Malignity in Paget's Disease

Piedad León Rubio, Marta Llanes Gómez, José Javier Pérez Venegas, and Sergio García Pérez

Sección de Reumatología, Hospital Universitario Puerta del Mar Cádiz, Spain

Clinical Case

An 88 year –old woman was diagnosed with Paget's disease of the right iliac bone 1 year before, without having any need for treatment. She came to the emergency room complaining of pain during the past 3 months, which worsened with movement, woke her up at night and was not relieved with common analgesics. Systemic and neurologic examination was normal and the musculoskeletal system exploration demonstrated pain upon palpation and limitation of active and passive movement of the right hip in all of its plains, without an increase in the volume of soft-tissue. The hematologic study, blood chemistry, and bone metabolism was normal, with a normal C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR), and an alkaline phosphatase (AP) of 130 (normal, 20-140) U/L. A simple x-ray of the pelvis showed a large lytic lesion in the core of a pagetic right iliac bone, with cortical destruction and fractures (Figure 1). A computerized tomography (CT) of the pelvis and hips was carried out, which confirmed the presence of a lytic lesion with a large amount of bone destruction, compatible with an osteosarcoma (Figure 2).

Diagnosis and Progression

A biopsy of the lesion was compatible with osteosarcoma. A bone gamma scan showed that the lesion was localized and did not affect other structures. The patient, who was not a candidate to surgical treatment due to her age, underwent palliative care for the pain. She died after 6 months due to an exacerbation of heart failure.

Discussion

Paget's disease is a disease of unknown cause whose prevalence depends on age, country, and geographical area; in Spain, prevalence is 1% of the population older than 55 years of age, while in the United Kingdom it is of the order of 4.5%. It is characterized by excessive and abnormal bone remodeling in localized areas of the skeleton. It can be observed as a casual finding on x-rays. Its clinical presentation varies according to the localization and distribution of the lesions[1,2]. Among the complications related to this disease, malignant degeneration appears in less than 1% of patients

Figure 1. *Simple pelvis x-ray, which shows a large lytic lesion in the core of the pagetic right iliac bone with cortical destruction and fractures.*

Correspondence: Dra. P. León Rubio.
Sección de Reumatología. Hospital Universitario Puerta del Mar.
Real, 250, 1.º A. 11100 San Fernando. Cádiz. España.
E-mail: piedadleonrubio@hotmail.com

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Figure 2. *Computerized tomography (with tridimensional reconstruction) of the pelvis that shows destruction of the right iliac bone.*

between 55-80 yeas of age, fundamentally in males and patients with polyostotic disease[3,4]. Cell types more frequently associated with the disease are osteosarcoma (50%-60%), fibrosarcoma (20%-25%), and chondrosarcoma (10%). Typical localizations are the femur, pelvis, and humerus, and are rarely present on vertebrae. They are usually accompanied by intense, recent-onset pain, which is resistant to treatment and is associated to an increase in soft-tissue volume. Occasionally a fracture can be the first symptom of sarcomatous degeneration. Radiologically, these lesions can be lytic and associated to a soft-tissue mass, with cortical destruction, bone fragmentation and /or fractures that show no sign of consolidating. An increase in ESR or AP can be related with the presence of sarcoma. In this case they had the peculiarity of being normal and were not helpful for patient follow-up. This fact has also been pointed out in the review by Cerdá et al,[4] where no correlation between biologic findings and malignization can be found.[5]

The therapeutic approach includes, depending on localization, a maximal surgical resection of the tumor mass, chemotherapy and, occasionally, radiotherapy. Nonetheless, prognosis is poor with a survival of 8% of patients at 2 years. Causes of death include lung metastasis or local complications of the disease.

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