Prilozi, Odd. biol. med. nauki, MANU, XXVIII, 2, s. 211‡222 (2007)
Contributions, Sec. Biol. Med. Sci., MASA, XXVIII, 2, p. 211–222 (2007)
ISSN 0351–3254
UDK : 616.718.4-006.34

Case Report

HIGH-GRADE SURFACE OSTEOSARCOMA – CHONDROBLASTIC TYPE

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A b s t r a c t: We present the case of an 8-year-old boy with pain in the distal part of the right thigh that occurred after a ball injury, without haematoma or oedema. The plain film showed exostosis of the distal part of the right femur. In another institution the "exostosis-like" formation was surgically removed and the histological finding was an aggressive type of chondroblastoma.

A few months later, on the site of the surgical intervention, a significantly enlarged painful solid mass appeared. Radiography showed a large dense mass at the level of distal femoral metadiaphysis with tumour matrix mineralization, and Codman's sign. Computed tomography showed a large soft tissue mass, which arose from the surface of the underlying cortical bone and signs of high-grade malignancy. CT is a valuable method in predicting high-grade malignancy. Arteriography was also done.

After operative treatment – seq. Campanacci, histology showed high-grade surface osteosarcoma of chondroblastic type. Pre- and post-operative chemotherapy was carried out. The boy died 2 years after diagnosis because of recurrent disease.

High-grade surface osteosarcoma has the worst prognosis, compared with other surface osteosarcomas. Therefore, timely diagnosis is indispensable.

Key words: high grade surface osteosarcoma, computed tomography, diagnosis.

Introduction

Osteosarcoma is a primary malignant bone tumour in which neoplastic cells produce osteoid. Most osteosarcomas arise intramedullaily, but some arise extramedullarily on the outer surface of the underlying bones, the so-called "surface osteosarcomas" [1]. Approximately 4–10% of osteosarcomas are located on the surface of the bone [2, 3, 4]. According to the nature of the tumour, its clinical, radiological and pathological findings, surface osteosarcomas have been classified into three groups: parosteal, periosteal and high-grade surface osteosarcomas [5, 6]. High-grade surface osteosarcoma is the rarest type, with incidence of < 1% of all osteosarcomas [7]. This is important in the surgical and medical care of the patients [5, 6], as well as for the prognosis.

Case report

We present the case of an 8-year-old boy with pain in the distal part of the right thigh, which occurred after mild ball injury. There were no clinical signs of haematoma or swelling. On the initial X-ray of the right femur, an "exostosis-like" tumour formation was detected, which was surgically removed with marginal excision. The histology showed an aggressive type of chondroblastoma.

A month later, on the site of the surgical intervention, a significantly enlarged painful and palpable solid mass appeared, and the patient was referred to the Orthopaedic Clinic, with suspected diagnosis of a malignant soft tissue tumour.

On physical examination a slightly tender, non-movable, solid mass was noted, in the distal part of the right thigh.

Sedimenation rate and serum levels of calcium, phosphorus and alkaline phosphatase were within the normal rates.

The first plain film showed a soft tissue mass on the latero-posterior aspect of the distal third of the femoral diaphysis with displacement of adjacent fat lines, and positive Codman's sign (Fig. 1). One month later, on plain film, an enlarged dense soft tissue mass could be seen which arose from the surface of the underlying bone, extending along the metaphysis too. There was mild cloudy tumour matrix mineralization (Fig. 2). The mass increased in size during the next month, with an increasing periosteal reaction, Codman's sign, and intensity of the tumour matrix mineralization (Fig. 3). Radiographs that were taken each month within a period of three months clearly showed the evolution of the disease, by presenting a progression of x-ray signs of high-grade malignancy.

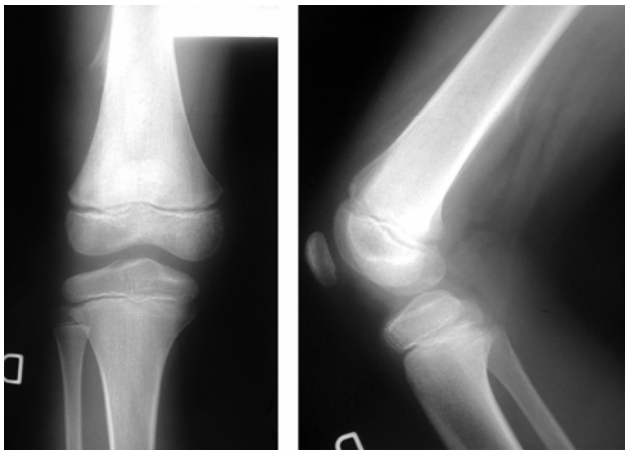

Figure 1 – Plain film showed soft tissue mass on latero-posterior aspect of distal third of the femoral diaphisis, with displacement of adjacent fat lines and positive Codman's sign

Слика 1 – Рендгенографија која покажува мекоткивна маса на латеро-постериорен аспект на дистална третина на дијафизата на фемурот, со поместување на соседните масни линии и позитивен Кодманов знак

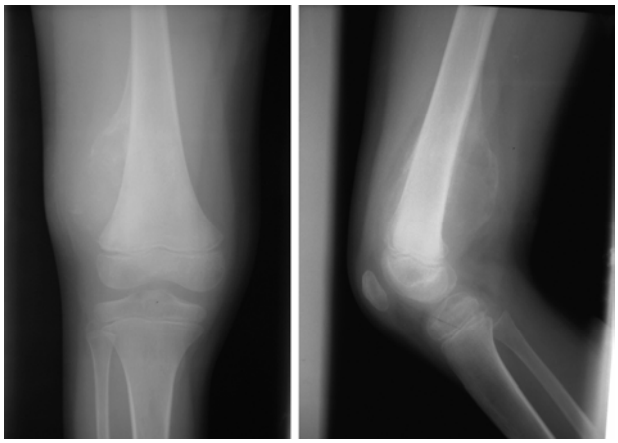

Figure 2 – Plain film, one month later; enlarged soft tissue mass, which arose from the surface of the underlying bone extending along the metaphysis. There is mild cloudy tumor matrix mineralization

Слика 2 – Рендгенографија еден месец подоцна, зголемување на мекоткивната маса, која потекнува од површината на коската и се протега долж метафизата. Постои умерена облачеста тумор матрикс минерализација

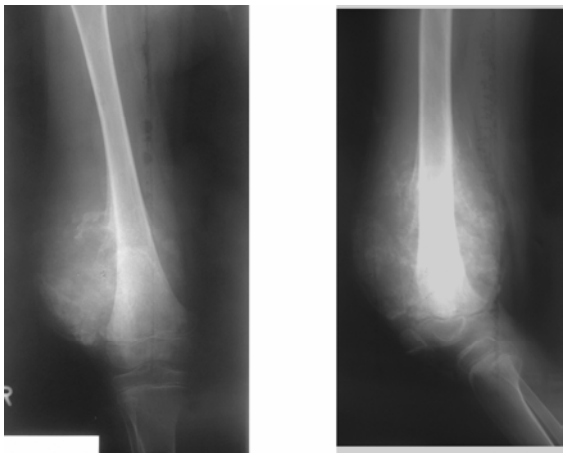

Figure 3 – Plain film, the mass increased in size over a period of next month, with severe periosteal reaction, Codman's sign and more intense tumor matrix mineralization

Слика 3 – Рендгенографија, масата ја зголемила големината во периодот на следниот месец, со јака периостална реакција, Кодманов знак и поинтензивна тумор матрикс минерализација

A transaxial CT scan at the level of the upper (Fig. 4), middle (Fig. 5) and distal parts (Fig. 6) of the tumour, demonstrated an inhomogeneous, large soft tissue mass (size 6–8cm), which arose from the surface of the underlying cortical bone. It was attached to the cortex on the broad base of the tumour with

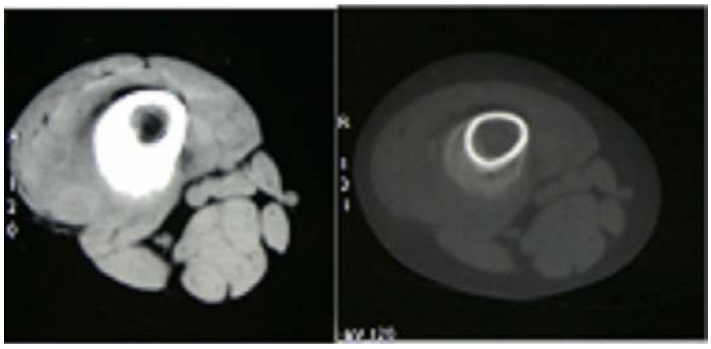

Figure 4 – Computed tomogram at the level of Codman' sign – proximal part of the tumor showed

Слика 4 – Компјутеризирана томографија на ниво на Кодмановиот знак ǂ прикажан е проксимален дел на тумороt

no defined cleavage plane or intramedullary extension. Within the soft tissue mass, there was centrally a slight cloudy tumour matrix mineralization, partially

irregular and peripheral disrupted egg-shell calcification. The cortex was not thickened and without peripheral disruption. The mass did not encircle or erode the bone. CT findings suggested a high grade of malignancy on the surface of the bone without intramedullar involvement.

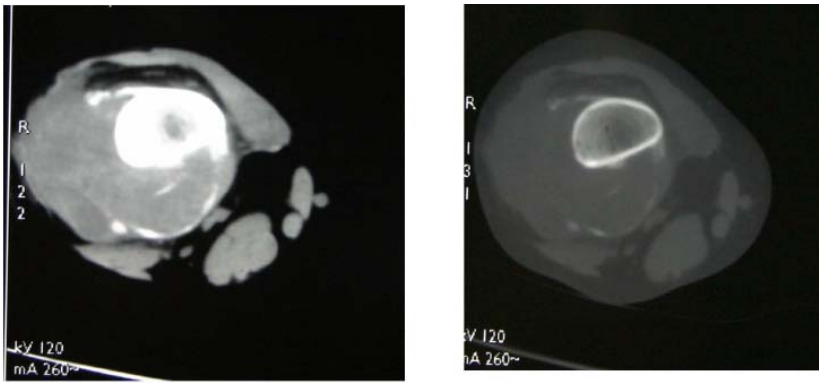

Figure 5 – Computed tomogram at medial part of the tumor demonstrated an inhomogeneous, soft tissue mass, which arose from the surface of the underlying cortical bone, with centrally cloudy tumor matrix mineralization, peripheral egg shell calcification

Слика 5 – Комūјуūеризирана ūомоґрафија на ниво на медијален дел на ūумороū кој ūрикажува нехомоґена мекоūкивна маса, која ūоūекнува од ūовршинаūа на коргексоū одоздола, со ценūрална облачесūа ūумор маūрикс минерализација и ūериферна како лушūа од јајце калцификација

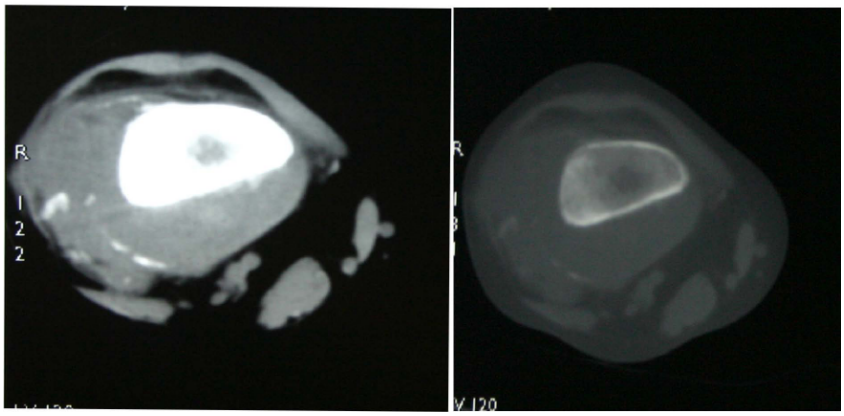

Figure 6 – Computed tomogram at distal part of the tumor. The mass do not encircle nor erode the bone

Слика 6 – Комūјуūеризирана ūомоґрафија на дисūален дел на ūумороū. Масаūа не ја заобиколува ниūу еродира коскаūа

Arteriography showed replacement of the superficial femoral artery at the tumour level, with discrete pathological blood vessels (Fig. 7). Bone scan with Tc 99m confirmed the solitary bone lesion on the distal part of the right femur.

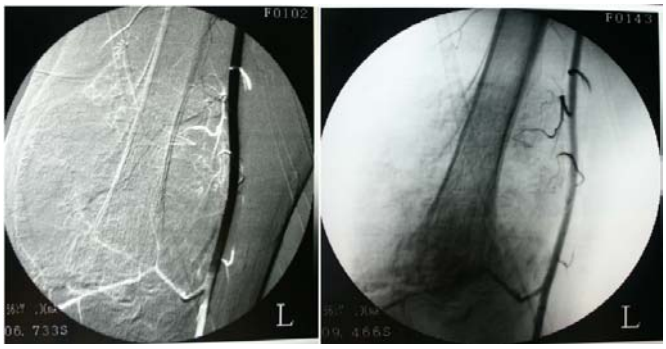

Figure 7 – Arteriography showed replacement of the superficial femoral artery at the tumor level, with discrete pathologic blood vessels
Слика 7 – Артериографијата покажува поместување на површната феморална артерија на ниво на тумороt со дискретни патолошки крвни садови

Open biopsy was carried out and the histology showed a high-grade surface osteosarcoma of the chondroblastic type.

Preoperative chemotherapy according to the COSS 96 regime was done. After 10 weeks of treatment, there was no significant tumour response and regression.

Operative treatment consisted of limb salvage procedure, with a wide excision and reconstruction of 15 cm of the distal right femur with overlying vasti, but preserving the rectus femoris. Reconstruction with Campanacci procedure, namely arthrodesis of the knee joint was done. Complete excision of the tumour, with clear surgical margins, was demonstrated on histopathological examination (Fig. 8).

A high-grade surface osteosarcoma of chondroblastic type was histologically verified (Fig. 9). The tumour tissue consisted of closely packed spindle cells with marked cellular and nuclear pleomorphism. The cells' nuclei were hyperchromatic, with prominent nucleoli and frequent atypical mitoses. Tumour cells with bizarre shapes and more than one nucleus were abundantly present. Deposits of slender lace-like seams of osteoid were found among the cells. In addition, among the tumour cells, there were lobules of highly malignant cartilage made of hyaline matrix and lacunae filled with atypical and pleomorphic cells. Diffusely, areas of haemorrhages and necroses were found. The tumour was designated as a poorly differentiated (Broders' grade 4) high-grade osteosarcoma of the chondroblastic type.

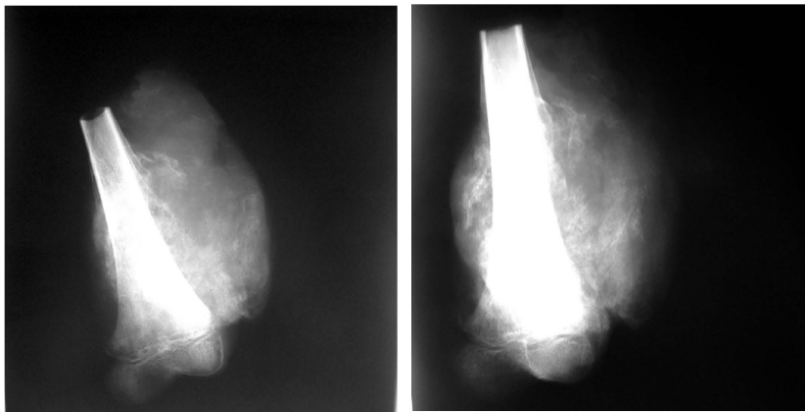

Figure 8 – X-ray of the specimen
Слика 8 – Рендгенографија на примерокот

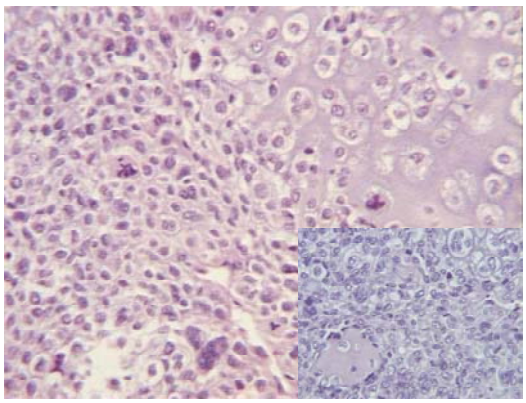

Figure 9 – HE x 400: Marked cellular and nuclear pleomorphism of malignant cells with bizarre shapes and frequent atypical mitoses. Lobules of highly malignant cartilage made of hyaline matrix and lacunae filled with atypical cells. Deposits of osteoid (insert)

Слика 9 – ХЕ x 400: Назначена целуларност и јадрен плеоморфизам на малигни клетки со бизарна форма и чести атипични митози. Лобули од високо малигна 'рскавица составен од хиалин матрикс и лакуни исполнети со атипични клетки. Депозити на остеоид (вметнат)

The treatment continued with postoperative chemotherapy. One year after surgery the patient was walking without difficulties with a normal gait, had no extensor lag and was pain free. Control CT showed no local recurrence of the disease, no distant metastases and laboratory findings were within normal range. The boy died 2 years after diagnosis because of recurrent disease.

Discussion

High-grade surface osteosarcoma is the rarest type, having an incidence of < 1% of all osteosarcomas [7]. It is considered more aggressive than that of the other surface osteosarcomas, classic osteosarcomas [1, 6, 8, 9].

In the larger studies, the ages at diagnosis have ranged from 8 years to 70 years, mean 25 years, 66% were males [6]. We present a case with early presentation of the disease, in the first decade of life of an 8-year-old boy.

High-grade surface osteosarcoma should be differentiated from other osteosarcomas arising on the surface of the bone because it is associated with a comparatively poor clinical outcome [6].

Previous reports described the similarity of radiological features of high-grade surface osteosarcoma to those of parosteal and periosteal osteosarcoma, but the histological features of the tumour are characterized by high-grade (grade III–IV) osteoblastic or fibroblastic type osteosarcoma [8, 11, 12]. Parosteal and periosteal are usually diaphysal and show perpendicular periosteal reaction, but high-grade surface osteosarcomas usually surround a much higher percentage of the bone circumference, and are more likely to invade the medullary canal that shows low attenuation at CT [13], Murphey MD *et al*. report on circumferential involvement of the host bone in 28% of patients, half of them greater than 50% [6]. In our case the tumour was localized on the distal part of the femur. CT clearly showed no intramedullar involvement. On the transaxial CT scan circumferential involvement was around 65%.

The amount of tumour matrix mineralization and distribution in the lesion varied from dense to moderate with a fluffy, immature appearance, predominantly at the base of the lesion [6, 10]. Tumours with less ossification tended to be less differentiated than tumours with more abundant ossification [16]. In our case the tumour was attached to the underlying bone with a broad base, where mild cloudy tumour matrix mineralization was present, mostly irregular and peripheral disrupted egg-shell calcification, suggesting a high grade of malignancy. In contrast to parosteal osteosarcomas, lucent zones between the tumour and underlying cortex were absent. Periosteal osteosarcomas typically show a speculated mineralization perpendicular to the long axis of the affected bone. Tumour matrix mineralization can be depicted on plain film, but CT is the method of choice for evaluation of localization, type and arrangement of mineralization.

Alteration of the underlying cortex of the affected bone was seen commonly in high-grade surface osteosarcoma, presented like: cortical destruction 17 lesions, cortical thickening 7 and both in 4 lesions [6]. In our case, CT showed that cortex was not thickened and was without peripheral disruption.

Periosteal reaction is rare with no tumour Codman's triangle or speculated periosteal reaction [6]. However, in our case there was a rapidly growing of tumour, with a severe periosteal reaction over a period of three months. Plain film well depicted Codman's triangle, that was also shown on CT. In addition, soft tissue invasion was observed in our case, which is minimal in parosteal osteosarcomas.

In the treatment of high-grade surface osteosarcoma, wide resection with an adequate surgical margin is required because of local recurrence. Marginal excision is strongly associated with increased risk of local recurrence [6, 10, 14].

The prognosis for high-grade surface osteosarcoma is worse than all other surface osteosarcomas. Patients have remained alive and disease-free a median of 10 years (range, 1.5–25.4 years) after diagnosis [15]. In our case the boy died 2 years after diagnosis, because of the recurrent disease.

Conclusion

"Exostosis-like" formations should be observed with caution.

The plain film presented aggressive development of this type of surface osteosarcoma. This case evidently shows the immense importance of timely diagnosis that is essential for adequate treatment.

CT is an excellent method for depicting the tumour localization and tumour characteristics, such as: type of tumour matrix mineralization, soft tissue involvement, absence of lucent zones between the tumour and underlying cortex, evaluation of the cortex and intramedullary involvement, which are important parameters for predicting high-grade malignancy.

CT gave information on all these parameters, which were confirmed histologically.

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Резиме

ПОВРШЕН ОСТЕОСАРКОМ СО ВИСОК СТЕПЕН НА МАЛИГНИТЕТ – ХОНДРОБЛАСТЕН ТИП

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Презентираме случај на 8-годишно момче со болка во дисталниот дел на десната надколеница, која се појавила по повреда со топка, без крвен подлив или оток. Рендгенографијата прикажува егзостоза на дистален крај на десен фемур. Во друга институција формацијата, личела на егзостоза, била хируршки отстранета и хистолошкиот наод бил агресивен тип на хондробластом.

Неколку месеци подоцна, на местото на хируршката интервенција се појавила значително зголемена болна солидна маса. Радиографијата прикажува голема дензна маса на ниво на дисталната феморална метадијафиза со тумор матрикс минерализација и Кодманов знак. Компјутеризираната томографија прикажува голема мекоткивна маса која потекнува од површината на кортексот со знаци за висок степен на малигнитет. КТ е вредна метода во предвидување на висок степен на малигнитет. Направена била и артериографија.

По оперативен третман – сек. Кампаначи, хистологијата покажала површен остеосарком со висок степен на малигнитет – хондробластен тип. Пред и постоперативна хемотерапија била направена. Момчето умрело две години по дијагнозата, поради повторување на болеста.

Површен остеосарком со висок степен на малигнитет - хондробластен тип има најлоша прогноза споредено со другите површински остеосаркоми. Поради тоа рано поставената дијагноза е мошне важна.

Клучни зборови: површен остеосарком со висок степен на малигнитет, компјутеризирана томографија, дијагноза.

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