Case Report

Osteopoikilosis; A Rare Incidental Spotted Bone Disease on CT

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Abstract: Osteopoikilosis is a benign, asymptomatic, sclerosing bone dysplasia with an autosomal dominant trait. The disease is characterized by diffuse symmetrical small round and ovoid radiopacities in the juxta-articular region of cancellous bone. We reported a rare case of a middle aged female presented with abdominal distension, whose radiological work up revealed an incidental osteopoikilosis.

Keywords: Osteopoikilosis, CT scan, spotted bone disease, benign, asymptomatic.

Introduction

Osteopoikilosis (OP) is a benign sclerosing bone dysplasia. It is an asymptomatic, autosomal dominant trait which may mimic many different bone pathologies.[1] It is also known as osteopathic condensans disseminata or spotted bone disease. The disease is associated with dermatofibrosis lenticularis disseminata (Buschke-Ollendorf Syndrome).[2] We report a rare case of osteopoikilosis in a middle aged female presented with abdominal distension, asymptomatic for any musculoskeletal pathology. Incidental diagnosis of osteopoikilosis was established during radiological workup. So far, we didn't find any published case report in Pakistani literature related to osteopoikilosis.

Case Report

A 42 years old female presented at outpatient department with a complaint of dull abdominal pain and distension for the past 3 months. She also had decreased apatite over the period of illness; however, there was no significant weight loss. No bowel related abnormality was narrated by the patient. Systemic review was unremarkable including musculoskeletal system. Clinical examination revealed abdominal ascites besides a non-tender abdomen, later confirmed on a transabominal ultrasound scan. Patient was sent to our department for cross-sectional imaging of abdomen with clinical diagnosis of intestinal tuberculosis. CT scan abdomen revealed gross ascites, however, the cause was not ascertained. A note was made of bilateral moderately dilated pelvicalyceal system, probably due to pelvi-ureteric junction (PUJ) obstruction. On skeletal review, multiple well defined subcentimeter (approximately 2-5 mm in diameter each) circular to ovoid hyperdense spots were visualized in the skeleton, more marked in vertebrae, pelvic bones and epiphyses of the femur. These hyperdensities were located in cancellous part of bone.

As the radiological findings were unexpected, an X-ray of the pelvis and shoulder were also acquired which confirmed the findings. All bones were free of any cortical erosion or periosteal reaction. The diagnosis of osteopoikilosis was established.

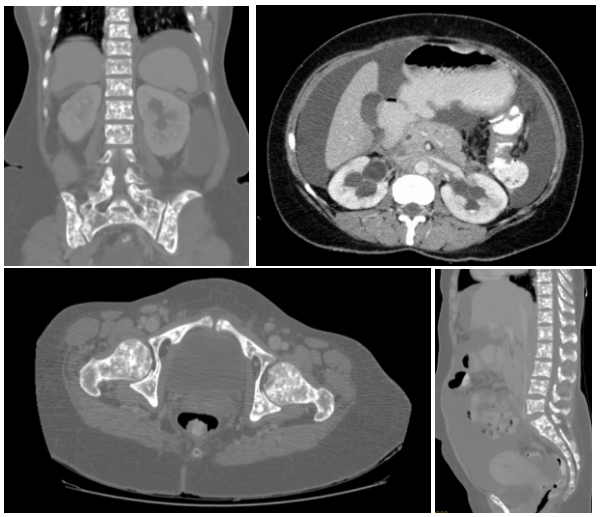

Fig-1: Gross ascites with bilateral moderately dilated pelvicalyceal system (PUJ obstruction). Multiple well defined 2-5 mm circular to ovoid hyperdense spots in the cancellous part of vertebrae, pelvic bones and distal end of the femur.

Being an incidental finding, patient was counseled that the condition was benign and static, that is not going to harm her. The diagnostic tap analysis of ascitic fluid turned out to be reactive in nature, and the patient was treated accordingly by the attending physicians.

Discussion

Osteopoikilosis is a rare bone dysplasia found in less than 0.1 per million. Radiologically, it is characterized by diffuse symmetrical bone islands, small round and ovoid radiopacities in the Juxtaarticular regions of

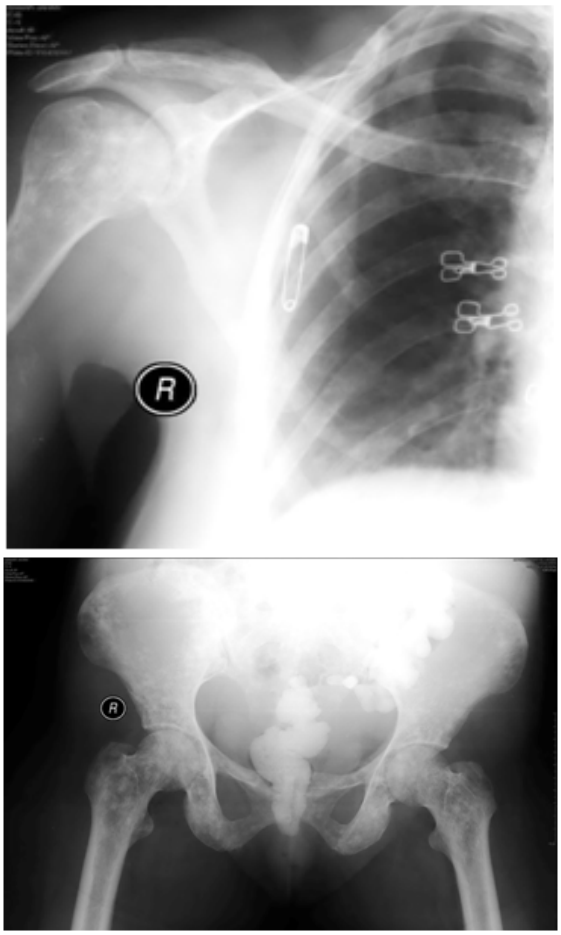

Fig-2: A/P projection of right shoulder shows multiple variable sized circular to ovoid radio-opacities in the cancellous part of proximjal humerus. A/P projection of pelvis revealing subcentimeter radio-opacities in the cancellous part of pelvic bones and proximal femur. Note the intruluminal contrast agent in the rectum due to oral preparation for CT abdomen.

bone are characteristic radiologic signs in cancellous bone which may appear at birth or during skeletal growth. It is usually found in the metaphyseal and epiphyseal regions of long bones, the carpals and tarsals, the end of large turbular bones and around the acetabula. It is occasionally associated with hereditary multiple exostosis, scleroderma and dermatofibrosis lenticularis disseminate.[3] Osteopoikilosis must be considered as a distinct clinical entity. Several diseases should be taken under consideration for its differential diagnosis, such as osteoblastic metastasis, tuberous sclerosis, Paget`s disease, mastocytosis, osteopathia striata, melorheostosis, synovial chondromatosis, sesamoid ossicles and Ollier`s disease.[4] As a general rule for metastasis, the radiographic pattern is lytic type and may cause subcortical destruction. The lesions in OP are symmetric, smaller, uniform size and do not cause cortical erosions.[5, 6] Due to the benign nature of OP, complications are very rare. Possible complications described in the literature are osteosarcoma, giant cell tumor and chondrosarcoma. Diagnosis is usually established with simple X-ray imaging. MRI may aid in differential diagnosis in more complicated cases, showing multiple circular or ovoid hypointense lesions located in proximal or distal epiphyses. In addition, radionuclide bone scan (scintigraphy) can also help distinguishing OP from osteoblastic bone metastases, but abnormal bone scan does not exclude OP. Bone scintigraphy demonstrates usually absence of radiotracer uptake in OP patients.[7] Since the condition is inherited, a family study should be done. Being asymptomatic and treatment is unnecessary.[8]

Conclusion

Since the benign bone lesions of osteopoikilosis may mimic the bone metastases and could easily be mistaken for metastatic disease, it is important that physicians be aware of the benign nature of this condition.

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