CASE REPORT

A fatal case of pure metaphyseal chondroblastoma

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SUMMARY

The chondroblastoma (CB) is a rare cartilaginous tumour; it represents less than 1% of all bone tumours. It is mostly localised at the level of the epiphysis of long bones. We report a fatal case of pure metaphyseal CB of the tibia in a 9-year-old boy whose pulmonary metastases developed soon after operative therapy of the primary tumour.

BACKGROUND

Chondroblastoma (CB) is a rare benign chondroid bone tumour, which occurs in young patients. It is the most common primary epiphyseal tumour in children. CB typically involves the epiphysis or epiphysis-equivalent apophysis of a long tubular bone.¹ Although the tumour may secondarily involve the metaphysis, reports of primary metaphyseal or diaphyseal CB without any epiphyseal or apophyseal involvement are exceptionally rare and frequently present as a diagnostic dilemma. Here we present a case of pure metaphyseal CB in a 9-year-old boy. Two months after surgery multiple lung metastases appeared. The purpose of this case report is not only to explain this uncommon site of origin (pure metaphyseal) but also to emphasise that patients with CB may have developed metastasis soon after presentation.

CASE PRESENTATION

The patient was a 9-year-old boy with a symptom of left leg pain. He gave no history of recent trauma and his parents denied recent febrile illnesses. On admission he was afebrile. Pain had increased markedly during the 1 month prior to presentation to our clinic. Pain was insidious in onset and gradually worsened causing difficulty in walking and was relieved by rest and analgesics. Pain was dull, aching in character and there was no radiation. On physical examination tenderness was present on deep palpation, but there was no local rise of temperature. His gait was notably antalgic. There were no distal neurovascular deficits. No appreciable lymphadenopathy was noted. There was no soft tissue involvement.

INVESTIGATIONS

Laboratory studies revealed normal serum chemistries and complete blood count and differential. Purified protein derivative test was negative. A plain radiograph of the left leg showed a well-defined lytic lesion in the metaphysis of tibia. It had narrow transitional zone without calcification or sclerosis. Periosteal reaction on the medial aspect of tibia was noted (figure 1). A MRI study (coronal T1-weighted image) revealed isosignal area in the

left metaphysis of tibia (figure 2). Short TI inversion recovery (STIR) coronal image showed a hypersignal intensity lesion with periosteal reaction and soft tissue oedema on the medial aspect of tibia (figure 3).

The imaging findings were thought to be most consistent with an osteosarcoma involving the left tibia. A chest X ray did not show any abnormalities. An incisional biopsy of the lesion revealed a monomorphous neoplastic proliferation of polygonal mononuclear cells with well-defined cytoplasmic borders and bright eosinophilic cytoplasm suggestive of chondroblasts, intermingled with scattered osteoclast-like giant cells. A fine network of pericellular calcification ('chicken-wire' pattern) was noted (figure 4). In addition mitotic activity and small foci of necrosis were also observed. The mononuclear cells were focally immunoreactive to S-100 protein but diffusely were immunoreactive to vimentin (figures 5 and 6). A diagnosis of chondroblastoma was confirmed.

DIFFERENTIAL DIAGNOSIS

Differential diagnosis involves giant cell tumour, chondromyxoid fibroma (CMF), chondrosarcoma, especially clear cell chondrosarcoma and eosinophilic granuloma (EG). We should say that giant cell tumour is almost exclusively seen in patients who are skeletally mature, whereas chondroblastoma tends to arise in skeletally immature patients. The presence of a sclerotic rim helps to differentiate chondroblastoma from giant cell tumour of the bone, which generally lacks a sclerotic border.² CMF occur in the metaphysis, whereas chondroblastomas typically occur in the epiphysis. In addition CMF are devoid of calcification and have a characteristic myxoid, pseudolobular pattern of organisation as well as more pleomorphic stellate cells.³ Clear cell chondrosarcomas also occur at the ends of long bones. However, the nuclei of the cells of clear cell chondrosarcoma are round and may show prominent nucleoli. Eosinophilic granuloma may appear in rare instances as a radiolucent epiphyseal lesion similar to chondroblastoma. However, a more heterogeneous collection of cells, including histiocytes, granulocytes and eosinophils, might help to differentiate it.⁴

TREATMENT

Thorough curettage of the lesion was performed. The bony defect was packed by bone grafting. Bone was harvested from iliac crest and was mixed with bone substitute to fill the cavity. Postoperatively, the leg was put on a splint for 2 weeks and then full-length plaster was applied for 1.5 months.

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Figure 1 A plain radiograph of the left leg showed a well-defined lytic lesion in the metaphysis of tibia. It had narrow transitional zone without calcification or sclerosis. Periosteal reaction on the medial aspect of tibia was noted.

OUTCOME AND FOLLOW-UP

Two months after surgery the patient was admitted with a sudden onset of shortness of breath. Laboratory studies disclosed a severe anaemia, with haemoglobin of 5 g/dL and a haematocrit of 15.2%. A chest CT showed multiple pulmonary metastases. It was therefore decided to administer one course of chemotherapy with doxorubicin, vincristin and cyclophosphamide. Despite this therapy, the patient showed no response, so chemotherapy was stopped. Two week later the patient's respiratory distress worsened and he was intubated. Two days later he died. The parents did not agree with autopsy.

DISCUSSION

In 1927, Kolodny⁵ was the first to describe a bone tumour presenting as a 'giant cell variant.' Ewing⁶ called it a 'calcifying giant cell tumour,' and later, in 1931, Codman⁷ termed it as 'epiphyseal chondromatous giant cell tumour.' The term benign 'chondroblastoma' was proposed by Jaffe and Lichtenstein⁸ in 1942 to describe this rare, benign and distinctive tumour composed of immature chondroblasts with a scant chondroid matrix. Although chondroblastoma affects people of all ages, a study of 104 cases by Bloem and Mulder⁹ revealed an average age of 16 years in tumours affecting long bones. Our patient

Figure 2 A MRI study (coronal T1-weighted image) showed isosignal area in the left metaphysis of tibia.

Figure 3 Short TI inversion recovery coronal image showing a hypersignal intensity lesion with periosteal reaction and soft tissue oedema on the medial aspect of tibia.

Figure 4 Monomorphous neoplastic proliferation of polygonal mononuclear cells with well-defined cytoplasmic borders. A fine network of pericellular calcification ('chicken-wire' pattern) was noted (H&E $\times 40$).

Figure 6 Tumour cells were strongly immunoreactive to vimentin.

was a 9-year-old boy. Patients usually present with symptom of pain and swelling, particularly if a pathological fracture is present. The presented case had leg pain. Most lesions (60%) involve the epiphysis and extend into the metaphysis, and a purely metaphyseal lesion is unlikely to be a chondroblastoma.¹⁰ However, there are situations where the tumour is located in non-physal locations.^{11–14} The presented case was an example. In one study the authors retrospectively reviewed 87 cases of chondroblastoma in patients with open physes at the time of diagnosis. They found out that the tumours were located in the epiphysis in 68% of the patients.¹⁵ Radiologically, chondroblastoma usually presents as a sharply demarcated oval or round lytic epiphysal defect surrounded by a rim of sclerotic bone. Cortical breakthrough is unusual. A scattered, stippled calcification appearance or a sparsely trabeculated pattern could be observed occasionally.¹⁶ In our case an X-Ray did not show calcification or sclerosis but periosteal reaction on the medial aspect of tibia was noted. MRI can help in the diagnosis and in the differentiation of a chondroblastoma. MRI is especially useful when plain radiographic findings are inconclusive.¹⁷ Immunohistochemically tumour cells coexpress vimentin and S100 protein.¹⁴ In our case tumour cells were focally positive for S100 protein. This finding coincided with the result of another study.¹⁸

Figure 5 Tumour cells were focally immunoreactive to S-100 protein.

From the available numbers, it seems that the metaphyseal and diaphyseal CB may not differ clinically and prognostically from typical epiphysal chondroblastomas. In contrast one study¹⁹ has been shown that non-epiphysal CBs were associated with a lower risk of recurrence. The reason may be related to this fact that non-epiphysal lesions are away from the epiphysis, so a more aggressive surgical approach can be taken. Therapeutic modalities for CBs have included curettage, en bloc resections and radiation therapy.²⁰ Our patient underwent thorough curettage of the lesion. The evolution of the chondroblastoma is mainly dominated by risk of local recurrence which varies between 5 and 38%.²¹ In 1942 Jaffe and Lichtenstein⁸ defined CB as benign tumour. However, in 1949 Copeland and Geschickter reported 25 cases of CB, among which 15 patients died within 9–18 months after the surgery and some cases even developed lung metastasis. Therefore, they proposed that CB is malignant.²² Later, Kyriakos *et al*²³ confirmed the existence of malignant CB, which was defined as CB, having histological characteristics of benign CB but with the ability to progress and metastasise aggressively. Histologically, most of these metastatic lesions are similar to conventional CB. Because of the lack of cellular criteria of malignancy it is impossible to predict the potential biological behaviour of CBs, in particular with respect to their ability to metastasise. However, the presence of tumour emboli in the primary lesion is highly suggestive of a subsequent development of metastatic disease.²⁴ In the presented case tumour emboli were not found but mitotic activity was considerable. It has been noted that the majority of metastases have only developed subsequent to some surgical therapy. In Kunze²⁴ study the period from the diagnosis of primary tumour to the detection of lung metastasis was average 8.4 years. It is suggested that iatrogenic factors contributing to the aggressiveness of this tumour, including breaching of cortex and accidental spillage at the time of curettage results in implantation of tumour tissue in soft tissues.²⁵ But there are a few reports that show unlike previously published examples of metastatic CB, these metastases developed before any operative manipulation of the primary tumour.²³

In our patient, the initial chest X-ray did not provide any information suggesting lung metastasis. However, considering that the metastatic nodules were very small, it was not surprising that these lesions could not be detected on chest radiography. A chest CT may be useful for detecting occult metastatic nodules in such patients. In addition PET scan is an exciting technology

and is often performed for the presence or absence of metastases²⁶ but unfortunately this method is not available in Iran. The five unique points about our case are: (1) the primary tumour site was unusual. (2) There was not sclerotic rim on X-Ray. (3) There were considerable mitotic activity. (4) The mononuclear cells were focally immunoreactive to S-100 protein. (5) Metastases developed very soon after operative manipulation of the primary tumour.

In conclusion CB is a rare benign chondroid bone tumour. Occasionally, however, CB develops more aggressively, with invasion of joint spaces, adjacent bones and even metastases. Metastases from CB are extremely rare soon after diagnosis. Despite its rarity, the possibility of metastasis as an accompaniment of primary CB merits consideration when this neoplasm is initially diagnosed. A preoperative plain chest X-ray is justified on all patients newly diagnosed with CB, because metastasis involves the lung most commonly and may be curable by early resection. CT imaging of the chest 1–2 months after primary tumour resection is also advisable to screen for metastases.

Learning points

- ▶ Although chondroblastoma (CB) may secondarily involve the metaphysis, reports of primary metaphyseal or diaphyseal CB without any epiphyseal or apophyseal involvement are exceptionally rare.
- ▶ From the available numbers, it seems that the metaphyseal and diaphyseal CB may not differ clinically and prognostically from typical epiphyseal CB.
- ▶ Occasionally, CB develops more aggressively and even metastases.
- ▶ Metastases from CB are extremely rare soon after diagnosis.
- ▶ CT imaging of the chest 1–2 months after primary tumour resection is also advisable to screen for metastases.

Competing interests None.

Patient consent Obtained.

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