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Research Paper

Surface osteosarcoma: Clinical features and therapeutic implications

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ABSTRACT

Introduction: Surface osteosarcoma are rare variant of osteosarcoma that include parosteal osteosarcoma, periosteal osteosarcoma and high grade surface osteosarcoma. These lesions have different clinical presentation and biological behavior compared to conventional osteosarcoma, and hence need to be managed differently.
Goal: The aim of this study is to analyze the clinico-pathological features and outcome of a series of surface osteosarcoma in an attempt to define the adequate treatment of this rare entity.
Patient and method: It is a retrospective and bicentric study of 18 surface osteosarcoma that were seen at the KASSAB's Institute and SAHLOUL Hospital from 2006 to 2013. The authors reviewed the clinical and radiologic features, histologic sections, treatments, and outcomes in this group of patients.
Results: Seven patients were male (38.9%) and 11 were female (61.1%) with mean age of 25 years (range from 16 to 55 years). Eleven lesions were in the femur and 7 in the tibia. We identified 11 parosteal osteosarcoma (six of them were dedifferentiated), 3 periosteal osteosarcoma and 4 high grade surface osteosarcoma. Six patients had neoadjuvant chemotherapy and all lesions had surgical resection. Margins were wide in 15 cases and intra lesional in 3 cases. Histological response to chemotherapy was poor in all cases. The mean follow up was 34.5 months. Six patients (33.3%) presented local recurrence and 8 patients (44.4%) presented lung metastases. Six patients (33.3%) died from the disease after a mean follow up of 12 months (6–30 months); all of them had high grade lesions.
Conclusion: Histological grade of malignancy is the main point to assess in surface osteosarcoma since it determines treatment and prognosis. Low grade lesions should be treated by wide resection, while high grade lesions need more aggressive surgical approach associated to post operative chemotherapy.

1. Level of evidence: IV

1.1. Introduction

Osteosarcoma are primary malignant bone tumors in which neoplastic cells produce osteoid. The spectrum of lesions is very wide, which can be distinguished either by their grade of malignancy, the differentiation of neoplastic cells or even according to their location related to the bone. Most of sarcoma arise intramedullarly, and only few of them develop on the outer surface of the cortex when they are called "surface osteosarcoma" [1]. They are classified into three subtypes: parosteal osteosarcoma (POS), periosteal osteosarcoma (PerOS) and high grade surface osteosarcoma (HGSO). They account for 5%, 1.5% and 0.5% of all osteosarcomas respectively [2]. Classic parosteal osteosarcoma (cPOS) originates from the outer fibrous layer of the periosteum. Its cells are fibroblasts. It is characterized by the low grade of anaplasia and slow indolent course. Occasionally, it presents a progression in malignancy and becomes dedifferentiated (DPOS). Periosteal osteosarcoma developes within the inner, germinative layer of the periosteum (the cambium layer) from cells differentiating in osteoblasts and chondroblasts. Its histological grade of malignancy is intermediate. HGSO arises from the surface of bone and it is entirely high grade, with a high mitotic activity identical to that of conventional OS. We report the clinical data and the oncological outcome of 18 cases of surface osteosarcoma and suggest the appropriate management.

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2. Patients and methods

We reviewed retrospectively the clinical data of 18 patients treated between 2006 and 2013 for a surface osteosarcoma. Patients were managed in 2 centers: Kassab's National Institute and Sahloul Hospital. Clinical data were available from patients' charts. Imaging records and histological slides were reviewed and discussed respectively with a radiologist and a pathologist that are specialized in musculo-skeletal pathology. Inclusive criteria were bone sarcoma producing osteoid originated at the surface of the bone regardless the grade of malignancy and the degree of medullar extension. The mean follow-up was 34.5 months (range: 6–96).

We analyzed the survival, local recurrence, and distant metastasis outcomes using survivorship techniques. Duration of symptoms was analyzed with *t*-test of student. We used Kaplan–Meier survival analysis to determine the cumulative probability of survival, survivorship free of recurrence, and survivorship free of metastasis. Differences in survivorship curves were evaluated using log-rank tests and p values less than 0.05 were considered significant. Statistical summaries, analyses, and plotting of survival curves were performed using SPSS version 22.

3. Results

3.1. Clinical findings

Seven patients were male (38.9%) and 11 were female (61.1%). The mean age of patients was 25 years (range from 16 to 55 years). All lesions involved the appendicular skeleton. Seven of them (38.9%) were in the right side and 11 (61.1%) were in the left side. The most common location was the knee (9 cases: 50%), with the distal part of the femur involved in 8 cases. The duration of symptoms before diagnosis ranged from 3 to 48 months (mean 12.6 months). cPOS had the longest duration compared to high grade lesions (22.5 vs. 12.1 months). The most common symptom was swelling, present in all patients. It was painful in 16 patients (88.8%). Only 4 patients (22.2%) had joint stiffness, mainly knee flexum. Clinical data are summarized in Table 1.

3.2. Imaging features

Roentgenographic findings were analyzed for all patients. Seventeen patients had been evaluated with magnetic resonance imaging and 10 patients had computed tomography.

Mainly, two patterns were observed. The first one corresponded to parosteal lesions and the second one to periosteal and high grade surface lesions.

- Parosteal tumors were the most frequent ones with 11 cases (61.1%). They had a predilection for the posterior cortex of the distal metaphysis of the femur (8 cases: 72.7%). One case involved the proximal metaphysis of the tibia and 2 cases were diaphyseal: one in the femur (case 1) and one in the tibia.
  Typically, the tumor had a melon-shaped, mineralized mass, pasted on the cortex (Figs. 1a and 2a). The tumor size ranged from 5 to 10 cm. The pattern of mineral was amorphous (50%) (Fig. 1a), lobulated (40%) (Fig. 2a) or striated (10%). There was a thin radiolucent zone between the tumor and the underlying bone (Figs. 1a and 2a) in 81.1% of cases. A speculated periosteal reaction was present in only one case (9%). The underlying cortex was eroded or destroyed in 4 cases (36.3%). In the larger, broad-based tumors, the cortex of the host bone was expanded by a new-bone formation in 60% of cases. Medullary involvement was clearly shown on plan radiographs in 5 cases (45.4%). It was focal in 2 cases (Fig. 2a) and diffuse in 3 cases.
  Seven of these lesions were evaluated by CT scan wich was useful to study the attachment of the tumor to the host bone, medullary involvement and the presence of radiolucency within the mineralized component (Figs. 1b and 2b).
  MRI was available in 10 patients. It best evaluated local extension of the tumor and especially medullary involvement. This was observed in 8 cases (72.7%). It was focal in 5 cases (Fig. 2c) and diffuse in 3 cases.
  Medullary extension was suspected radiologically in two cases of the cPOS (50%) and in all cases (100%) of DPOS.
- The second pattern was that of PerOS (3 cases) and HGSO (4 cases). All lesions were diaphyseal and periosteal reaction was constant. Typically, lesion had saucer shape appearance characterized by a cortical thickening at the periphery with Codman triangle and a non mineralized soft tissue mass at the center (Figs. 3a and 4a). The soft tissue mass was characterized by an aggressive periosteal reaction perpendicular to the osseous long axis and causes scalloping that affected only the thickened cortex (Fig. 3a). There was no abnormality in the adjacent medullary canal. Fine ring calcifications were seen in 3 cases (42.8%) and they were related to the chondroblastic differentiation of the lesion (Fig. 4a).

HGSO were radiologically more aggressive. While PerOS affected only one part of the diaphyseal cylinder (Fig. 3a), HGSO were circumferential in 2 cases (Fig. 4a). Soft tissue mass was less mineralized in HGSO than in PerOS.

All patients were evaluated by MRI. Lesion margins were well defined with a pseudocapsule which corresponds to the periosteum (Figs. 3b and 4b). The marrow signal intensity was normal in all patients with PerOS (Fig. 3b) and abnormal in HGSO (Fig. 4b and c). In later cases, bone marrow invasion was focal in 3 cases (Fig. 4b) and diffuse in one case. MR images obtained after intravenous gadolinium administration showed a marked degree of enhancement in all patients. The enhancement pattern was the thick peripheral and septal enhancement with nodularity pattern in all cases (Figs. 3b and 4c).

3.3. Histological findings

Tumors were classified into the following subtypes:

- *Parosteal osteosarcoma* (*11 cases*: *61.1%*): Among parosteal tumors, 5 (45.4%) were low grade lesions (cPOS) with hypocellular spindle_cell stroma that contained well differentiated bone trabiculae (Fig. 1f). Spindle cells showed minimum cytological atypia and rare mitotic activity. Three lesions were graded 1 and two graded 2. Four were fibroblastic and one chondroblastic. In one case, the osseous trabeculea of the neoplasm had cement lines simulating the appearance of Paget disease and in one case they had fibrous dysplasia appearance.
  Six of the parosteal osteosarcomas (54.6%) were considered to have undergone dedifferentiation (DPOS); that is, a high grade spindle cell sarcoma co-existed with a lower grade, more typical parosteal osteosarcoma (Fig. 2e). Four cases were recognized at the time of the initial diagnosis (synchronous lesions) and in two cases, dedifferentiation occurred at the time of recurrence (metachronous lesions). Cartilaginous component was identified in 4 cases (66%). In two cases, cartilage was predominant. Three cases were osteoblastic and one fibroblastic.
  Medullary involvement was observed histologically in only one case in the group of cPOS, but was constant in the dedifferentiated lesions.
- *Periosteal osteosarcomas* (*3 cases*: *16.6%*): Tumors were diagnosed histologically as moderately differentiated (Grade 2 or 3). All cases were predominantly chondroblastic (Fig. 3c). Bone marrow was not invaded in any case.