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Case Report

Extended oligoarticular juvenile idiopathic arthritis with multiple enchondromatosis: A case report

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ABSTRACT

Juvenile idiopathic arthritis (JIA) is the most common chronic rheumatic disease in childhood. Since there is no pathognomonic diagnostic criterion, the diagnosis is made by excluding other arthritis in childhood. Multiple Enchondromatosis, also known as Ollier's disease, is an ossification disorder often seen in the metaphyseal and diaphyseal regions of long bones or the metaphyseal regions of tubular bones in the hands and feet. Pain, shortening, deformity, fracture, and transformation into malignancy may appear. Here we report a case with extended oligoarticular juvenile idiopathic arthritis (oligo JIA) who developed enchondromatosis during clinical follow-up. While the patient was in remission without medication, he presented with swelling in the hand fingers and enchondromatosis lesions with expansile lytic characteristics were seen on the radiograph. Comorbid diseases can be added in the clinical follow-up of JIA patients. Physical examination is important in terms of added comorbid disease. Our case report is important because it is the first case in which these two diseases are seen together in the literature.

Keywords: Deformity, enchondromatosis, Juvenile Idiopathic Arthritis

INTRODUCTION

Juvenile idiopathic arthritis (JIA) is defined as a heterogeneous group of diseases that includes arthritis that begins before the age of 16 and lasts longer than 6 weeks. It is the most common chronic rheumatic disease in childhood.[1] The diagnosis of JIA is based on the exclusion of other causes of chronic arthritis in childhood.[2] According to the International League of Associations for Rheumatology (ILAR) classification, JIA is classified into 7 subgroups based on clinical and laboratory findings in the first 6 months. Oligoarticular juvenile idiopathic arthritis (Oligo JIA), described as 4 or fewer joint involvement during the first 6 months of the disease, accounts for 50-80% of all JIA patients. Persistent type is described as four or fewer joints involved during the all disease course. The extended type is described as more than 5 joints involved after the first 6 months.[3]

Multiple enchondromatosis was described by Ollier in 1899. It is an ossification disorder often seen in the metaphyseal areas of the tubular bones or in the metaphyseal and diaphyseal areas of the long bones. Enchondromas can cause pain, shortening, deformity, and fractures in the bones.[4] Transformation to chondrosarcoma is observed in approximately 20-50% of cases.[5]

CASE REPORT

A seven-year-old male patient presented with pain and swollen knees at the age of 2. He has been suffering from bilateral knee

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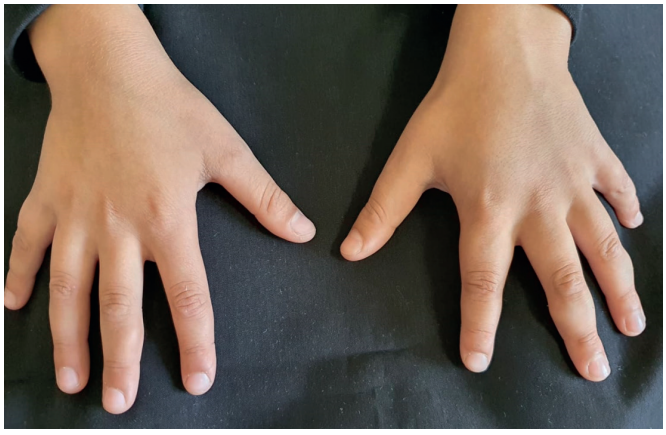

Figure 1. Swelling in the 3th finger of the right hand, the 3th and 4th fingers of the left hand.

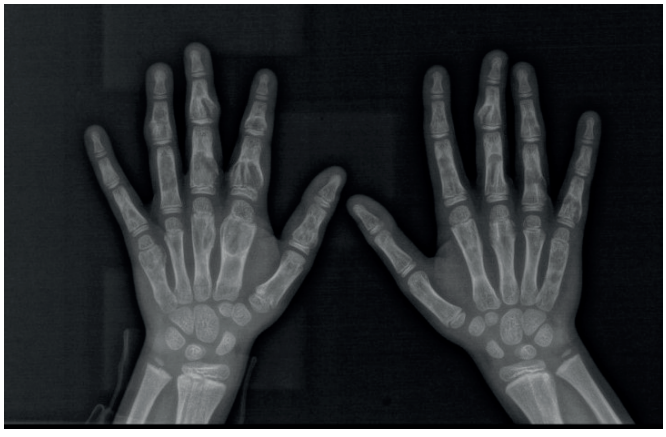

Figure 2. Expansile lytic lesions in the medullary area in the metaphysodiaphyseal area on both metacarpals and phalanges (predominantly in the proximal and middle phalanges).

arthritis for 2 months. His erythrocyte sedimentation rate (ESR) was 67 mm/h, C-reactive protein (CRP) was 15.59 mg/L, anti-nuclear antibody (ANA) was 1/1000, Rheumatoid Factor was 6.1 IU/mL, and HLA B27 was negative. Knee ultrasonography showed bilateral massive suprapatellar effusion. The patient was diagnosed with oligo JIA and treated with intra-articular steroid injection in bilateral knee joints. Both ankles and wrist joints were involved after one year. Because of the increased number of articular involvement, the patient was diagnosed with extended oligo JIA. The patient was treated with methotrexate. Methotrexate was discontinued after one year of remission. After three years of remission, the patient was admitted with swelling and deformity on the fingers. His physical examination showed expansion and deformities on the right 3rd middle

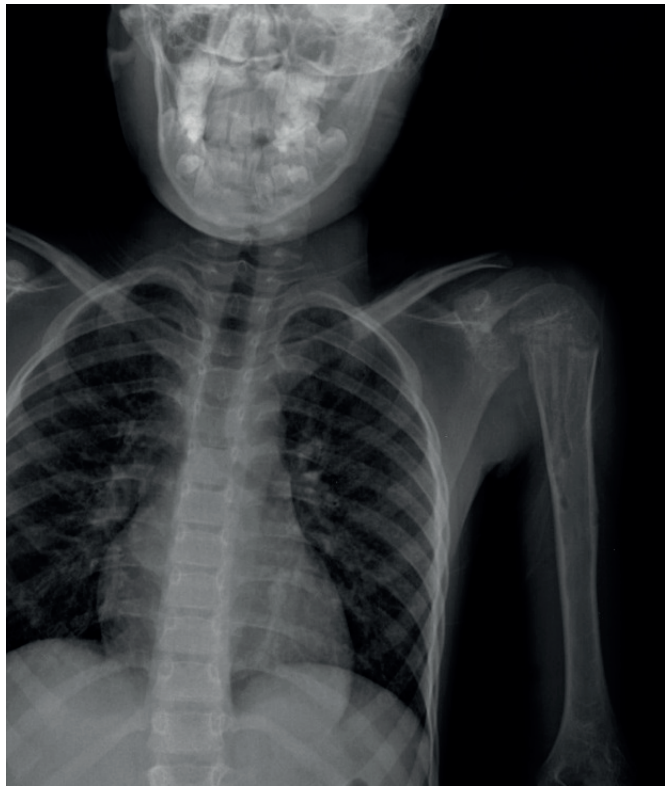

Figure 3. A lytic lesion in the medullary area extending to the left humerus proximal metaphysodiaphyseal area.

phalanx and left 3rd and 4th middle phalanges (Figure 1). The systemic and joint examinations were normal. The radiographic examination revealed expansile lytic lesions in the medullary area at the metaphysodiaphyseal area in the metacarpals and phalanges of both hands (Figure 2) and on the left humerus proximal metaphysodiaphyseal area (Figure 3). The patient was diagnosed with multiple enchondromatosis after radiographic examination.

DISCUSSION

We presented the case of a patient with extended oligo JIA and concomitant multiple enchondromatosis. Juvenile idiopathic arthritis is the most common chronic rheumatic disease in childhood. If not treated early and properly, it can cause severe deformity and disability. Non-steroid anti-inflammatory drugs, intraarticular and systemic corticosteroids, methotrexate, and biological drugs are used for treatment.[6]

Multiple enchondromatosis is a non-hereditary bony deformity that is most commonly seen in the tubular bones of the hand and foot and in the metaphyses of long and flat bones. Expansive masses and deformities occur as a result of the proliferation

of cartilage cells.[7] The disease affects both genders and all age groups. Small tubular bones of the hand are involved in 40-65% of cases; mass formation is mostly seen in the proximal phalanges, metacarpals, middle phalanges, and distal phalanges, respectively.[8] Our case also presented the involvement of fingers, which is consistent with the literature.

Juvenile idiopathic arthritis is characterized by periods of remission and activation. While some JIA subtypes have inactive diseases with treatment, some subtypes have inactive diseases without treatment.[9] Deformities may develop in patients who have persistently active diseases or who do not receive appropriate treatment. Finger deformities can be detected in inflammatory rheumatic diseases, especially polyarticular JIA, and in non-inflammatory conditions. Non-inflammatory diseases such as mucopolysaccharidosis, skeletal dysplasias, and pachydermodactyly can cause deformities.[10] Our case also presented with finger deformities during the follow-up remission period. Our patient had extended oligo JIA, and the metacarpophalangeal and proximal interphalangeal joints could be involved in the disease course. However, the absence of arthritis findings on physical examination and the detection of multiple enchondromas on the radiographs showed that a new comorbid disease was added. A good physical examination and advanced radiological imaging are necessary for the differential diagnosis. We diagnosed the disease by direct radiographs showing expansile lytic lesions on the fingers and humerus. Patients with multiple enchondromatosis may require surgical intervention for pathological fracture, deformity, or suspected malignant transformation.[11]

Our patient is the first case diagnosed with extended oligoarticular JIA and enchondromatosis together. This patient should be followed up for possible complications and malign transformation.

Ethical approval

The patient's parents provided informed consent for the publication of the report.

Author contribution

Surgical and Medical Practices: AYB, EK, ZFK, APK, MHP; Concept: APK, MHP; Design: APK, MHP; Data Collection or Processing: AYB, EK; Analysis or Interpretation: AYB, ZFK; Literature Search: AYB; Writing: AYB, APK. All authors reviewed the results and approved the final version of the article.

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Conflict of interest

The authors declare that there is no conflict of interest.

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