

Primary fibrosarcoma of the distal femur. Report of a rare case

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Abstract— Fibrosarcoma of bone is a relatively rare malignant neoplasia, representing less than 5% of all bone tumors. It is distinguished from other bone sarcomas by the presence of spindle-shaped tumor cells located among intertwining bundles of collagen fibers, without any other type of histological differentiation, such as the formation of cartilage or bone.

In the present report, we present a rare case of primary fibrosarcoma in the distal femur in a 45-year-old patient with complaints of mild pain and swelling in the right knee joint for approximately 6 months. After imaging evaluation, a biopsy was performed; in the second stage, a subsequent resection of 16 cm of the distal femur and its replacement with a tumor endoprosthesis was performed. Three months postoperatively, no local recurrence or metastasis of the tumor was observed.

Keywords— fibrosarcoma, diagnosis, surgical treatment.

1. Introduction

Fibrosarcoma (FS) of bone is a malignant mesenchymal tumor characterized by immature proliferating fibroblasts or undifferentiated anaplastic spindle cells. On immunohistochemical study, the tissue is negative for the specific markers characteristic of other bone tumors, so the diagnosis of FS is a diagnosis of exclusion. FS of the bone can be primary or less often secondary after radiotherapy in the area or other pathological conditions (Paget's disease, chronic osteomyelitis, progression or dedifferentiation, mainly from chondrosarcoma, bone marrow infarction, fibrous dysplasia, etc.)^{8,12}.

MacDonald and Budd⁹ in 1943 and Phemister¹¹ in 1948 were among the first to distinguish FS of bone as a separate neoplasm of the skeleton. Its incidence ranges from 4% to 8% of all primary bone tumors, occurring in the second to seventh decades of life. Secondary FS comprises approximately 25-30% of bone FS. FS involves the metaphyses of long bones, especially the distal femur. Isolated diaphyseal or epiphyseal localizations are extremely rare. Exceptionally, multiple fibrosarcomas are also found.

The most important prognostic sign is the histological grade. Treatment for FS of the bone is wide resection, amputation, or disarticulation^{1,8,12}.

FS is characterized by slow progression of complaints; patients usually report mild to moderate pain in the area of the lesion. After a significant increase in size and the presence of an extraosseous component, a dense to firm tumor mass could be palpated. In undifferentiated FS, the clinical picture is crucial, with rapid deterioration of the general condition^{1,8}.

2. Case report

A 45-year-old patient was admitted to our clinic with complaints of mild but persistent pain and swelling in the area of the right knee. No clinical evidence of circulatory and neurological deficits in the limb was established. The complaints existed from 6 months. No enlargement of regional lymph nodes or distant metastases was detected. After a biopsy, a primary FS of the distal femur was diagnosed. The planning of the radical removal of the tumor was after evaluation of the roentgenography (Figure 1) and MRI with contrast (Figure 2). A successful radical resection of the tumor 16 cm from the knee joint was performed, and the defect was replaced by a tumor endoprosthesis (Figure 3). The postoperative period was uneventful, with prompt rehabilitation of the knee following the clinical protocol. The patient was referred to the oncodispensary for observation. Three months postoperatively, the patient did not use mobility aids, a flexion of 65 degrees was reported in the knee joint (Figure 4), and no local recurrence was observed (Figure 5).

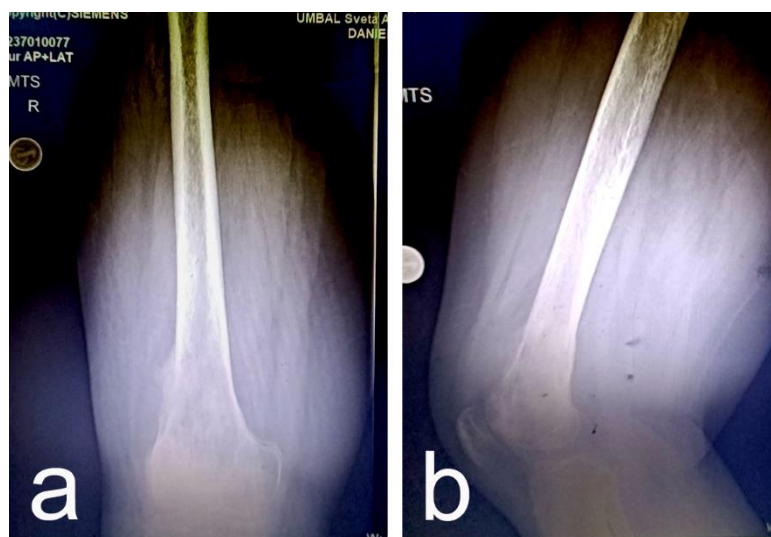


Figure 1. Preoperative roentgenography

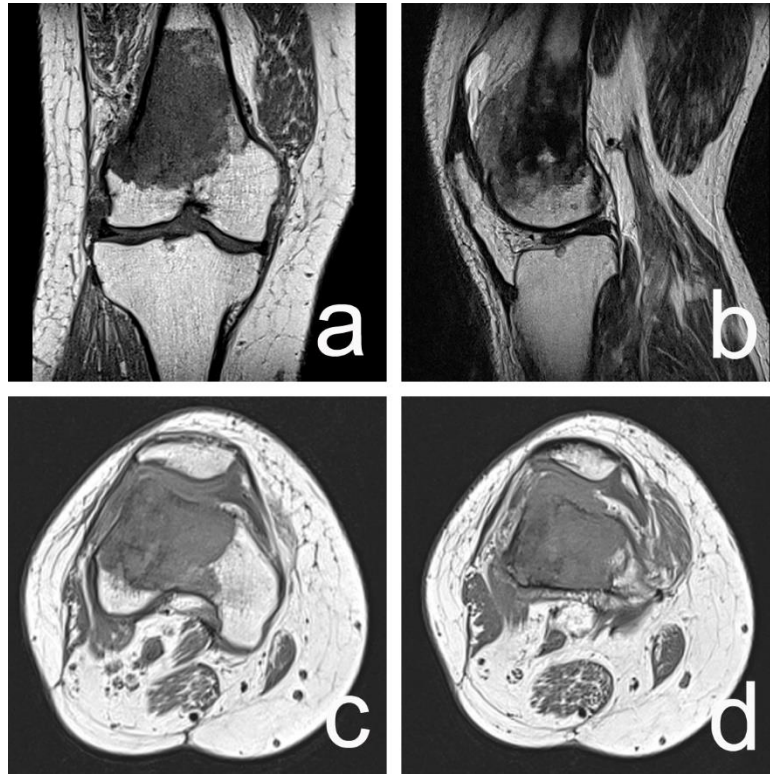


Figure 2. Preoperative MRI

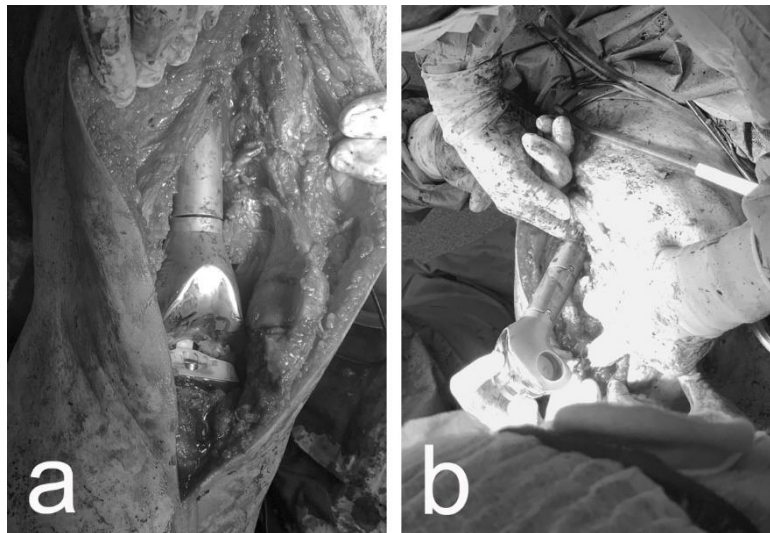


Figure 3. Intraoperative photography



Figure 4. Postoperative photography

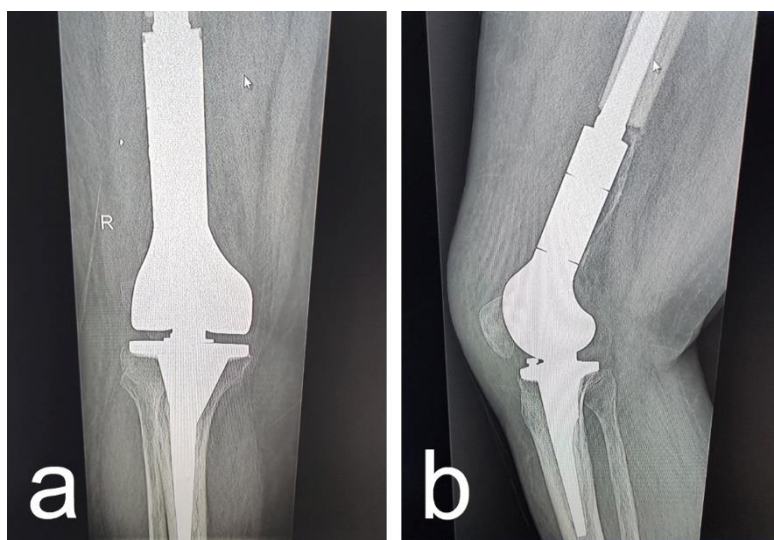


Figure 5. Postoperative roentgenography after 3 months

3. Discussion

The most common localizations of bone FS are the metaphyseal region of the distal femur, the proximal tibia, and the pelvic ring. Clinical manifestations are not specific; however, pathologic fractures are a common complication and are seen in approximately 18% of patients at their first presentation to the physician^{1,8}.

For the early diagnosis of FS, imaging studies are of great importance. In general, FS is described as a poorly circumscribed osteolytic lesion. Most often, the inner side of the cortex is eroded or completely destroyed. No periosteal reaction was observed. However, it should be noted that the radiographic picture is not specific, and the differential diagnosis includes giant cell tumor of bone, malignant fibrous histiocytoma, telangiectatic osteosarcoma, lymphoma, solitary plasmacytoma, and metastatic carcinoma. The absence of calcification or ossification is of diagnostic importance, as such findings are commonly seen in chondrosarcoma and conventional osteosarcoma. CT and MRI present in detail the localization of the formation, its relationship to nearby vascular-nerve bundles, and the presence or absence of metastases^{1,7,8}.

The correct diagnosis and radical treatment are the key factors for survival. Radical surgery consists of the complete resection of the tumor within wide ranges and corresponding chemotherapeutic treatment, despite the lack of clinical data on its effectiveness. The type of surgical procedure (en bloc resection, disarticulation or amputation) depends mainly on the stage, local status and location of the tumor. Amputation is indicated when radical resection cannot be performed or in cases of local recurrence after en bloc resection^{1-7,8}.

Because of the significant rarity of FS of bone, the role of chemotherapy in these patients has not been well studied, and the regimens used thus far are those of osteosarcoma treatment^{8,10}.

Despite the low incidence of local recurrence, the 5-year survival rate was low (28%). This can be attributed mainly to the development of metastatic disease in most patients (63%). The presence of local recurrence is a poor prognostic marker. FS metastasizes most often to the lung or other parts of the skeleton^{8,10}.

4. Conclusion

Adequate surgical treatment by wide resection or amputation is the treatment of choice for patients with primary FS of bone. The most important prognostic factors are the age of the patient, the localization and the stage of the tumor. In the presence of recurrence after resection, disarticulation or amputation is the method of choice, as well as chemotherapy, although outcomes are not well studied due to the rarity of the FS of bone.

5. References

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