A Rare Cause of Polyarthralgia: Osteopoikilosis

Poliartraljinin Nadir Bir Sebebi: Osteopoikilozis

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Abstract

Osteopoikilosis or spotted bone disease is a rare bone dysplasia of unknown etiology. It is characterized by an abnormality in bone maturation process and often found incidentally on radiologic examination. This disease, which has an autosomal dominant inheritance, has numerous, annular or oval, sclerotic lesions with regular borders. Although it is frequently asymptomatic, rarely polyarthralgia and joint swelling may occur. In this case, we aimed to show that the etiology may be a rare syndrome such as osteopoikilosis in the patient with the complaint of polyarthralgia.

Keywords Arthralgia; Osteopoikilosis; Osteosclerosis

Öz

Osteopoikilozis veya benekli kemik hastalığı, etiyolojisi bilinmeyen nadir bir kemik displazisidir. Kemik olgunlaşma sürecinde anormallik ile karakterizedir ve sıklıkla radyolojik incelemede tesadüfen bulunur. Otozomal dominant kalıtıma sahip olan bu hastalık, düzenli sınırları olan çok sayıda, dairesel veya oval, sklerotik lezyonlara sahiptir. Sıklıkla asemptomatik olmasına rağmen nadiren poliartralji ve eklemlerde şişlik görülebilir. Bu olguda poliartralji yakınması olan hastada etiyolojinin osteopoikilozis gibi nadir bir sendrom olabileceğini göstermeyi amaçladık.

Anahtar Kelimeler *Artralji; Osteopikilozis; Osteosklerozis*

INTRODUCTION

Osteopoikilosis (OPK) is a rare, autosomal dominant, benign and osteosclerotic bone disease. It can be seen in any age group and there is no gender discrimination. Patients are generally asymptomatic and detected by incidental radiographs. Although many patients are asymptomatic; some patients may have joint pain. Symmetrical, numerous, small, well-defined, homogeneous, circular or oval sclerotic lesions clustered in the epiphysis and metaphysis of long bones are seen in the periarticular areas on the graph.[1] Osteopoikilosis can be confused with osteoblastic metastases, tuberous sclerosis and bone mastocytosis because of the similarity of radiological images.[2] If the patient has pain Non Steroidal Anti Inflammatory Drugs (NSAID) are often used as an option, analgesics such as acetaminophen and opioids can also be used. Rare active lesions have been treated with bisphosphonate therapy; but the results are controversial.[3]

Osteopoikilosis is histologically defined as bone islands that form dense core structure and are not associated with bone marrow in trabecular or cancellous bone. Some hypotheses have been made to explain the mechanisms of pain in OPK. Increased 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.[4] In this case; a case of OPK, which may rarely occur with polyarthralgia, will be presented.

CASE REPORT

50-year-old woman with polyarthralgia, especially in the ankles, for about 2 years; she applied to our physical medicine and rehabilitation clinic of xxxxxx University Training and Research Hospital. The patient had diabetes mellitus disease and was using 1000 mg of metformin daily. The patient, who had a cesarean operation once before, had no other operation or history of trauma. The patient's symptom at presentation was polyarthralgia. There was no redness or warmth in the any joint. The patient had no weight loss, fever, and night sweats. On physical examination, the range of motion in all joints was complete; and her muscle strength examination was normal. There was minimal swelling and tenderness lateral side of the right ankle, there was no warmth and color change. There was tenderness in both hands 2., 3., 4. and the 5. metacarpophalangeal joint. The Neer, Hawkins and Speed's test of the patient, which said that the pain in her right shoulder increased for the past month, were negative. Her systemic examination was normal. There was no skin sign called dermatofibrosis lenticularis disseminata or Buschke Ollendorff syndrome.

The patient, who has no symptom in her rheumatological questioning, has been to the physical therapy clinic several times before, she has not received any diagnosis, and she has been using non-steroidal anti-inflammatory drugs and vitamin D due to her polyarthralgia; she also stated that she benefited from them partially. Routine medical workups and bilateral ankle radiographs of the patient were requested. Rheumatoid Factor, Anti Nuclear Antigen, Cyclic Citrulline Peptide values, which were previously investigated for rheumatological disease suspicion, were normal. The patient's vitamin D level, electrolyte values, thyroid function tests, whole blood test, erythrocyte sedimentation rate and C-reactive protein level were normal. She had no complaints of inflammatory pain. She had no chronic widespread pain at multiple tender points, joint stiffness, and systemic symptoms (e.g., mood disorders, fatigue, cognitive dysfunction, and insomnia). The patient who has no feature in routine medical workups; but multiple, well-circumscribed, round, sclerotic lesions were determined on the ankle radiographs. Afterwards, x-ray radiographs were requested to the other joints of the patient and existing lesions were observed in almost all joints (Figure 1, 2, 3).

Scintigraphy was requested for osteoblastic bone metastasis exclusion due to diffuse lesions and polyarthralgia. The scintigraphy result was reported as normal. The patient, who was also consulted to the Orthopedics and Trauma-

tology clinic, was diagnosed with OPK. It was planned to follow up with NSAID and to continue their regular follow-up without any aggressive or invasive intervention. Informed consent was obtained from the patient and permission was obtained from the patient for the presentation of the case.

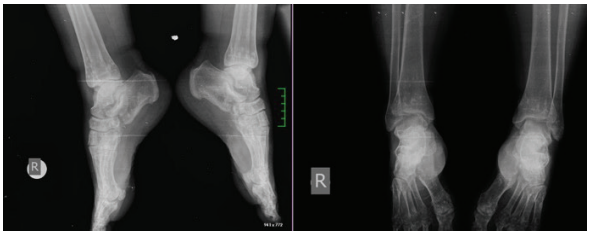

Figure 1: Anteroposterior and lateral radiograph of ankles. Numerous sclerotic lesions in both ankles and metatarsophalangeal joints

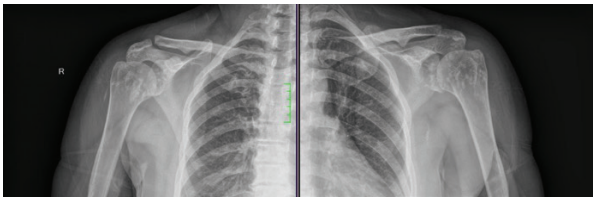

Figure 2: Anteroposterior radiograph of the shoulders. The humeral head and widely distributed, small, well-circumscribed and circular sclerotic bone lesions around the glenoidal cavity.

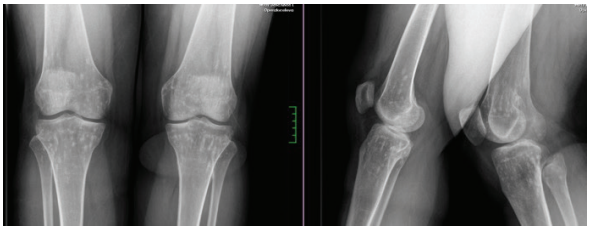

Figure 3: Anteroposterior and lateral radiograph of knees. There are multiple well-defined sclerotic lesions around both knee joints

DISCUSSION

Osteopoikilosis is an asymptomatic osteosclerotic bone disease in which sporadic and familial cases have been reported in the literature, whose pathogenesis has not been fully elucidated. The cases are mostly diagnosed incidentally on typical radiographic findings on direct radiographs.

However, it rarely causes polyarthralgia and joint swelling. Pain is not a dominant feature of OPK but in 15%–20% of patients, slight joint pain and effusion have been reported.[5] Patients with OPK should be monitored since—although the clinical course is benign—occasional complications or coexisting pathological conditions have been reported including premyelopathic syndrome due to spinal canal stenosis, hip fracture, malignant degeneration, and other disorders requiring medical surveillance.[6] It is common to consider diseases that affect the musculoskeletal system such as rheumatoid arthritis, vitamin deficiencies, fibromyalgia, etc. in pre-diagnoses in patients admitted to physical therapy clinics due to polyarthralgia. However, while these frequent diseases are considered at the forefront, rare diseases should also be taken into consideration and current diagnosis methods should be used effectively, especially imaging.

Bone scintigraphy is largely normal in OPK and this feature is important in distinction it from osteoblastic bone tumors. In addition; asymmetry, variation in size, axial skeleton involvement, osseous destruction and periostal reaction differentiates osteoblastic metastases from OPK.[7] In the study of Carpintero et al. which included ten OPK patients, four patients complained of abdominal and/or urological symptoms and had undergone abdominal radiography, three had injuries, two complained of joint pain (accompanied in one case by effusion in the knee), and the tenth patient was referred because her brother had been diagnosed elsewhere as having OPK. In same study three patients had associated bone disorders (two displayed osteopathia striata and the other exostosis of the anterior aspect of the ankle, together with bone condensation consistent with melorheostosis).[8] In our case, there was no family history and no associated bone disorders was detected. Serdaroglu et al. showed in their case reports that lesions

may be metabolically active and their size may change and become denser over time.[9] Inci et al. showed in their case report that lesions may be disappear with time.[10] The scintigraphy and bone formation biomarkers examined in our case revealed that the lesions were not active. In addition, disappear of lesions was not observed during follow-up.

In conclusıon when a rare disease such as OPK affecting multiple joints is encountered, differential diagnosis should be made with metastatic bone tumors, and differential diagnosis methods such as bone scintigraphy should be used for this purpose. Knowing the typical radiological appearance and involvement sites of this rare situation is important for the diagnosis of the disease. Informing the patients about the diagnosis on this subject also prevents unnecessary examination when they apply to other centers. Radiological recognition of OPK is important in order to eliminate diagnostic difficulties, avoid invasive diagnostic procedures and prevent unnecessary further examination. We also aimed to emphasize the importance of thinking about rare causes such as OPK, in addition to rheumatological diseases, in patients suffering from polyarthralgia.

Conflict of interest

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Kaynaklar

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