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Case Report

Osteopoikilosis – Spotted bone disease

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Introduction

Osteopoikilosis is an asymptomatic, autosomal dominant, sclerosing dysplasia characterized by multiple, discrete round or ovoid radio densities in cancellous bones. Initially described by Albers-Schonberg in 1915 it is occasionally associated with cutaneous lesions as well as other osteo-sclerotic skeletal disorders.

Case report

A 40-year old male patient with history of trauma to the right elbow was referred to the radiology department for radiographic examination. Clinical examination was unremarkable. The patient did not have any other co-morbidity. The radiograph of the right elbow revealed multiple, well defined, ovoid to lanceolate foci of increased density clustered in the

epiphyseal and metaphyseal regions (Fig. 1). There was no evidence of any osseous injury.

On questioning the patient, no past or family history of similar osseous lesions could be elicited for any radiographic examination of either the individual or his family members. His laboratory and biochemical parameters inclusive of serum alkaline phosphatase, ESR and complete blood count were within normal limits.

Similar lesions involving bilateral wrists (Fig. 2), hips and shoulders (Fig. 3) were noted on detailed skeletal survey of the patient. In addition to the long bones, involvement of the carpus was noted bilaterally. There was no evidence of cortical thickening or periosteal reaction seen in association with these lesions. Visualised soft tissues were normal. No spinal involvement was observed.

For further characterization of these lesions unenhanced CT of the pelvis with suitable gonadal shielding was performed. It revealed multiple, symmetrical, well defined round to oval sclerotic foci involving epiphyseal and metaphyseal regions of femora, acetabulum, pubic rami, ischium as well as juxta articular location of sacro-iliac joints (Fig. 4). These findings were characteristic of Osteopoikilosis.

Discussion

Osteopoikilosis (Osteopathia condensans disseminata; Spotted bone disease) is an asymptomatic sclerotic dysplasia initially described by Albers-Schonberg and Ledoux-Lebard and associates in the early twentieth century. This disorder is seen in both men and women and may become evident at any age, although it is rare to find this disease below age of 3 years.

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Fig. 1 – AP and lateral radiographs of the right elbow: small, discrete, sclerotic lesions involving the epiphyseal and metaphyseal regions of humerus, radius and ulna are seen.

Both inherited and sporadic forms have been reported. An autosomal dominant pattern that becomes more and more prominent in each succeeding generation has also been described.^{1,2}

Clinical manifestations, if present, are usually mild. Dermatological abnormalities in form of closely situated, elevated, whitish fibrocollagenous infiltrations (dermatofibrosis lenticularis disseminata or the Buschke–Ollendorff syndrome),

a predisposition to keloid formation and scleroderma like lesion are seen in approximately twenty five percent of the cases. Associations with dwarfism, dystosia, premyelopathic syndrome due to spinal stenosis, organ anomalies (coarctation of aorta, double ureter) and in 15–20% of patients mild articular pain with or without joint effusion have also been reported.^{2,3}

Radiologically the abnormality appears as small, well defined, ovoid or lanceolate sclerotic lesions arranged

Fig. 2 – Radiograph both wrists PA: multiple, small sclerotic lesions in periarticular distribution involving the distal radius and ulna, carpals and the metacarpals noted.

Fig. 3 – Frontal radiograph of the right shoulder revealing multiple periarticular, small sclerotic lesions involving the humerus and scapula.

scintigraphy. The sclerotic lesions in mastocytosis and tuberous sclerosis on the other hand are less well defined, asymmetric and show lesser periarticular preference.⁶ These findings along with exclusion of the primary malignancy by imaging of the prostate, serum PSA levels, endoscopy, colonoscopy, breast imaging and other ancillary tests may help to arrive at the diagnosis of Osteopoikilosis.

However, Mungovan et al suggested that in a setting of characteristic radiological findings of Osteopoikilosis an abnormal bone scan does not exclude its diagnosis.⁷

To conclude multiple, symmetrical, well defined, periarticular sclerotic lesions merit consideration of Osteopoikilosis along with other differential diagnosis before costlier investigations are undertaken.

Conflicts of interest

All authors have none to declare.

Fig. 4 – Unenhanced CT of the pelvis shows multiple, small, discrete, sclerotic lesions in symmetric periarticular distribution of hip and SI joints.

symmetrically around the joints, more commonly the epiphysis and metaphysis of long tubular bones.^{1–4} Histopathologically these lesions correspond to lamellar osseous tissue containing Haversian systems.^{3,4}

In an epidemiological study of familial Osteopoikilosis by Benli et al a predominance of these lesions was found in the phalanges of hand (100%) followed by carpal bones (97.4%), metacarpals (92.3%), foot phalanges (87.2%), metatarsals (84.4%), tarsal bones (84.6%), pelvis (74.4%), radius (66.7%), ulna (66.7%), sacrum (58.9%), humerus (28.2%), tibia (20.5%) and fibula (12.8%).⁵

On serial radiographs these sclerotic may show an increase or decrease in size or number with occasional disappearance in children, in sharp contrast with adults in whom these lesions remain more or less static.¹

Differential considerations for this abnormality include osteoblastic metastases, tuberous sclerosis and mastocytosis. The blastic metastases primarily involve the axial skeleton, demonstrate osseous destruction and show uptake on bone

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