

CASE REPORT

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Osteopoikilosis: Pain as a Presenting Symptom in Three Family Members

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Abstract: Osteopoikilosis is a rare asymptomatic sclerosing bony dysplasia of benign origin. It is usually found incidentally on radiological examinations. Familial occurrence indicates a genetic milieu with autosomal dominant pattern. Here, we present a case report of a young woman suffering from pelvic pain due to osteopoikilosis (OPK). The same disorder was later found in her son and daughter.

Keywords: Osteopoikilosis, pain, familial occurrence

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Introduction

Osteopoikilosis, spotted bone disease or osteopathia condensans is a rare asymptomatic bone dysplasia of unknown etiology.¹ It is characterized by an abnormality in bone maturation process and often found incidentally on radiologic examination. An autosomal dominant inheritance has been proposed for OPK. Albers-Schonberg was the first to describe this uncommon sclerosing bone dysplasia in 1915.² Incidence in both sexes is identical and it can occur at any age. Overall incidence of OPK has been claimed to be one in every 50,000 subjects.³ Radiologic signs of OPK are homogeneous, small, well-defined symmetric round and ovoid radio opacities lesions. The commonly involved sites are metaphyses and epiphyses of long bones, scapulae, pelvis, carpi and tarsi.⁴

Pain is not a prominent feature of OPK, but in some patients, pain could be a presenting symptom of the disorder.

Case Report

A 46 year old female patient was referred to our pain clinic, complaining of pelvic pain starting 8 years ago. She reported the presence of diffuse pelvic pain mostly located at both hip joints. Her pain was constant, but worsened when climbing stairs and walking for long distances. The quality of pain was vague and its severity was rated as 5–6 on the visual analogue scale. She did not report any history of past or recent trauma or systemic disease and was not taking any medications. There was no visible gait abnormality or postural deviations. Inspection of pelvis showed bilateral symmetry of muscle bulk and there were no scars or bruising. Palpatory examination revealed slight tenderness over the great trochanters and symphysis pubis, however no trigger points were located.

On physical exam, range of motion of the hip joints was normal with no restriction. Neurologic examination revealed no abnormality and provocative tests such as Thomas Test, Patrick's Test, femoral stretch test and straight leg raising were normal. Sacroiliac joint tests were also normal. No sign of cutaneous connective tissue involvement was noted. On laboratory, all findings for complete blood count, erythrocyte sedimentation rate, C—reactive protein, rheumatoid factor, serum electrolytes, alkaline phosphatase, calcium, phosphate and thyroid function tests were normal.

Pelvic X-ray showed multiple oval shaped radiodensities of various degrees on the upper heads of femur, acetabulum and symphysis pubis (Figs. 1, 2): osteopoikilotic lesions has also found in the foot X ray of the patient. In order to rule out metastases, whole body bone scintigraphy was performed and the result was normal. Abdomino pelvic ultrasonography was also normal.

In light of these characteristic lesions and findings without cancer red flags, a diagnosis of Osteopoikilosis was made. Non-steroidal anti-inflammatory drug, Celecoxib, was prescribed. She was found to be pain free and asymptomatic in a follow up visit after the first month. Two months later, she returned to our clinic along with her 11-year-old son and sixteen year old daughter. Both complained of occasional pain in their legs. Plain radiographs revealed the same characteristic ovoid sclerotic lesions of OPK in distal end of Tibia, Fibula (fig 3, 4) and foot. Further work up showed no other abnormalities except for an exostosis in medial aspect of tibia in her son's X ray.

Discussion

This current report presents a 46 year old female patient and her son and daughter diagnosed with OPK. An epidemiologic study on 53 patients affected with OPK indicated that the frequent sites for OPK, were phalanges (100%), carpal bones (97.4%), metacarpals (92.5%), foot phalanges (87.2%), metatarsals (84.4%), tarsal bones (84.6%), pelvis (74.4%), femur (74.4%), radius (66.7%), ulna (66.7%), sacrum (58.9%), humerus (28.2%), tibia (20.5%), and fibula



Figure 1. X ray of the patient pelvis, showing multiple symmetric and sclerotic lesions.



Figure 2. Foot X ray of the 16 year old daughter, showing osteopoikilotic lesions.

(2.8%).⁵ In our cases, we found multiple oval shaped radiodensities of various degrees on the upper heads of femur, acetabulum and symphysis pubis.

Characteristic radiologic signs of OPK consist of numerous 2–10 mm ovals or round shaped densities,



Figure 3. Ankle X ray of 11 year old boy, showing ovoid sclerotic lesions in distal end of Tibia and Fibula.



Figure 4. Knee joint X ray of 11 year old son, showing ovoid sclerotic lesions in distal end of Tibia and Fibula. Circle indicates an exostosis in medial aspect of Tibia.

symmetrically distributed within epiphyses and metaphyses of long bones. These lesions appear as dense radiopaque spots.⁶ In the current study, pelvic X-ray showed multiple oval shaped radiodensities of various degrees on the upper heads of femur, acetabulum and symphysis pubis.

Pain is not a dominant feature of OPK but in 15%–20% of patients, slight joint pain and effusion have been reported.⁴ The main complaint in our patients, which caused them to seek medical help, was persistent pain in affected areas. Patient examination was not remarkable and no deformity or dysfunction could be documented. Some hypotheses have been made to explain the mechanisms of pain in OPK. It is presumed that joint pain is an incidental finding in the course of OPK. Increased localized bone metabolism at the location of the lesion, irritation of joint capsule attachment by sclerotic areas and increased intraosseous pressure due to venous stasis at the areas of lesion could produce joint pain.⁷ In nearly 25% of patients, OPK is associated with Buschke—Ollendorf



Syndrom.⁸ However, we could not find any dermatologic conditions as those in Buschke—Ollendorf Syndrome in our patient or her family members.

It is clinically significant to differentiate OPK from metastatic osteoblastic lesions. Therefore, the major differential diagnosis should be considered for osteoblastic metastases, mastocytosis and tuberous sclerosis.⁹ Asymmetry, variation in size, axial skeleton involvement, osseous destruction and periosteal reaction differentiates osteoblastic metastases from OPK. In addition, scintigraphy can help distinguish OPK from osteoblastic bone metastases but abnormal bone scan does not exclude OPK.¹⁰

In our case, there were no hot spots indicating increased uptake of radiotracer in bone scan. Because of the familial nature of the disorder, when the patient came back with two offspring (her son and daughter), they were also evaluated and were found to have the same characteristic lesions as their mother. We did not ask for bone scintigraphy in her son and daughter.

Although the risk of malignant transformation with OPK is very rare, some complications such as osteosarcoma,¹¹ giant cell tumors¹² and chondrosarcoma¹³ have been reported in the literature. Therefore, we regularly followed up with the patient and other affected members of her family.

In conclusion, OPK is an asymptomatic natural variation with benign nature but in patients with characteristic radiologic findings, the patients may be presented with pain. Familial occurrence of OPK in relatives of the affected person should also be considered. Thus, well-timed follow-up visits of the patients are recommended to survey other conditions which may require treatment.

Disclosure

This manuscript has been read and approved by all authors. This paper is unique and is not under consideration by any other publication and has not been published elsewhere. The authors and peer reviewers of this paper report no conflicts of interest. The authors confirm that they have permission to reproduce any copyrighted material. According to institutional ethical rules, an informed consent was obtained from the patient for herself and her under 18 year old offspring.

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