Case Report

Chronic Myeloid Leukemia Presenting with Avascular Necrosis of Femur Head

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Abstract

We report the case of a 15 years girl in whom avascular necrosis (AVN) of right femur head was the presenting feature of chronic stable phase of chronic myeloid leukemia (CML). To date, only three case of CML with AVN have been reported. So, in view of rarity of this condition, a similar case of CML presenting as AVN of femur head is being reported.

Introduction

Bone involvement in chronic myeloid leukemia (CML) is seen in 3 to 5% of all patients. A skeletal abnormality developing during chronic stable phase of CML usually represents ectopic areas of myeloid metaplasia, the first sign of acceleration into blastic transformation.

Avascular necrosis (AVN) of bone is considered as one of the uncommon osseous manifestations of leukemias. It is usually seen in 0.12% to 10% of lymphomas and acute leukemias (especially acute lymphoblastic leukemia) particularly after treatment with corticosteroids or radiotherapy. It is very rare to discover AVN in CML. To date, only few case of CML with AVN have been reported in English literature worldwide. We are reporting the case of a young girl with CML who presented with AVN of the femur.

Case Report

A 15 years girl presented initially to the orthopaedic department with complaint of pain around right hip joint for three months associated with difficulty in walking. There was no history of fever, weight loss or any bleeding diathesis. Physical examination revealed only mild pallor and 6 cm firm splenomegaly. There was tenderness and limitation of all movements at right hip joint and the rest of the physical examination was normal.

Investigations revealed - haemoglobin 10.8 g/dl, total leucocyte count of 290 X 10^9/L, differential leucocyte count- neutrophils 38%, eosinophils 2%, basophils 5%, lymphocytes 3%, promyelocytes 2%, myeloblast 2%, myelocytes 38% and metamyelocytes 16%, and platelet count 250 x 10^9/L. Neutrophil alkaline phosphatase (NAP) score was very low-2 (normal 40-100). The bone marrow aspirate demonstrated a hypercellular marrow with myeloid hyperplasia without any evidence of blastic transformation. The patient was found to be Philadelphia chromosome positive.

A plain radiograph of the right hip joint revealed flattening of the right femoral head, reduction in the epiphysial height of the femur head, widening of the joint space and patchy areas of sclerosis and lucencies of the femur head suggestive of AVN of the femur. A contrast-enhanced computed tomography (CECT) of the same joint confirmed the diagnosis of AVN of the femur head (Fig. 1). The left hip joint was normal.

A closed core biopsy taken from right femur head showed hypercellular leukaemic foci, mild interstitial edema along with focal areas of fibrosis and osteosclerotic changes of the bony trabeculae (Fig. 2).

The rest of the investigations including the alkaline phosphatase, uric acid, calcium and phosphorus and tests for liver and kidney functions were normal.

A final diagnosis of CML with AVN of the right femoral head consequent to leukaemic infiltration was made and

Fig. 1: Contrast-enhanced CT scan of the right hip joint.

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Received: 19.6.2001; Accepted: 21.5.2002
The various theories proposed are leucostasis, vascular compression by leukaemic foci or vascular obstruction by microthrombi in presence of disseminated intravascular coagulation. In the present case the core biopsy from femur head revealed hypercellular leukaemic foci, which presumably by causing vascular compression, could have lead to AVN.

Additionally, osteonecrosis was seen in femoral heads in the other three cases which was not demonstrated in the biopsy obtained in our case. As the core biopsy, which was performed in this patient, was a blind procedure, it probably did not include the area of osteonecrosis, which however was evident on the X-rays and CECT. Osteonecrosis is probably due to hypervasularization of juvenile femoral head and possibly explains why this complication is not seen in adult femur heads.

REFERENCES

DISCUSSION
Skeletal lesions are seen very infrequently in chronic phase of CML - in 3-5% of all cases. The various skeletal lesions described in CML include osteolysis, osteoporosis, osteosclerosis, periosteal reaction and fractures. As highlighted previously, a skeletal abnormality developing during the chronic phase of CML may represent ectopic areas of myeloid metaplasia, the first sign of acceleration into blastic transformation. Bone pain is present in the majority of individuals with skeletal lesions, and symptoms may occur before any abnormality is detectable on radiological assessment.

Only a few cases of CML presenting with AVN of bone are reported. All these cases including the present one occurred in young individuals (< 17 years). The mechanism of bone necrosis in untreated CML is poorly understood.