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Dedifferentiated parosteal osteosarcoma with high-grade osteoclast-rich osteogenic sarcoma at presentation

Abstract  We report a case of a 32-year-old woman who presented with parosteal osteosarcoma of the distal femur with simultaneous dedifferentiation to a high-grade osteoclast-rich osteogenic sarcoma. This pattern of dedifferentiation is rare, particularly at the time of presentation. We are aware of three other somewhat comparable cases in the literature; however, none is quite similar to our case.

Key words  Parosteal osteosarcoma · Osteoclast-rich osteogenic sarcoma · Concurrent · Dedifferentiation · Femur

Introduction

Dedifferentiation of tumors is a biologic event that is clinically significant. This usually involves transformation of a low-grade malignant neoplasm that follows a slow progressive course, to a high-grade malignant tumor with a greater metastatic potential and a more rapid lethal clinical course.

Case report

A 32-year-old woman presented in August 1992 with pain in her left knee of several weeks’ duration following a twisting injury. A roentgenogram showed a large mass in the popliteal fossa intimately related to the posterior lateral aspect of the distal femur (Fig. 1). MRI confirmed the presence of a lobulated mass attached to the lower posterior femur by a broad ossified base. Ossification was mostly at the base not the periphery. There was no involvement of the underlying medullary bone (Fig. 2). An open incisional biopsy of the mass was performed. Histologically, the lesion consisted of two distinct components (Fig. 3). One component showed parallel “streamers” of woven bone separated by a cellular fibrous tissue with minimal cellular atypia (Figs. 4, 5). There was an abrupt transition to a second component, which showed an osteoclast-rich sarcoma (Fig. 6) with atypical mitoses, scattered anaplastic giant cells (Fig. 7), and focal production of osteoid (Fig. 8). The lesion was initially regarded as a giant cell tumor and wide excision was recommended. A few weeks later, the patient underwent an en bloc resection of the distal left femur. This included the tumor (6×4.1×6 cm) with underlying cortical table (8×2×0.5 cm). The initial biopsy at that time was reviewed and a revised diagnosis of parosteal osteosarcoma with dedifferentiation to high-grade osteoclast-rich osteosarcoma was made. The en bloc resection specimen showed no residual component of parosteal osteosarcoma. The tumor consisted of a high-grade osteosarcoma with numerous osteoclast-like giant cells. The margin of the specimen was involved by tumor. Postoperatively, the patient received three courses of chemotherapy (adriamycin and cisplatinum). Three months later, an above-knee amputation of the left leg was performed. A white-gray lobulated tumor mass was found in the posterior distal femur, measuring 4.8×1.0×1.5 cm. It showed several histological patterns. Some areas showed pleomorphic spindle cells, abundant osteochondroid matrix, and scattered osteoclast-like giant cells. Other areas were composed of sheets...
of osteoclast-like giant cells in a background of mononuclear cells with numerous mitoses (up to two per high-power field, 40x objective). The tumor showed 80% necrosis. The histological features of parosteal osteosarcoma were not identified. The patient received postoperative cyclophosphamide, actinomycin D, and bleomycin. In October 1993, she developed metastases to the right and left lower lobes of the lungs. Both tumor nodules were surgically excised. In February 1995, September 1995, and May 1996, tumor nodules were again removed from both lungs. The metastatic tumor deposits consisted of osteoclast-rich osteosarcoma with osteoid production. The patient has had no further recurrences.

Discussion

Parosteal osteosarcoma is a rare neoplasm which in some series accounts for approximately 5% of all osteosarcomas [1, 2]. It is a well-differentiated, predominantly fibro-osseous variant of osteosarcoma that arises from the juxtacortical region of long bones. The most common site is the posterior aspect of the distal femur. Although the peak patient age is in the second decade, the tumor usually occurs at a later age than conventional intramedullary osteosarcoma.

Radiologically, these tumors are eccentrically placed, lobulated, and densely mineralized lesions that appear to be “pasted on” the cortex. The base of the lesion is usually more ossified and the outermost portions generally are the least mineralized. The lesion is attached to the underlying cortex by a broad base with a lucent zone present between the underlying cortex and the tumor [3, 4]. In one study of parosteal osteosarcomas, the underlying cortex was considered normal in approximately 50% of cases. In approximately 25% the cortex was thickened and in the remainder it was destroyed [5].

The histology of parosteal osteosarcoma is that of a fibro-osseous lesion with relatively well-formed osseous trabeculae, commonly arranged as parallel “streamers” separated by cellular fibroblastic tissue with minimal nuclear atypia. There is usually minimal to no involvement of the underlying medullary bone. The cortex is often thickened and deformed.