PRIMARY BONE LIPOSARCOMA IN CHILDREN

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ABSTRACT

Introduction. Even though the soft tissue liposarcoma is a frequent tumor, the primary bone liposarcoma is very rare, being localized especially in the long bones.
Material and meyhod. We present the case of a 14 years old female child, hospitalized in the „Sf. Maria" Emergency Clinic Hospital for Children Iasi accusing pain, functional impairment and tumefaction in the right arm, symptoms which suddenly occurred about 24 hours prior to presentation, following a falling trauma on the right arm. The bone x-ray emphasized a tumor in the proximal part of the humerus bone, associated with a pathological fracture.
Results. A biopsy form the tumor was performed and the pathology report (histology and immunohistochemistry) documented a malignant liposarcomatous proliferation.
Conclusion. The final pathology diagnosis, correlated with the clinical findings, which excluded the possibility of a bone metastasis, was that of a primary bone liposarcoma. Case particularity: very rare tumor, witch presented with a pathological bone fracture. In establishing the diagnosis of a bone liposarcoma it is very important to exclude a bone metastasis from a liposarcoma with a primary localization other than the bone, as well as other primary bone tumors. The prognosis seems to be better than in the osteosarcoma but liposarcoma presents a higher rate of local recurrence and systemic dissemination.

Keywords: bone, liposarcoma, child

INTRODUCTION

Primary bone liposarcoma are extremely uncommon tumors, despite the fact that the bone marrow is rich in adipose cells. The disease is sufficiently rare to justify reporting isolated cases.

CASE PRESENTATION

Female child, 14 years-old, is admitted in the Pediatric Surgery Department for pain, functional impairment, tumefaction localized in the right arm, symptoms with a sudden onset 24 hours prior to presentation. She had no pathological history and a good performance status at the time of the presentation.

The local exam: 1/3 proximal region of the right hummers bone tumefaction, of firm consistency, painful both spontaneously and when mobilized, limiting the passive and active movements in the scapulohumeral joint.

The laboratory tests revealed an inflammatory syndrome: neutrophilia, elevated fibrinogen and lymphopenia.

The radiological exam of the right arm reveals an osteolytic lesion in the 1/3 proximal region of the humerus and a right humerus pathological fracture.

The CT scan is suggestive for an osteosarcoma and a pathological fracture, without displacement.

Surgery is performed and multiple biopsies from the bone tumor and the peritumoral areas are done,

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one drain tube was installed and postoperative immobilization in swaddle for two weeks was performed.

The extemporaneous exam of the tumor performed during the surgical intervention on frozen sections coloured with Blue toluidine is suggestive for a liposarcoma, which is why a special frozen coloration is performed – Scharlach. The suspicion is thus strengthened by the presence of the fatty drops from the tumor cells cytoplasm.

Prints: May-Grumwald-Giemsa staining– infrequent tumor cells with atypical cyto-nuclear features and optical empty vacuoles in some of the cells. Infrequent inflammatory cells present.

Paraffin: fragment from the extemporaneous exam and number 3 surgical piece represents an adipocyte-like tumoral cellular proliferation with a marked cyto-nuclear pleomorphism and sarcomatous vessels in the stroma. Infrequent bone lamellas with an "isolated" aspect and the following characteristics:

1. connective tissue is infiltrated with tumor proliferation bone lamellas
2. tumor proliferation with myxoid areas and round cells areas.
3. tumor infiltrate in the bone tissue
4. tumor proliferation, myxoid cells areas, round cells areas, tumor necrosis.

Immunohistochemistry:

CD34 – positive in the vessel's walls and the presence of capillary with a "branch" aspect

S100 – positive in some tumor cells like adult lipocyte and negative in all other tumor cells

Ki67 – positive in different proportion – 50% in myxoid areas and 80% in round and pleomorphic cells areas.

Final diagnosis: Bone liposarcoma with round cells, high malignancy grade, G3, T2a

STAGING. TREATMENT. EVOLUTION

The thoraco-abdominal CT scan revealed multiple secondary hepatic and vertebral lesions (D9, D10, D11, L1, L3), suggesting secondary pulmonary lesions.

In this context we classified the liposarcoma as stage IV and initiated a chemotherapy protocol recommended in soft tissue sarcoma in advanced stages: Ifosfamide + Doxorubicin (Ifosfamide 1500 mg/m^2 with Mesna (urological protection) days 1-2-3-4 and Doxorubicin 20 mg/m^2/day, days 1-2-3). The protocol is to be repeated every 3 weeks, according to Worden et al (1). Unfortunately, response to treatment was unsatisfactory and the disease extensively recurred locally three months from the diagnosis.

DISCUSSIONS

The bone liposarcoma is exteremely rare tumor in pediatric population with an incidence of less than 0,1% of all primary bone tumors (2). All age groups may be affected, but the usual onset is seen in adulthood (3), while the children are rarely afflicted (only 5 cases being currently reported) (2,4, 5,6,7). The most frequent localization is in the long bones: tibia, femur, humerus (3, 8), while wide bones (scapula, skull) and vertebrae are seldom affected (9). The clinical features are dominated by a painful tumor mass and the radiology exam de-

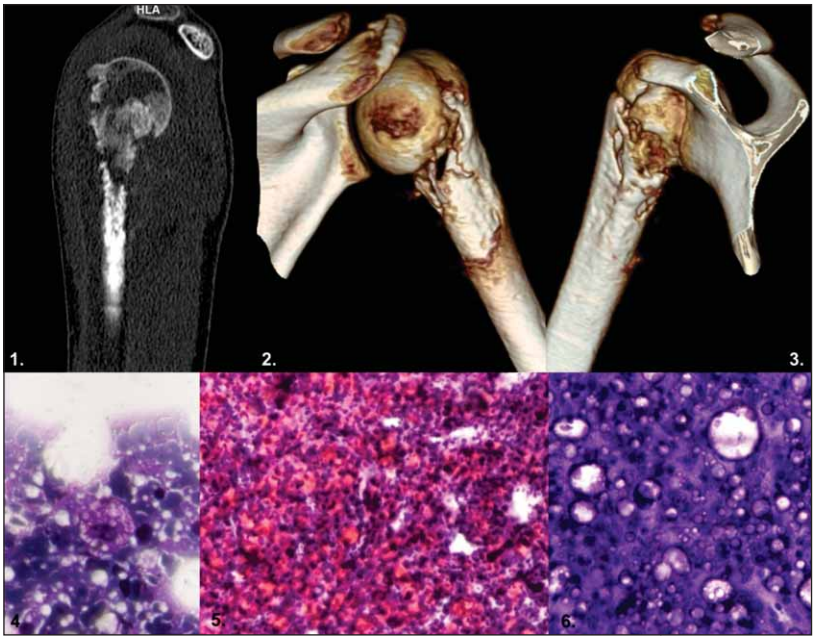

FIGURE 1.
1 – Humeral CT scan, lateral;
2 – Humeral CT reconstruction, median view;
3 – Humeral CT reconstruction, lateral view;
4 – Print. MGG, x400;
5 – Extemporaneous exam. Scharlach, x 100;
6 – Extemporaneous exam. AT, x200

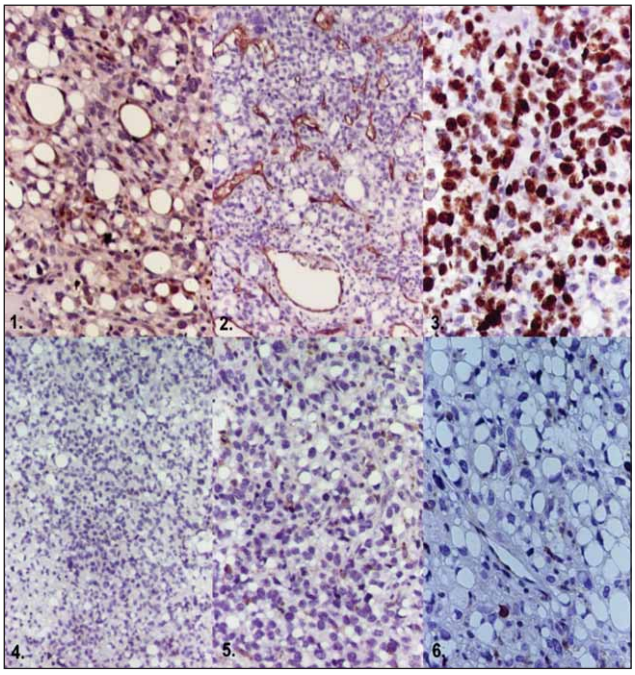

FIGURE 2. *1 – S100 x100; 2 – CD34 x100; 3 – Ki67 x100; 4 – CK AE1/ AE3 x100; 5 – CD68 x100; 6 – CD45 x200*

scribes a limited, expansive, osteolytical bone tumor. CT/MRI describes the lypidic component of the tumor. The histological variants are similar to those of the soft tissue liposarcoma (3): lipoma-like well differentiated liposarcoma, myxoid/round cells liposarcoma, pleomorphic liposarcoma. The differential diagnosis includes the soft tissue liposarcoma with local bone extension/metastasis (3), primary bone tumor, lypoma and other malignant tumors like osteosarcoma and Ewing sarcoma. Surgical excision remains the main form of therapy (broad excision/amputation and prosthesis). The role of chemotherapy is controversial but may be helpful for high- grade tumours (protocols for soft tissue liposarcoma (2)) and also local radiotherapy.

The prognosis is related to the histological type: pleomorphic liposarcoma has the least favorable prognosis (3) with an overall survival of less than three years from the diagnosis (10). Well-differentiated and myxoid types have a 100% and 88% 5-year disease free survival rates respectively (4).

The prognosis of the bone liposarcoma is generally unfavorable, but it is better than that of osteosarcoma (2).

Tumors with poorly circumscribed margin or those with local recurrences after surgery have a poor prognosis, although they rarely metastasize (5). Repeated local recurring liposarcomas may evolve into a high grade sarcoma with metastatic potential. There is a high probability of local recurrence and metastasis, most commonly to the lung (9), (10).

CONCLUSION

Bone liposarcoma is a very rare bone tumor in children; the presentation in our case was that of a pathological bone fracture. The advanced stage (IV) at the moment of the diagnosis and the lack of response to the chemotherapy led to the unfavorable evolution.

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