Case report 656

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Clinical information

A 55-year-old man complained of progressively worsening pain in the left ankle. The patient had a history of generalized psoriasis which had been treated 5 years before by extensive local corticotherapy. He was a heavy smoker and suffered from a severe arthropathy. Alcohol abuse was also noted, without pancreatitis. Physical examination revealed tenderness in the distal portion of the left leg. Laboratory findings and blood studies were within normal limits.

Radiographs of the left ankle, including tomographs, demonstrated a lytic, poorly margined, polycyclic area in the distal tibial metaphysis (Fig. 1 A, B). Periosteal reaction and endosteal erosion with posterior cortical bone destruction were present. No soft tissue mass was detected. In other metaphyseal regions of both tibiae, there were sharply demarcated zones of increased density suggesting multiple medullary bone infarcts (Bls). Furthermore, frontal roentgenograms of the right hip showed necrosis of the femoral head, clinically latent.

A 99mTc-MDP bone scan demonstrated increased uptake in the distal end of the left tibia. The scan was otherwise normal. CT scans of the left tibial metaphysis demonstrated a lytic tumor eroding the posterior cortical, having its origin in a centrosseous calcified area (Fig. 1 C).

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**Diagnosis: Malignant fibrous histiocytoma in a previous bone infarct**

Magnetic resonance imaging (MRI) demonstrated in the four metaphyses of both tibiae and in the distal right femur serpiginous areas of low signal intensity on T1- and T2-weighted images (Fig. 2) characteristic of mature bone marrow infarction. Apart from these lesions, MRI showed in the distal third of the left tibia cortical erosion, suggesting malignant degeneration of one of the Bls (Fig. 3A). No soft tissue mass was detected.

An open biopsy confirmed cortical destruction by a soft tumor mass transgressing the periosteum. Microscopical examination revealed multiple sarcomatous cells, growing in a storiform pattern (Fig. 4). Numerous focally necrotic zones, dystrophic calcifications, and nonviable lamellar bone trabeculae were present in the tumor.

The patient underwent an under-the-knee amputation. On gross examination, the metaphyseal cortical bone was transected. In a sagittally cut section there was a yellowish area in the medullary cavity corresponding macroscopically to infarcted bone. Around the area of the infarction, a gray soft tissue mass was present, destroying the posterior cortical bone and periosteum (Fig. 3B, C). Microscopically, this sarcomatous tumor developed a triphasic pattern of cells: spindle cells with foamy cytoplasm, plump cells with histiocytic appearance, and giant cells. The spindle cell component was largely predominant and arranged in a storiform pattern. In some areas, numerous capillaries were present, suggesting an hemangiopericytoma-like pattern. Electron-microscopic evaluation confirmed the presence of fibroblastic, myofibroblastic, and malignant histiocytic-like cells. The lesion was diagnosed as a malignant fibrous histiocytoma around a bone infarct, without metastasis. No adjuvant treatment was prescribed.

**Discussion**

Although malignant fibrous histiocytomas (MFH) is well known among the tumors of the somatic soft tis-
MFH of bone is rare (1%–5% of primary bone tumors) and sometimes develops in association with pre-existing bone abnormalities (20%–25% of MFHs), like bone infarct (BI) [14]. However, MFH arising within or associated with skeletal infarction is exceptional, since only 32 cases (reviewed in [4, 7]) have been tabulated in the literature. Analysis of the reported cases reveals that such patients had frequently large, multiple, metaphyseal medullary infarcts of the long bones. These BIs were related to decompensation sickness [15], vasculitis [13], alcohol abuse [7], sickle cell anemia [8, 15], or steroid therapy [2, 17]. As in the present case, these conditions have more often been associated with asecptic necrosis of the epiphyses, which never degenerate. A long interval between the occurrence of BI and MFH diagnosis is frequently observed.

Our patient had abused alcohol and received steroid therapy through extensive topical steroids for his generalized psoriasis. In addition, our case documents the existence of a previous BI present for more than 5 years before the MFH was diagnosed. An earlier arteriogram (1983) had clearly demonstrated its presence.

Typically, MFH associated with BI [1] occurs in a preponderence of men over 40 years of age. It involves almost exclusively the femur and the tibia. Pain and swelling are the main complaints.

Radiologically, MFH is characterized by a destructive, purely osteolytic, ill-defined, metaphyseal medullary area. The cortex is frequently destroyed. Periosteal reaction and soft tissue mass are often noticed. Radiological studies also demonstrate associated mature BIs characterized by large, serpiginous, medullary calcifications [5].

CT shows the osteolytic tumor and its relationship with the centrosseous calcifications of bone marrow infarction. Furthermore, endosteal erosion and soft tissue mass are easily detectable. Operative planning requires knowledge of the local extent of the tumor, and CT is commonly required to locate it precisely. 99mTc-MDP and 67Ga citrate scans demonstrate intense accumulation of the radiopharmaceuticals in the tumor [12], and the remainder of the scan can be used to search for possible osseous metastases. The usefulness of angiography [9] has been reported in planning the operative treatment.

MRI is a sensitive examination in the detection of avascular necrosis of the femoral head [3, 11] and the knee [19]. Typically, a low signal area is present on T1- and T2-weighted images. In some patients, the margins of the lesion become hyperintense on long pulse sequences, showing an inflammatory tissue at the reactive interface. These MR features are similar for medullary bone infarction [18]. Thus, serpiginous metaphyseal zones of low signal intensity are detected in T1-weighted images, as well as similarly unenhanced signal in the T2-weighted sequences. A thin band of low signal delimits the margins of the BI [2, 16]. In some cases, the central portion is isointense with the surrounding normal bone marrow, indicating fatty areas.

In addition, our report suggests the ability of MRI to discriminate malignant sarcomatous transformation of medullary BIs by demonstrating diaphyseal cortical erosion. Furthermore, MRI delimits the tumor mass and determines the presence or absence of soft tissue invasion. Despite its classic lack of specificity, contrasting with its high sensitivity in detecting skeletal tumors, the association of medullary infarction with malignant transformation allows prediction of the presence of an MFH.

Histologically, the following characteristics typify MFH [14]: mononuclear ovoid to spindle histiocytic cells growing in a storiform pattern (characteristic but not pathognomonic of MFH), anaplastic multinucleate giant cells, evidence of phagocytosis, heavy reticulin production, and fat production. MFH of bones exhibits a wide range of histological patterns. With this in mind, Huvos et al. [10] have subdivided MFH into three variants according to the histological appearance of growth: the predominantly fibroblastic pattern, the predominantly histiocytic or xanthomatous morphologic variant pattern, and the malignant giant cell tumor type. In our case the tumor had a predominantly fibroblastic pattern.

BIs are characterized by dense bland fibrosis, dystrophic calcifications, and foci of nonviable lamellar woven bone. Around the BI, a chronic reparative process develops at the revascularizing margins, and the proliferation of histiocytes is an essential component of this process. It has been suggested [15] that, after many years, in some instances, the histiocytic component may undergo sarcomatous transformation. Thus, the tumoral histiocyte can differentiate along two major directions: towards phagocytic functions or fibrogenic functions. In the present case, the localization of the tumor around the BI reinforces this chronic reparative theory for the malignant transformation of BIs [13].

In summary, a case is presented of a 55-year-old man with a MFH in a previous BI. The poor prognosis of such secondary MFH is related in part to the difficulty of its early diagnosis, its aggressive behavior, and its tendency to metastasize (mainly to bone sites and lungs). Nevertheless, although biopsy of asymptomatic bone marrow infarction is not indicated, MRI seems consistent in detecting sarcomatous transformation of symptomatic BIs and plays an important role in the patient's preoperative evaluation.

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References