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Bone island and hand involvement – A short review

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ABSTRACT

Bone island or enostosis is an enigmatic lesion affecting the skeleton system. As these anomalies have limited clinical implications owing to an asymptomatic course with most cases diagnosed incidentally, the exact etiopathogenesis is not clearly understood. A lot has been studied regarding radiological features and morphology despite scarce clinical cases and relevant studies. More research is warranted to provide in-depth details of these lesions that has potential to enlighten us with newer aspects of bone metabolism and properties. These lesions might just not be an aberration and may offer important clues about bone behavior and morphogenesis. Bone islands, when in large number, are associated with equally fascinating disorder called osteopoikilosis. This review, however, does not include osteopoikilosis on purpose. Hand involvement, although rarer, has been described and a short review is attempted to acknowledge that apart from relevant facts on diagnosis and management.

Key words: *Enostosis, bone island, diagnosis, management, imaging, hand*

Introduction

Bone island or enostosis is a rare benign lesion that often is an incidental radiological finding of dense sclerotic focus inside of a bone. It can mimic neoplastic pathology and thus requires knowledge about the entity to prevent unnecessary investigations. Bone islands represent a locus of compact, cortical bone inside a cancellous bone and considered developmental aberration that is mostly asymptomatic. Older literature describes these as hemartomas, but the probable etiology of these lesions is congenital or developmental one with defective enchondral ossification as reason [1]. Knowledge of this lesion and its features can help anticipate its presence and may reduce burden of unwarranted investigations in selected instances.

Bone Island Within Hands

The hand is an uncommon site for these lesions as most have predilection for long bones, ribs, spine, and pelvis. The reported incidence in the growing hand has been reported in a study to be 3.8% between ages of 5 and 13 [2]. These lesions are usually asymptomatic and scaphoid was found to be the most common location. Like the lesions in various other studies, certain of them also show positive scintigraphy later confirmed by biopsy to be a bone island as described in one report of the lesion in os capitate [3].

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A recent study showed a significant link of bone islands with leprosy more so with lepromatous type and borderline cases [4]. Hand and foot radiographs of patients with Hansen disease were compared to age and sex matched normal subjects in the abovementioned study. Significant presence of bone islands were found with Hansen disease of long duration. With no clear reason, the authors attributed this to genetic predisposition or side effect of disease itself.

Etiology and Involvement

These lesions are considered stable, benign and non-progressive but some of these may be symptomatic especially if more than 2 cm in size when they are referred to as giant bone islands. But small islands, though unusual, may also present with pain [5]. These lesions are common in pelvis, femur, ribs, facial and other long bones and usually are smaller than 2 cm.

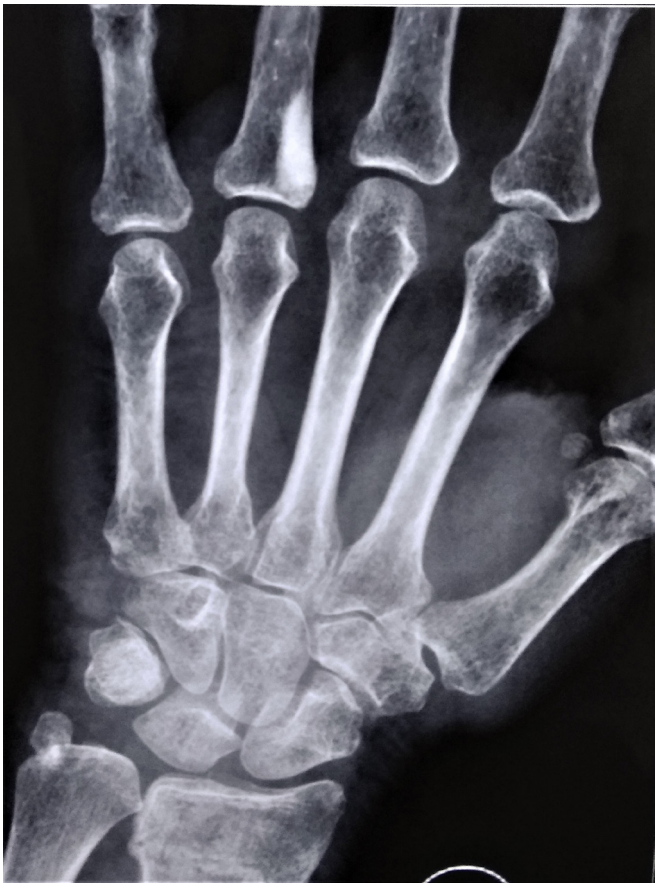

Figure 1. Radiograph showing a dense sclerotic lesion on fourth proximal phalanx.

but larger, symptomatic lesions or those involving rare sites are sporadically reported [6]. Carpal bones, tarsal bones and spine are also involved by the lesion. In long bones these are found to be involving epiphysis and metaphysis. One report of enostosis with diaphyseal exostosis is reported in a family [7].

In one report, the responsiveness of bone islands to factors affecting bone metabolism like hormonal influence was noted. The bone island was reported to disappear in case of hyperparathyroidism and reappeared on surgical removal of parathyroid adenoma [8]. This finding highlights importance of further research in this aspect of the disorder. Five lesions in a series were described to enlarge proportionate to growth in adolescent cases thus suggesting their metabolic participation [9]. Though not commonly found in immature skeleton, growth of more than 25% over six months should be cautiously assessed with biopsy to rule out neoplastic element [10]. Multiple bone islands have been associated with various other disorders like osteopoikilosis (osteopathia disseminate), striped osteopathy (Voorhoeve disease), and malrheostosis [11].

Investigations

The lesions present as homogenously dense lesion in a cancellous bone with radiating bony streaks blending with native trabecular bone (Figure 1 and 2). The lesion may be round, oval or oblong in shape. These peripheral steaks are also referred to as 'thorny radiations', feather like spicules or brush borders and are better appreciated in CT than radiographs. Magnetic resonance imaging (MRI) shows a dark black spot corresponding to the lesion without any surrounding edema or related pathology (Figure 3). There is no periosteal reaction or destruction is seen. Low signal intensity in all views resembling cortical bone is thus found in MRI.

CT scan usually show a low attenuation focus in this lesion showing inert nature. These enigmatic lesions, however, carry a scope for enlargement in size or spontaneous shrinkage overtime [12].These homogenously dense sclerotic foci appear with low attenuation

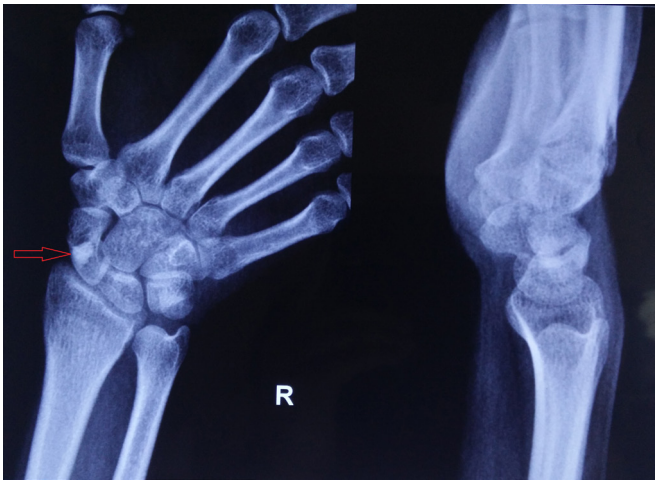

Figure 2. Radiograph of wrist showing dense, sclerotic focus in scaphoid region (arrow) with otherwise normal scaphoid and rest carpal bones.

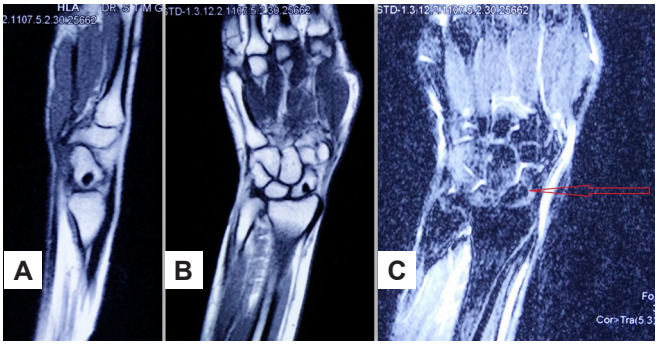

Figure 3. MRI sections showing low attenuated lesion in sagittal **(A)** and coronal **(B)** plane showing no bone edema or associated pathology. The lesion appears similar in T-2 weighted images also **(C)**.

signal both in CT and MRI to assist in diagnosis. Meticulous and specific CT attenuation is required to better diagnose or differentiate these lesions from osteoblastic metastasis as described in a study comprising large series [13]. The study highlights importance of specific attenuation thresholds in order to differentiate osteosclerotic islands from untreated osteoblastic metastatic lesions. The osteosclerotic metastatic lesions also appear hot on bone scan but in MR imaging on fluid sensitive sequences may reveal a halo of bone marrow edema surrounding the lesion [14].

Bone scintigraphy coupled with histopathology may be tried in cases with dubious details [15]. These lesions mostly appear as cold spot in scintigraphy, the feature that may help distinguish it from neoplastic differentials. However a small percentage of lesions may have aggressive look in scintigraphy and histopathological assessment is thus confirmatory [16]. Some of these lesions with increased osteoblastic activity present as active lesion on scintigraphy [17]. Thus, the morphological assessment on radiographs, CT and MRI is better to rely upon than scintigraphy for the diagnosis. The large lesions may be followed up with periodic imaging to check increase in growth in future [1,18,19]. Lesions with increased uptake and polyostotic pattern have also been reported [20]. In patients suffering from lung cancer, spectral CTs has been found to be better at differentiating osteoblastic metastasis from bone islands in a large study of 122 lesions (43 bone islands and 79 metastatic lesions) but these cases represented lesions in vertebral bodies [21].

Microscopic examination reveal compact lamellar bone with normal Haversian systems surrounded by spongiosa with no mitoses or atypical nuclei. Radiating trabeculae of the lesion blending with native host cancellous trabeculae can also be appreciated.

Management

As most of these are incidental findings owing to their asymptomatic status, small islands are managed conservatively by observation. Some of the small bone islands, however, may be symptomatic and may require surgical treatment [19,22]. One such reported case of hip pain, limp and subluxation due to small island adjacent to growth plate of proximal femur has been described with transmetaphyseal curettage as effective management [22]. This method can be successfully used in symptomatic hand lesions if the articular surfaces are not involved. Small carpal bones might require cautious curettage as the small cross sectional area may predispose to iatrogenic fractures complicating the procedure.

Giant lesions may warrant future evaluation if there is increase in size. Symptomatic lesions or those with increased uptake on scintigraphy require biopsy for ruling out sinister underlying disorder. Osteosarcoma like low grade central type is prominent differential diagno-

sis requiring exclusion [23]. Other differentials may be osteoid osteoma, enchondroma, osteoblastoma, bone infarcts, healing non-ossifying fibroma among others. Besides it, the supernumary islands may be part of well described disorders and require identification like osteopoikilosis, osteopathia striata and melorheostosis. There are numerus described sclerosing bone dysplasia and at certain times one or more of the type concomitantly involve i.e. overlap syndrome. All these are supposed to be defects of bone formation or resorption and may have interlacing pathogenesis that future research might unearth [24].

The literature is sparse regarding overall prevalence and surgical management of these lesions and no definitive guidelines are established. The unanimous point highlighted in nearly all studies is to observe the quiescent lesions and investigate the symptomatic ones to determine those lesions as source of symptoms. Meticulous assessment of clinical diagnosis should be coupled with judicious imaging studies and surgical management should be reserved for recalcitrant cases.

Conflict of interest statement

The authors have no conflicts of interest to declare.

References

1. Greenspan A. Bone island (enostosis): current concept-a review. Skeletal Radiol 1995;24:111-5.
2. Limb D, Agrawal Y. The distribution of bone islands and juxta-articular bone cysts in the growing hand. J Hand Surg (British and European Volume) 2006;31:441-4.
3. Araki S, Otani T, Watanabe K, Yamaji T. Positive bone scan in a bone island: A histologically verified case in os capitatum. Acta Orthop Scand 1989;60:369-70.
4. Carpintero P, García-Frasquet A, Tarradas E, Logroño C, Carrascal A, Carreto A. Bone island and leprosy. Skeletal Radiol 1998;27:330-3.
5. Gerges M, Glard Y, Launay F, Jouve JL, Bollini G, Cottalorda J. Small bone islands: unusual clinical symptomatology. Orthopedics 2014;37:e79-82.
6. Park HS, Kim JR, Lee SY, Jang KY. Symptomatic giant (10-cm) bone island of the tibia. Skeletal Radiol 2005;34:347-50.
7. Kaganova ED, Efimov FF. Familial disease of diaphyseal exostosis and enostosis. Ortop Travmatol Protez 1958;19:75-7.
8. Hoffman Jr RR, Campbell RE. Roentgenologic Bone-Island Instability in Hyperparathyroidism: Case Report. Radiology 1972;103:307-8.
9. Onitsuka H. Roentgenologic Aspects of Bone Islands. Radiology 1977;123:607-12.
10. Motamedi K, Seeger LL. Benign bone tumors. Radiol Clin North Am 2011;49:1115-34.
11. Cerase A, Priolo F. Skeletal benign bone forming lesions. Eur J Radiol 1998;27:91–7.
12. Petrikowski CG, Peters E. Longitudinal radiographic assessment of dense bone islands of the jaws. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 1997;83:627-34.
13. Ulano A, Bredella MA, Burke P, Chebib I, Simeone FJ, Huang AJ, et al. Distinguishing Untreated Osteoblastic Metastases From Enostoses Using CT Attenuation Measurements. Am J Roentgenol 2016;207:362-8.
14. Schweitzer ME, Levine C, Mitchell DG, Gannon FH, Gomella LG. Bull's eyes and halos: useful MR discriminators of osseous metastases. Radiology 1993;188:249–52.
15. Greenspan A, Stadalnik RC. Bone island: scintigraphic findings and their clinical application. Can Assoc Radiol J 1995;46:368-79.
16. Araki S, Otani T, Watanabe K, Yamaji T. Positive bone scan in a bone island: A histologically verified case in os capitatum. Acta Orthop Scand 1989;60:369-70.
17. Davies JAK, Hall FM, Goldberg RP, Kasdon EG. Positive bone scan in bone island. J Bone Joint Surg Am 1979;61:943-5.
18. Greenspan A, Stadalnik RC. Bone island: scintigraphic findings and their clinical application. Can

Assoc Radiol J 1995;46:368-79.
19. Greenspan A, Steiner G, Knutzon R. Bone island (enostosis): clinical significance and radiologic and pathologic correlations. Skeletal Radiol 1991;20:85-90.
20. Caballes RL, Caballes RA. Polyostotic giant enostoses with strongly positive radionuclide bone scan. Ann Diagn Pathol 2004;8:247-51.
21. Dong Y, Zheng S, Machida H, Wang B, Liu A, Liu Y, et al. Differential diagnosis of osteoblastic metastases from bone islands in patients with lung cancer by single-source dual-energy CT: advantages of spectral CT imaging. Eur J Radiol 2015;84:901-7.
22. Diab MG, Glard Y, Launay F, Jouve JL, Bollini G, Cottalorda J. Small Bone Islands: Unusual Clinical Symptomatology. Orthopedics 2014;37:e79-82.
23. Brien EW, Mirra JM, Latanza L, Fedenko A, Luck J Jr. Giant bone island of femur: case report, literature review, and its distinction from low grade osteosarcoma. Skeletal Radiol 1995;24:546–50.
24. Greenspan A. Sclerosing bone dysplasias-a target-site approach. Skeletal Radiol 1991;20:561-83.