Multicentric malignant transformation of multiple exostoses

Abstract We treated a patient with large multiple chondrosarcomas derived from multiple cartilaginous exostoses. One sarcoma originated in the left pubic bone and the other sarcoma in the posterior aspect of the greater trochanter of the left femur. Thirty months after hindquarter amputation, the patient is alive without relapse. This is the first report of a patient with synchronous multiple malignant transformation of multiple cartilaginous exostoses.

Key words Multiple osteosarcoma · Malignant transformation · Chondrosarcoma

Introduction

Osteochondroma is the most common bone tumor and represents approximately 35–50% of the benign bone tumors [4]. Occasionally it appears as multiple osteochondroma and is hereditary. Malignant transformation of multiple cartilaginous exostosis has been reported to occur in 5–25% of the patients [4, 7]. Large lesions of centrally located bones have a greater potential for malignant transformation and should receive special attention [8]. Multiple malignant transformation of the benign cartilaginous lesions is extremely rare, and is reported only in Ollier’s disease [3, 6]. We treated a patient with two synchronous chondrosarcomas at different sites where previous osteochondromas had been located.

Case report

A 71-year-old man had a history of small hard bony masses in the left inguinal region and left knee joint dating back to adolescence. In December 1988, enlargement of the tumor in the left inguinal region and proximal thigh became evident. He had felt slight pain in the inguinal area for 2 months. In November 1993, he was referred to our hospital. He had no family history of multiple bony masses or bone sarcomas.

He went about on crutches. At his first visit, his left leg was extraordinarily hypertrophied. In the left inguinal area, a football-sized bony hard mass was palpable. His left hip joint was contracted. In the medial side of the left knee was a palpable egg-sized tumor and in the ventral portion of the femur a first-sized bony hard mass. Blood cell count and erythrocyte sedimentation rate were within normal limits. C-reactive protein was 0.6 (normal range: <0.5). The other serum chemistries were within normal limits.

Radiographs of the pelvis depicted a football-sized tumor in the left hip area. It had the appearance of a single osteochondroma and is hereditary. Malignant transformation of the tumors originating from the pubic bone and the proximal femur was impossible on the roentgenogram (Fig. 1). In the left proximal tibia (Fig. 2) and diaphysis of the anterior surface of the left femur, tumor masses were identified on the roentgenogram. At the hip joint level, CT demonstrated a large, high-density, cauliflower-like tumor protruding from the left pubic bone into the an-
terior side of the left hip (Fig. 3). In the intertrochanteric region of the posterior surface of the left proximal femur, another tumor was located, which protruded into the soft tissue (Fig. 4). This tumor had a low-density cap compared to the bone protruding from the femur. The bone scan showed large hot areas in the left pubic bone, hip region, and proximal femur. Urinary bladder and colon were shifted and not infiltrated by tumor. On T1-weighted MR images, the tumor of the pubis and proximal femur consisted of low signal intensity masses. On T2-weighted images, the postero-inferior area of the tumor from the pubis and the posterior area of the tumor originating from the greater trochanter had a high signal intensity (Fig. 5).

Preservation of the sciatic nerve and femoral artery would have been impossible if a safe surgical margin was selected. In November 1993, the patient underwent hindquarter amputation. Thirty months after surgery, neither local recurrence nor metastasis has developed.

Macro- and microhistology
After surgical treatment, the resected specimen was analyzed macro- and microhistologically (Fig. 6). Big tumors were identified in the os pubis (26 cm in diameter), trochanteric area (18 cm in diameter), anterior side of the distal femur (12 cm in diameter), and proximal tibia (5 cm in diameter). Tumors originating from the pubis and the trochanteric area were diagnosed as chondrosarcomas. In the pubic tumor, there was a cartilage mass measuring 17x13 cm. In the trochanteric area, the cartilage cap of the tumor was over 2 cm in thickness.