Massive osteolysis in the pelvis—a case report

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In 1985, a 42-year-old woman felt pain in her left hip while playing volleyball, but continued to play occasionally because her symptoms were mild. In 1987, the pain worsened. Past and family histories showed no remarkable findings.

She consulted a local physician and radiolucency and fractures of the left ischium and pubis were noted on plain radiographs (Figure 1) and CT. A diagnosis of lymphangioma of the bone was made by incisional biopsy. Curettage and autogenic bone grafting were performed. The left pelvic osteolysis progressed and in 1988, 1 year after surgery, the bone graft was resorbed. In 1990, 3 years after surgery, the sacrum was also involved and osteopenia of the left femur was detected. A few years later, radiographs showed dislocation of the left hip with progression of the osteolysis of the left ilium. Fracture of the pelvis occurred twice. Treatment with calcitonin and vitamin D failed to prevent progression of the disease.

Figure 1. Radiograph taken in 1987 after initial consultation at a previous hospital showed radiolucency and fracture of the left ilium and pubis.

In 1996, the hip pain recurred after a fall and she was referred to our hospital. She could walk with 2 crutches. She complained of pain on motion, but no pain at rest. There was a swelling of the left buttock and hip joint, but no tenderness and no heat. Active motion of the left hip was limited. The circumference of the right thigh was 47 cm and that of the left thigh 54 cm.

Radiographs showed almost complete osteolysis of the left pelvis and partial osteolysis of the sacrum (Figure 2). The left femur was also affected and had fractured. 3-D-CT showed a large osteolytic lesion (Figure 3). MR images showed that the left pelvis had vanished and was replaced by an area of low signal intensity on T1-weighted images (Figure 4). A high signal intensity area was found on T2-weighted images (Figure 5). Bone scintigraphy showed loss of nor-

Figure 2. Initial radiograph taken in 1996 at our hospital showed almost complete osteolysis of the left pelvis and partial osteolysis of the sacrum. The left femur was also affected and fractured.

Figure 3. On 3-dimensional CT, a broad osteolytic lesion was demonstrated.

Figure 4. The left pelvis had vanished and was replaced by an area of low signal intensity on T1-weighted MR images, as indicated by the arrows.

Figure 5. A high signal intensity area was found in the same area on T2-weighted images. Arrows indicate high signal intensity area of the tumor.

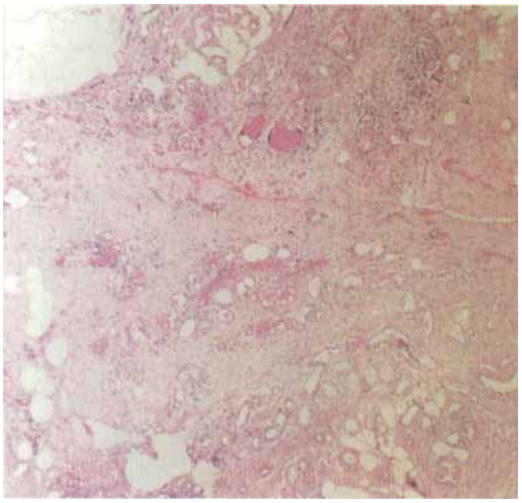

Figure 6. In the parosteal soft tissues and the bones, dilated vessels with thin walls were noted.

mal uptake in the left pelvis. ^{201}Tl scintigraphy showed no abnormal uptake. Angiography of the left internal iliac artery was normal.

On incisional biopsy, profuse amounts of clear and colorless fluid, similar to spinal fluid, flowed from the incision. Histologic examination demonstrated inconclusive findings, although dilated vessels were noted in the bone and in the parosteal soft tissues (Figure 6).

40 Gray of irradiation was administered to the lesion. During 6 months after radiotherapy, no progression was seen and the patient could walk with an orthosis and 2 crutches with slight pain. But 1 year after radiotherapy, there were signs of progress.

Discussion

Massive osteolysis, Gorham-Stout syndrome, Gorham's disease or disappearing bone disease is rare, with about 140 reported cases (Kulenkampff et al. 1990, Green et al. 1995). The disease leads to partial or complete disappearance of the affected bones (Gorham and Stout 1955, Cannon 1986). It usually occurs in persons younger than 40 years of age. Pelvis, shoulder bones or long bones are most frequently involved (Kulenkampff et al. 1990, Green et al. 1995). Symptoms are mild compared to the radiological findings and a pathological fracture may lead to the diagnosis (Kulenkampff et al. 1990, Cannon 1986). In our case, with almost complete bone loss of the left pelvis and dislocation of the left hip, the patient did not complain of pain at rest. Histology shows intraosseous hemangiomatous and/or lymphangiomatous changes (Kulenkampff et al. 1990, Stöve and Reichelt 1995). Life span is rarely limited, but there is no effective treatment of the disease. Calcitonin, vitamin D, excision with bone grafting, excision with prosthetic replacement, radiotherapy and chemotherapy have been tried with varying degrees of success (Kulenkampff et al. 1990, Green et al. 1995).

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