CASE REPORT

Calcaneal osteosarcoma: a rare cause of heel pain in the paediatric population

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SUMMARY

Osteosarcoma is the most common primary non-haemopoietic malignant bone tumour in children and adolescents. However, it rarely occurs in the calcaneus with only a few case reports in the literature. We report a case of a 14-year-old boy with calcaneal osteosarcoma, who presented with heel pain followed by swelling. The pain was initially thought to be related to a benign process and treated with analgesics, delaying the diagnosis. We discuss the clinical presentation, the differential diagnosis, multi-imaging and pathological findings of a calcaneal osteosarcoma, its clinical outcome and the importance of early diagnosis to improve outcome.

BACKGROUND

Osteosarcoma is the most common non-haemopoietic primary malignant neoplasm of the bone in children and adolescents. The calcaneal location of osteosarcoma is very rare, representing less than 1% of all osteosarcoma, and is rarely reported in the literature particularly in young patients. We report an unusual case of a 14-year-old patient presenting with heel pain followed by a rapidly growing heel mass. Although primary malignancy of the foot is uncommon, we emphasise the importance of maintaining high suspicion for malignancy in paediatric patients presenting with similar symptoms, since early diagnosis is crucial in improving outcome and prognosis.

CASE PRESENTATION

A previously healthy 14-year-old boy of non-consanguineous parents and no known genetic abnormalities initially presented to his paediatrician with a 1-month history of heel pain and limping and no reported major trauma or fever. The pain was thought to be related to sport activity and responded initially to analgesics. However, over the next few weeks, the pain increased and a heel swelling appeared. At that time, an orthopaedic surgeon was consulted. On physical examination, a stiff, immobile and tender mass located in the posteroinferior aspect of the left heel was palpated, with no overlying skin changes.

INVESTIGATIONS

Radiographs (figure 1A) and a CT scan (figure 1B) of the left ankle and foot revealed a densely sclerotic mass with aggressive periosteal reaction and sunburst appearance involving the calcaneus, with extension into the adjacent soft tissues. The imaging features of the mass were consistent with a malignant bone-forming tumour, mainly

osteosarcoma. MRI (figure 1C–F) was performed to evaluate the disease extent showing a heterogeneous mass involving almost the entire calcaneus with areas of low signal intensity due to sclerotic bone formation. Following contrast administration (figure 1F), there was a heterogeneous enhancement of the tumour, with areas showing no enhancement related to the densely sclerotic bone component. The tumour reached the articular surface with the talus and cuboid as well as the tip of the lateral malleolus. Superiorly, the tumour reached the posterior capsule of the ankle joint, associated with ankle joint effusion. There was also a reactive bone marrow oedema within the navicular, cuboid and talus bones. There was infiltration and oedema of the surrounding soft tissues. A CT-guided percutaneous biopsy of the lesion was consistent with osteosarcoma, osteoblastic type (figure 2). Bone and CT scans of the chest, abdomen and pelvis showed no evidence of metastatic disease at the time of diagnosis.

TREATMENT

The patient was started on chemotherapy (cisplatin, methotrexate and doxorubicin). Following chemotherapy, there was clinical improvement in the size of the heel mass and in the pain. Below-knee amputation was performed after the completion of three cycles of chemotherapy. Macroscopically, the tumour measured 7 × 6 × 5.5 cm in its greatest dimensions. It consisted of a tan-white yellow heterogeneous, smooth and firm mass, located in the body of the calcaneus, reaching the articular surfaces with the talus and cuboid bones. Microscopically, the bone was substituted by the 7 cm osteoblastic tumour invading the soft tissues. There was 50% overall necrosis within the mass, with residual osteoblastic osteosarcoma, which is considered as a poor response to chemotherapy (<90% necrosis).

OUTCOME AND FOLLOW-UP

Chemotherapy was continued postoperatively, with cisplatin, doxorubicin and methotrexate, for a planned total of 32 weeks of treatment. However, 3 months after the surgery, the patient started complaining of pain at the distal aspect of the amputated limb. Several radiographs of the leg were normal and the pain was attributed to phantom limb pain. However, owing to the patient's increasing pain, MRI of the affected limb was performed and it showed diffuse tumour infiltration of the left tibia and femur (figure 3A,B). Simultaneously, the patient developed lung metastases, associated with malignant pleural effusion, resulting in

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Figure 1 Lateral radiograph of the left ankle (A) showing a large sclerotic lesion involving the calcaneal bone. Sagittal CT scan of the left ankle and (B) showing the large destructive heterogeneous tumour of the calcaneus is seen, with significant dense bone formation. Sagittal T1-weighted MRI (C) and coronal STIR (short tau inversion recovery) (D) of the left ankle showing a predominantly low signal intensity tumour reaching the articular surfaces with infiltration of the surrounding soft tissues. Coronal T1-weighted images with fat saturation before (E) and after (F) gadolinium administration reveal heterogeneous enhancement of the tumour.

dyspnoea and requiring chest tube insertion for symptom relief (figure 3C,D). The poor outcome was discussed with the family, chemotherapy was stopped and the patient was shifted to palliative care. He passed away 11 months after the initial presentation from diffuse metastatic disease to the lungs and bones, with superimposed pulmonary infection.

DISCUSSION

Osteosarcoma is the most common non-haemopoietic primary malignant neoplasm of the bone in children and adolescents, although it accounts for less than 1% of all cancers diagnosed in the USA.¹ It produces an osteoid matrix and variable amounts of cartilage matrix and fibrous tissue.² A majority of

Figure 2 Histopathology from a percutaneous biopsy of the tumour (A–B) revealing malignant sarcomatous cells (arrowhead) arranged in cords and nests with associated osteoid deposition (arrow) (mag. $\times 100$ and $\times 200$). Pathology images after amputation postchemotherapy (C–D) show residual osteoblastic osteosarcoma characterised by nests of malignant sarcomatous cells encompassed by abundant osteoid deposits. Note the mitotic figure in the centre of the field (arrow) (mag. $\times 100$ and $\times 200\times$).

Figure 3 Coronal T1-weighted MRIs of the left leg stump (A) and left thigh (B) showing diffuse heterogeneous abnormal low signal intensity involving the entire length of the tibial stump. Multiple low signal rounded lesions involve the distal femoral epiphysis and metaphysis (B). CT scan of the chest (C) shows a calcified metastatic nodule in the right lower lobe (arrow). A follow-up CT scan of the chest (D) shows a few metastatic lung nodules (arrows), one of which is calcified (left arrow), along with pneumonic infiltrates, atelectatic changes and bilateral pleural effusions.

osteosarcoma cases occur in adolescence, with a male predominance at all ages.³ It is classified by WHO into conventional (osteoblastic, chondroblastic and fibroblastic), telangiectatic, small cell, low-grade central, secondary, periosteal, parosteal and high-grade surfaces, with the conventional type being the most common.^{2 3} The therapeutic options and prognoses for different types of osteosarcoma differ from each other. Correct diagnosis and analysis of extent are essential and require accurate evaluation by imaging, in particular MRI.

Osteosarcoma most commonly arises in the metaphyses of long bones of the appendicular skeleton; particularly the distal femur, proximal tibia and proximal humerus.³ However, epiphyseal and diaphyseal involvement have been described. Generally, osteosarcoma can involve any part of any bone.^{4 5} Involvement of the pelvis, fibula, facial bones and spine is unusual. The clavicle, ribs, skull, scapula and small bones of the feet and hand are rarely affected. Osteosarcoma of the foot bones is the least common, accounting for only 1% of all cases,⁶ with 75% arising from the calcaneus or metatarsal bones.⁷ Osteosarcomas of the foot have been reported mostly in adults,⁷ and only a few cases of calcaneal osteosarcoma have been reported in children and adolescents, such as our patient. Osteosarcomas of the foot are more likely to be low grade and have higher reported survival rates,⁷ in contrast to the case we present. In addition, it has reported association with Werner syndrome, hereditary retinoblastoma, Li-Fraumeni syndrome and Rothmund-Thompson syndrome,⁸ which our patient did not have.

The symptoms are non-specific, often delaying the diagnosis. Pain on walking and foot swelling are the most commonly reported presenting symptoms. The most common conditions that may present as heel pain and swelling in a paediatric patient include calcaneal apophysitis and retained foreign body. In rare instances, fractures and tumours of the calcaneal bone may be encountered. Benign tumours are known to affect the paediatric heel and are more common than malignant tumours. These include unicameral bone cyst, aneurysmal bone cyst (ABC), giant cell tumour, osteoid osteoma, osteoblastoma and non-ossifying fibroma.⁹ Osteosarcoma is the most common

malignant tumour of the calcaneus, followed by Ewing sarcoma and chondrosarcoma.⁹ The pain and swelling caused by osteosarcoma may improve initially with conservative treatment, leading to misdiagnosis and delay in treatment. The delay between the onset of symptoms and the diagnosis ranged from 1 to 24 months in a review of 52 cases of calcaneal osteosarcoma performed by Choong *et al*¹⁰ at the Mayo Clinic. Therefore, bone malignancy of the foot should be taken into consideration when a paediatric patient presents with foot pain and swelling, to reach an early accurate diagnosis and improve the outcome, precluding delay in the diagnosis. Hence, the primary evaluation of such a presentation should include radiographic assessment.

The reported subtypes of calcaneum osteosarcomas include the conventional osteosarcoma, low-grade central osteosarcoma, telangiectatic osteosarcoma, periosteal osteosarcoma and small-cell osteosarcoma.¹¹ Osteoblastic type is the most common and is usually densely sclerotic with ill-defined borders and extra-compartmental bone production. Mixed tumour presents as an osteolytic expansile lesion with sclerotic rim and internal trabeculations, along with cortical destruction and extraosseous extension.⁷ An intramedullary variant of osteosarcoma of the foot may have the radiographic features of a lytic lesion with a thin sclerotic border and no cortical destruction. This variant constitutes approximately 5% of all osteosarcomas and is associated with better survival rates.¹² Telangiectatic osteosarcomas of the calcaneal bone may radiologically present as an expanding lytic lesion with fluid-filled cystic spaces, very similar to ABC.⁷ Owing to the lack of fascial barriers and thinner cortices, osteosarcoma of the foot usually presents with the involvement of the surrounding compartments, and MRI is essential in assessing the extension. Our patient's radiographs show classic features of osteoblastic osteosarcoma and MRI confirmed the presence of surrounding soft tissue infiltration, which allowed the accurate assessment of the extension, thus guiding surgical intervention.

The treatment of bone sarcomas varies depending on the stage. The standard treatment of patients with conventional

osteosarcoma consists of a combination of chemotherapy and surgery. Usually, limb-salvage surgery is possible in most bone sarcomas of the long bones. However, osteosarcoma of the foot usually is not amenable to limb-salvage surgery because of the poor compartmentalisation of the tumour in the foot and the subsequent extension to the surrounding foot compartments,¹⁰ such as in our patient. Therefore, below-knee amputation is usually the adopted form of treatment with satisfactory results in most cases. Limb-salvage surgery in calcaneal osteosarcoma has been reported for the first time in 1998,¹² with only a few additional reports in the literature.¹¹ It involves the prosthetic replacement of the calcaneus and results in successful eradication of the tumour with no local recurrence of the disease after several years of follow-up, while allowing for an optimal, functional use of the limb. However, limb-sparing surgery can be offered only to patients with intramedullary tumours that show

a good response to neoadjuvant chemotherapy.¹² Before the use of neoadjuvant chemotherapy, the 2-year survival rate for patients with osteosarcoma was 15%. Survival after preoperative and postoperative chemotherapy currently increased to reach 60–70%, depending on the response of the tumour to neoadjuvant chemotherapy,¹² which is graded as good if 90% or more of chemotherapy-induced tumour necrosis is observed.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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Learning points

- Primary malignancy of the foot is uncommon.
- Heel pain and swelling in a paediatric patient are usually associated with benign conditions, including calcaneal apophysitis, retained foreign body and fractures.
- Benign tumours are known to affect the paediatric heel and are more common than malignant tumours.
- Paediatricians and radiologists should be aware that heel pain is not always benign in nature, avoiding a delay in diagnosis and allowing early treatment and good outcome.
- When a paediatric patient presents with long-standing heel pain, malignancy should be included in the differential diagnosis and good physical examination followed by appropriate imaging modality is essential in the evaluation.

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