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Images in Clinical Rheumatology

43-Year-Old-Male With a Right Pelvic Mass☆

Varón de 43 años con masa pélvica derecha

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Clinical Case

A 43-year-old male came to the clinic due to mechanical lower back pain, which had lasted for 2 years. Physical examination showed painful limitation for right leg flexion and extension, with no neurological deficit. Laboratory analysis highlighted hemoglobin 10.6 g/dl, normal ESR and GGT 111 U/L (0–40). Simple X-ray (Fig. 1), CT (Fig. 2) and MR (Fig. 3) of the hips evidenced a 90 mm×80 mm×80 mm mass in the right hemipelvis with an osteolytic component which affected half of the cotillus, iliac and pubic ramus, displacing the bladder and subjacent structures leftward.

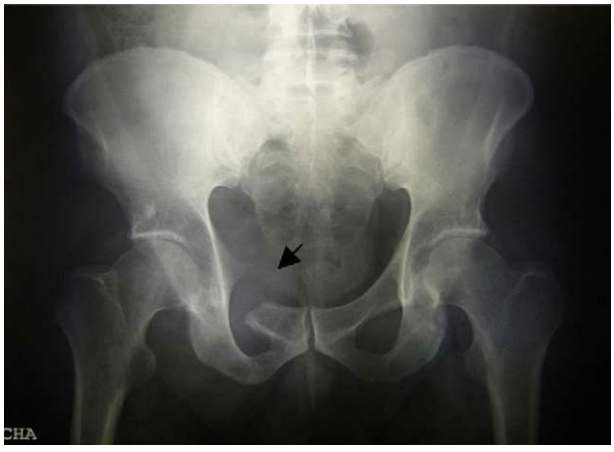

Fig. 1. Pelvic X-ray showing osteolysis (arrow) of the superior pubic ramus, with irregular bony margins and loss of cortical bone with no periosteal reaction.

Diagnosis and Progression

The histological study of an ultrasound guided biopsy concluded that the patient presented a peripheral primitive neuroectodermal tumor (PNET)/Ewing's sarcoma (ES). Chemotherapy was started with alternating cycles of vincristin/cyclophoshamide/adriamicine-iphosphamide/etoposid.

Discussion

Primitive neuroectodermic tumors are neoplasias with a differentiation to neuronal tissue which, if involving peripheral nerves, is called peripheral primitive neuroectodermic tumor.[1] Because it shares the t(11;22) translocation with ES, it is considered the same disease. This tumor represents 3%–6% of solid tumors and 1.4%–1.8% of malignant processes, with an incidence of 3 cases/million/inhab./year.[2] 90% of the cases appear between 5 and 30 years and is more common in men. It is manifested by pain (in the pelvis, femur or humerus),[3,4] swelling and, sometimes, fever, weight loss, anemia and leukocytosis.

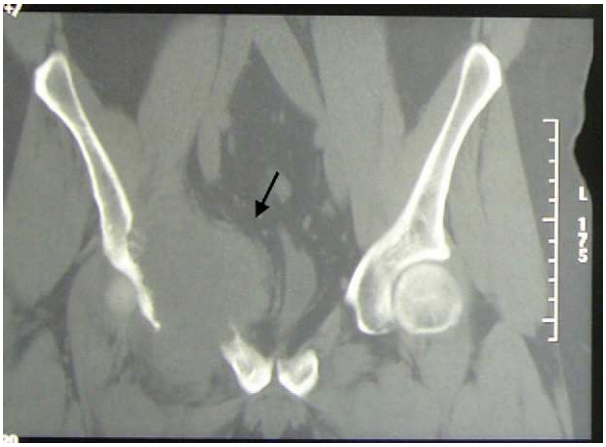

Fig. 2. Pelvis CT: lysis of the medial or superior portion of the acetabulum extended to the iliac and superior pubic ramus, associated with a large soft tissue mass measuring 12 mm×74 mm×73 mm (arrow), infiltrating muscle.

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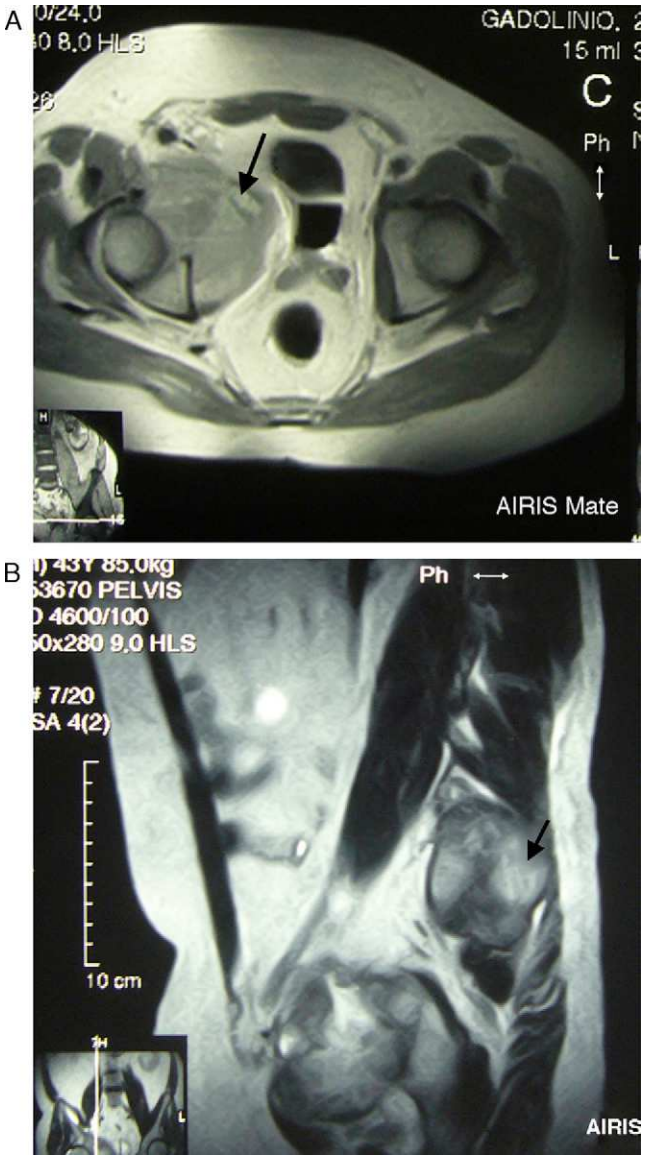

Fig. 3. (A and B) Axial and coronal MR of the hips RM in T2 sequence with contrast. A voluminous mass in the right hemipelvis is shown, affecting the anterior half of the cotillum, the iliac and pubic ramus, displacing the urinary bladder and adjacent structures leftward, with an isointense signal with bone and increased intensity with contrast, except in areas of intense necrosis.

X-ray findings reflect osteolysis, erosions, periostitis and soft tissue masses, as in our case. Immunohistochemistry is important to establish the differential diagnosis.[5]

Important findings in our patient were: age at presentation was uncommon as was the insidious nature of its progression, with no constitutional symptoms, fever or laboratory abnormalities. The natural history of PNET/ES is unknown although it tends to be aggressive in tumors developed in deep areas, when size is large or those presenting translocations.[6] A combination of surgery, radiotherapy and chemotherapy attain an increase in survival and disease-free survival.[7,8]

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