Pain on right knee with limping problem in an adolescent with aplastic anemia

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A 16 year-old girl was admitted to hematology department with menometrorrhagia, and pain on right knee with limping problem. In her medical history, she had been treated with corticosteroids intermittently for idiopathic thrombocytopenic purpura. We noticed leucopenia and anemia in her complete blood count. After detailed evaluation the definite diagnosis was idiopathic severe aplastic anemia. In physical examination, she was exhausted and seemed cushingoid. There were petechia and ecchymosis on her bilateral lower extremities and strias in abdominal skin besides having no organomegaly. The patient complained of pain and constraint of movement on her right knee. Perimeter of right knee was 42 cm, while left knee was 40 cm. Erythrocyte sedimentation rate was 140 mm/hr and C-reactive protein was 3.6 mg/dl (0-0.8). Plain radiography revealed cortical disarray at distal femur, tightness in joint interval, and heterogeneous sclerosis on her right knee (Figure 1). Bone scintigraphy showed non-specific alterations. We evaluated the patient in order to eliminate infections, including osteomyelitis, tuberculosis, salmonellosis, and brucellosis. There were no sign of infection. Magnetic resonance imaging (MRI) revealed radiologic findings (Figures 2 and 3). Written informed consent was obtained from the patient's parents.

What is your diagnosis?

RADIOLOGIC DIAGNOSIS

Osteonecrosis. Obvious cortical disarray at distal femur, chronical osteonecrotic alterations (heterogeneous hypointense in T_1, heterogeneous hyperintense in T_2) extending from joint surface to the epiphysis line were apparent in MRI (Figures 2 and 3).

DISCUSSION

Aseptic osteonecrosis, also known as avascular necrosis of bone, is a disorder characterized by segmental death of one more osseous site. Osteonecrosis

Fig 1. Cortical disarray at distal femur, tightness in joint interval, and heterogeneous sclerosis. Plain radiography of right knee

Fig 2. Right knee, MRI T_1

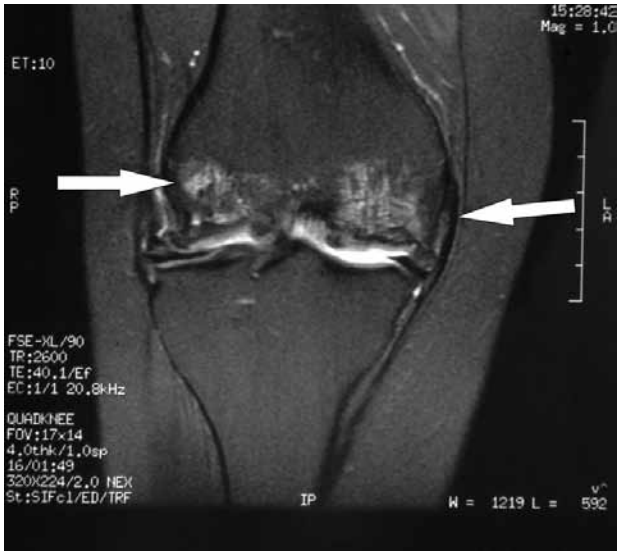

Fig 3. Right knee, MRI T_2

can accompany a wide variety of disease spectrum. Conditions accompanied with osteonecrosis in children and adolescents are Leg Calve Perthes disease, congenital dysplasia, Gaucher disease, hemophilia, sickle cell hemoglobinopathies, renal failure with osteodystrophy, disorders treated with prolonged use and/or high doses of glucocorticosteroids. The association between steroid use and the development of osteonecrosis has been known for fifty years (1). Although pathogenesis is multifactorial, increased fatty infiltration in the bone marrow of patients with aplastic anemia is a key factor in the development of osteonecrosis (2). Corticosteroids increase the probability and severity of osteonecrosis in aplastic anemia patients. Osteonecrosis is commonly multiarticular and bilateral. Knee and hip joints are the most frequent affected sites. The clinical spectrum of osteonecrosis ranges from asymptomatic patients to patients with significant bone pain and loss of function, sometimes necessitating total joint replacement (3). Early diagnosis is important for the treatment of the osteonecrosis. Bone scintigraphy is more efficient to diagnose in the early phase of osteonecrosis (4). However, MRI has been proven to be the technique of choice to identify osteonecrosis.

References

1. Sala A, Mattano LA Jr, Barr RD. Osteonecrosis in children and adolescents with cancer -an adverse effect of systemic therapy. Eur J Cancer 2007;43:683-9.
2. Park J, Jun J, Kim Y, et al. Osteonecrosis of the hip in patients with aplastic anemia. J Korean Med Sci 2002;17:806-10.
3. Wang GJ, Cui Q, Balian G. The Nicolas Andry award. The pathogenesis and prevention of steroid-induced osteonecrosis. Clin Orthop Relat Res 2000;370:295-310.
4. Mitchell DG, Rao VM, Dalinka MK, et al. Femoral head avascular necrosis: correlation of MR imaging, radiographic staging, radionuclide imaging, and clinical findings. Radiology 1987;162:709-15.