Primary parosteal osteosarcoma of the rib

Abhishek Shah\textsuperscript{a}, Hongxi Ma\textsuperscript{a}, Xiaoyan Sun\textsuperscript{a} and Dianbo Cao\textsuperscript{a,*}

\textsuperscript{a} Department of Radiology, The First Hospital of Jilin University, Changchun, China
\textsuperscript{b} Department of Pathology, The First Hospital of Jilin University, Changchun, China

* Corresponding author. Department of Radiology, The First Hospital of Jilin University, Changchun, Jilin Province 130021, China. Tel: +86-431-88782911; fax: +86-431-85654528; e-mail: caodianbo@126.com (D. Cao).

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Abstract

Osteosarcoma is a common primary malignant tumour of bones that produces an osteoid matrix. Parosteal osteosarcoma is an uncommon neoplasm and principally affects the long bones, especially in the distal femur, proximal tibia and proximal humerus. Rarely, the tumour may arise in a flat bone, and rib involvement is very infrequent. An unusual case of primary parosteal osteosarcoma of the rib in an asymptomatic 28-year old female is presented here. A chest X-ray film suggested an intrapulmonary homogeneous mass, while a computed tomography scan revealed a broad-based calcified mass attached to the inner cortex of the right fourth anterior rib. The patient underwent a wide excision of the tumour together with adjacent intercostal muscles and chest wall reconstruction. Postoperative histopathological outcome was consistent with primary parosteal osteosarcoma.

Keywords: Rib • Parosteal osteosarcoma • Computed tomography • Treatment

CASE REPORT

A 28-year old woman was admitted to our hospital because of the detection of an intrapulmonary mass. A physical examination revealed no abnormality. Haematological evaluation was within the normal limit. There was no past history of radiotherapy, chemotherapy or Paget\textsuperscript{b} disease. On a chest X-ray, a lobulated, intrapulmonary homogeneous mass adjacent to the right 4–6th ribs was detected. No destruction of the underlying rib or adjacent ribs was noted. A computed tomography (CT) scan revealed a broad-based mass measuring 3.6 cm × 4.0 cm attached to the inner cortex of the right fourth anterior rib (Fig. 1). The mass, outpouching into the right lung, was lobulated, associated with scattered calcification adjoining the inner margin of the rib. There was no mediastinal lymphadenopathy or any other lesions over the body except for the right chest on CT. The fourth rib tumour was extensively removed together with adjacent intercostal muscles, and the chest wall was reconstructed. In gross appearance, the tumour was greyish white in colour, measured 5 cm × 3.8 cm × 3 cm and had a lobulated surface (Fig. 2). The resected specimen was confirmed to be parosteal osteosarcoma on histopathological analysis. The patient refused chemotherapy and was well at 6 months of follow-up.

DISCUSSION

Osteosarcoma is a malignant tumour of connective tissue that produces an osteoid matrix and variable amounts of cartilage matrix and fibrous tissue. Parosteal osteosarcoma accounts for ~3–4% of all osteosarcomas and 1.6% of primary bone malignancy [1]. The peak incidence of parosteal osteosarcoma is in the third decade of life. It usually involves the posterior aspect of the distal femur (in 60% of cases), proximal humerus or theibia. Histologically, the lesions are frequently well differentiated, with a well-formed osteoid within a spindle cell stroma. Medullary involvement occurs in <25% of cases. Osteosarcoma occurring as a primary tumour in the rib is rare and there are only a few case reports existing in the literature as a primary parosteal osteosarcoma of the ribs [2, 3]. The natural history of this disease may extend to five years or longer before metastasis occurs. Unlike conventional osteosarcoma, these tumours are commonly painless, leading to a delay in presentation and a large tumour size at diagnosis [4]. On the contrary, Osawa et al. [5] have reported a case with haemorrhagic shock due to the rupture of osteosarcoma of the ribs in the thoracic cavity, which may be due to the invasion of the tumour in the intercostal vessels or due to the rupture of tumour itself. So, when a case of haemorrhagic pleural effusion is encountered, the possibility of osteosarcoma should be kept in mind.

Radiologically, on a chest X-ray film, it is difficult to differentiate whether the lesion is from the ribs, lung or pleura. Occasionally, it can help us to know about the presence of a mass lesion which is densely calcified. In our case, a chest X-ray showed a lobulated mass of homogeneous opacity in the right lung, highly suspicious of a pleural or lung lesion. Sabloff et al. [6] also presented a similar case of extraskeletal osteosarcoma of the pleura where it was difficult to identify the originating site on the basis of chest X-ray findings. In such a circumstance, a CT scan is the best choice as it can clearly reveal the origin, location, component and extent of the tumour, especially in the complicated anatomic location. Parosteal osteosarcoma often appears as dense, calcified masses, but in our case subtle calcification was found on the CT images. A large lesion may encircle the
In addition to CT scans, MRI can be a supportive tool to evaluate the extension of tumour towards soft tissue, other adjacent structures and medullary extension, as it is more clearly visualized by MRI [2]. Otherwise, MRI does not possess any extra significance in comparison with CT scans in diagnosis and cost effectiveness. While making the diagnosis radiologically, differential diagnoses of calcified masses should be considered, such as metastatic tumour, chondrosarcoma, myositis ossificans and pleural osteosarcoma.

The treatment of parosteal osteosarcomas generally consists of local excision with a wide margin [7]. Exactly what size of margin is optimal remains unresolved in the management of this malignancy. In >80% of patients, local recurrence appears in the absence of a wide resection. So, adjuvant chemotherapy or radiotherapy should be considered for those patients who undergo a palliative operation.

In conclusion, this case highlights the fact that rib parosteal osteosarcoma, although rare, should be considered in the differential diagnosis when a contained-calcified mass centred in a rib is detected on a CT scan. Imaging studies, especially CT scans, play a crucial role in the diagnosis of osteosarcoma because the diagnosis is based on a combination of histopathological and imaging features.

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**REFERENCES**