Case report

Alveolar soft part sarcoma of bone

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Introduction

Alveolar soft part sarcoma (ASPS) is a rare soft tissue tumor that accounts for approximately 0.5%–1.0% of soft tissue sarcomas, commonly originating from the deep soft tissue.1 In 1999 Park et al. first reported six cases of primary ASPS of bone and showed their radiological findings of the bone origin.2 We report a case of ASPS arising in the ilium, which was shown by computed tomography (CT) and magnetic resonance (MR) images.

Case report

A 33-year-old Japanese woman suffered from left buttock pain for 9 months, with a palpable firm mass over the left iliac crest. She had been bedridden because of increasing pain for 1 month prior to her initial visit to our hospital. On physical examination, a firm mass with a poorly delineated margin, measuring approximately 15 × 10 cm, was palpable over the left iliac crest. Routine laboratory examinations did not indicate any abnormality.

Plain radiographs revealed an ill-defined osteolytic lesion without any mineralization (Fig. 1). CT scanning demonstrated a destructive process in the left ilium with bilateral soft tissue masses, exhibiting a central low density area (Fig. 2). MR imaging showed an expansile iliac tumor with large intra- and extrapelvic soft tissue masses. T1-weighted images showed moderately high signal intensity with a central low-signal area associated with an intra- and extrapelvic soft tissue masses (Fig. 3a). Short T1 inversion recovery (STIR) images showed moderately high signal intensity with foci of signal void corresponding to dilated blood vessels (Fig. 3b). Angiography of the left common iliac artery revealed a hypervascular tumor (Fig. 4a), which stained densely in the capillary phase (Fig. 4b) and venous phase (Fig. 4c). The chest radiographs showed multiple nodular lesions in both lungs.

An open biopsy was carried out. Histology revealed solid nests of tumor cells arranged in an alveolar pattern and separated by thin-walled, sinusoidal vascular channels. The rounded or polygonal tumor cells exhibited centrally located round nuclei with prominent nucleoli and abundant eosinophilic granular cytoplasm (Fig. 5). Periodic acid-Schiff (PAS) preparations demonstrated diastase-resistant, positive granules in the cytoplasm (Fig. 6). Electron microscopic examination revealed intracytoplasmic crystalline structures showing filaments arranged in a parallel fashion. Also seen were electron-dense secretory granules (Fig. 7).

Radical tumor resection was not possible because of multiple lung metastases. The tumor was treated with embolization, intraarterial infusion chemotherapy based on pirarubicin- (80 mg/m² × 3) and ifosfamide-based intravenous chemotherapy (1.0 g/m² × 3). We added radiation therapy with 65 Gy in total. These treatments did not appear to be effective. Three years later metastases to the brain developed. She died of the disease 62 months after the initial presentation. Consent for publication has been obtained from her families.

Discussion

Alveolar soft part sarcoma has rarely been reported3–5 since the first description by Christopherson et al.6 in 1952. Most of these lesions occur in the deep soft tissue, especially in the lower extremities. Its histogenesis is still unknown. MyoD1 protein expression suggests that ASPS represents an unusual variant of skeletal muscle tumor.7 Wang et al. reported that an immunocytochemi-

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ical and biochemical study of myogenic regulatory protein expression did not indicate a skeletal muscle tumor.\(^8\)

In 1999 Park et al.\(^2\) reported six cases of ASPS primarily involving bone and described their radiological findings. The primary sites of the tumor were the femur in three patients, the fibula in two, and the ilium in one. Both the tibia and the fibula were involved in one of the patients. The radiographic features were nonspecific but suggestive of a malignant nature. All cases showed bone destruction with ill-defined margins, and their epicenter was in the bone. The findings indicated that the tumors occurred primarily in bone.

In our patient, cross-sectional imaging demonstrated that the epicenter was located in the ilium, and the

Fig. 1. Plain radiograph of the left hemipelvis shows a large, ill-defined osteolytic lesion in the ilium

Fig. 2. Computed tomography (CT) shows a destructive lesion in the left ilium that extends to the adjacent soft tissue

Fig. 3. Magnetic resonance images (MRI) of the left pelvis. a T1-weighted image shows a large mass of moderately high signal with a low-signal area in the central portion. b Short T1 inversion recovery (STIR) images shows a mass of high-signal intensity in the left ilium with an extraosseous component involving the gluteal muscle. Signal voids are seen in the tumor