trying to correlate the disease activity, patient symptomatology and tracer uptake.

We conclude that 99mTc-MDP bone scan is probably the most helpful investigation in cases of bony leg pain where dysplasias like Ribbing disease are suspected.

REFERENCES

1. Seeger LL, Hewel KC, Yao L, Gold RH, Mirra JM, Chandnani VP, et al. Ribbing disease (multiple diaphyseal sclerosis): Imaging and differential diagnosis. AJR Am J Roentgen 1996;167:689-94.
2. Shier CK, Krausicky GA, Ellis BI, Kottamasu SR. Ribbing's disease: Radiographic-scintigraphic correlation and comparative analysis with Engelmann's disease. J Nucl Med 1987;28:244-8.
3. Makita Y, Nishimura G, Isegawa S, Ishii T, Ito Y, Okuno A. Intrafamilial phenotypic variability in Engelmann disease (FD): Are ED and Ribbing disease the same entity? Am J Med Genet 2000;91:153-6.
4. Felson DT, Chaisson CE, Hill CL, Totterman SM, Gale ME, Skinner KM, et al. The association of bone marrow lesions with pain in knee osteoarthritis. Ann Intern Med 2001;134:541-9.
5. Reimann WR, Fischer KC, Ritter JH. Painful transient tibial edema. Radiology 1994;192:195-9.
How to cite this article: Damle NA, Patnecha M, Kumar P, Gadodia A, Subbarao K, Bai C. Ribbing disease: Uncommon cause of a common symptom. Indian J Nucl Med 2011;26:36-9.

Source of Support: Nil. Conflict of Interest: None declared.

8. Inaoka T, Shuke N, Sato J, Ishikawa Y, Takahashi K, Aburano T, et al. Scintigraphic evaluation of pamidronate and corticosteroid therapy in a patient with progressive diaphyseal dysplasia (Camurati-Engelmann disease). Clin Nucl Med 2001;26:680-2.

9. Mari C, Catafau A, Carrio I. Bone scintigraphy and metabolic disorders. Q J Nucl Med 1999;43:259-67.

10. Chapurlat RD, Meunier PJ. Fibrous dysplasia of bone. Baillieres Best Pract Res Clin Rheumatol 2000;14:385-98.

11. Ziran N, Hill S, Wright ME, Kovacs J, Robey PG, Wientroub S. Ribbing disease: Radiographic and biochemical characterization, lack of response to pamidronate. Skeletal Radiol 2002;31:714-9.