A RARE PRESENTATION OF LUNG METASTASIS IN OSTEOSARCOMA: A RAPIDLY ENLARGING LUNG LESION

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Abstract

Osteosarcoma is the most frequent primary malignant mesenchymal bone tumour in children and adolescents. Although the lung is the most common site of its metastasis, to the best of our knowledge, it is infrequent to have hypervascular pulmonary metastasis, particularly in the post-operative period. Herein, we report a case of a 15-year-old boy who presented with a rapidly enlarging lung mass on a background of osteosarcoma of left proximal tibia. The progressively enlarging right lung mass was detected as an opacity on a chest radiograph, three months post-surgical resection of the osteosarcoma. Computed tomography of thorax revealed a contrast-enhancing hypervascular right lung mass. This was complicated with intra-lesional haemorrhage post-biopsy. Histopathological examination (HPE) confirmed metastatic osteosarcoma. We discuss the rarity of this occurrence and its imaging findings.

Keywords: Osteosarcoma, Pulmonary Metastasis Osteosarcoma, Hypervascular Pulmonary Metastasis, Haemorrhagic Pulmonary Metastasis

Introduction

Osteosarcoma is defined by the presence of malignant mesenchymal cells producing osteoid or immature bone (1). This high-grade tumour is the most commonly occurring primary bone sarcoma comprising 60% of all bone cancers in individuals younger than 20 years of age (2, 3). The most common sites of osteosarcoma are the metaphysis of long bones, whereby 42% found in the femur (75% distal), 19% in the tibia (80 % proximal) and 10% in the humerus (90% proximal) (4).

The majority of osteosarcoma metastases are spread via the haematogenous route, and the lung is its most common site, which accounts for 98% of cases (5). There are approximately 20-40% of osteosarcoma patients who present with pulmonary metastasis at the time of diagnosis, and they usually appear as calcified nodules (6, 7). Osteosarcoma with lung metastasis is very common; however, osteosarcoma with hypervascular pulmonary metastasis is an extremely rare presentation.

Case report

We report a 15-year-old boy who was diagnosed with osteosarcoma of the left tibia and had undergone above-knee amputation (AKA) one month after failure of response to neoadjuvant chemotherapy. He was planned for post-operative chemotherapy; however, due to the left AKA stump infection that required multiple wound debridements, his chemotherapy was postponed.

During the oncology clinic follow up at three months post-AKA, he complained of right pleuritic chest pain for three days, associated with cough and whitish sputum. The serial chest radiographs (CXR) post-surgery showed no lung lesion (Figure 1A). However, the CXR acquired on the clinic day revealed a new lesion at the right middle zone (Figure 1B). The subsequent CXR performed after nine days demonstrated a substantial enlargement of the right lung lesion (Figure 1C). Contrast-enhanced CT thorax was performed four days after the last CXR and showed a significantly enhancing right upper lobe mass with intralesional and peripheral vascularity, associated

with a massive right pleural effusion (Figure 2). Following the CT finding, an ultrasound-guided tru-cut biopsy of the mass was carried out. Unfortunately, the procedure was complicated with haemorrhage whereby the patient developed shortness of breath and bleeding at the puncture site immediately after the biopsy. Although the pre-biopsy haemoglobin (Hb) level has gradually dropped from 13.3 g/dl to 10.1 g/dl over a week, fortunately, the post biopsy level remained stable. The repeat CXR demonstrated complete right hemithorax opacity with significant contralateral mediastinal shift (Figure 1D). The subsequent CT thorax revealed enlarging right lung mass with evidence of active bleeding and massive right pleural effusion (Figure 3).

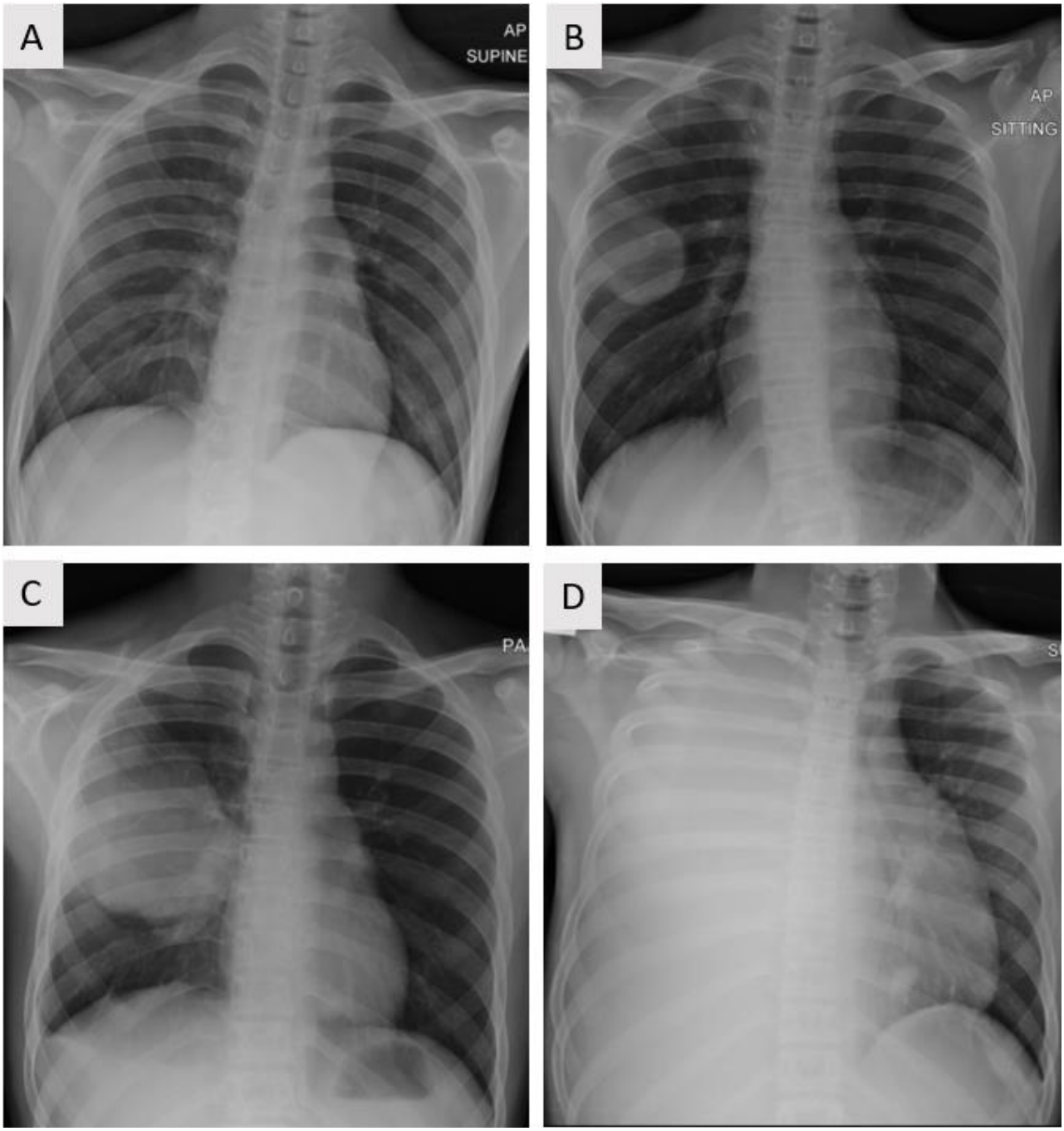

Figure 1: Serial chest radiographs in anteroposterior (AP) view show normal lung fields at the initial stage (A). A well-defined homogenous opacity is identified at the right middle zone three months later (B) which has progressively increased in size within nine days (C). Post ultrasound-guided biopsy reveals complete opacification of the right hemithorax (D)

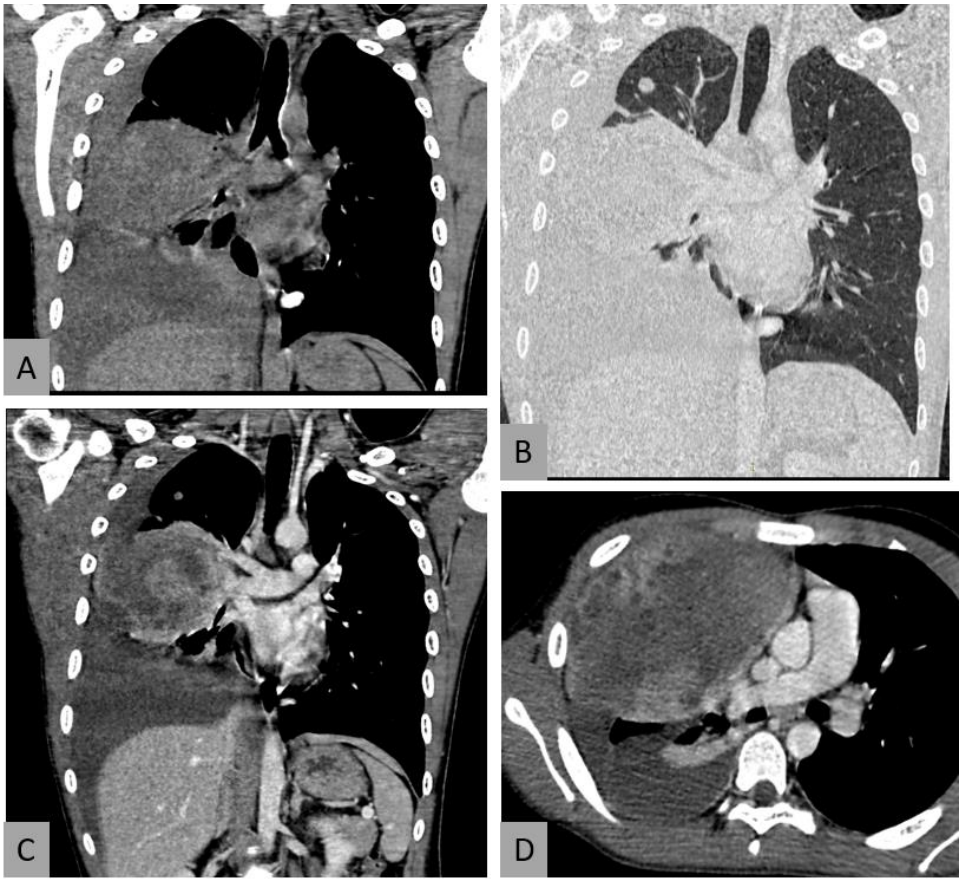

Figure 2: (A) Non-contrast CT thorax in coronal view shows right lung mass arising from the upper lobe with presence of right pleural effusion. (B) Lung window setting also reveals another small lung nodule at the same lobe. In the coronal (C) and axial (D) views of the post-contrast study, the mass appears large and heterogeneously enhanced with evidence of intralesional and peripheral vascularity

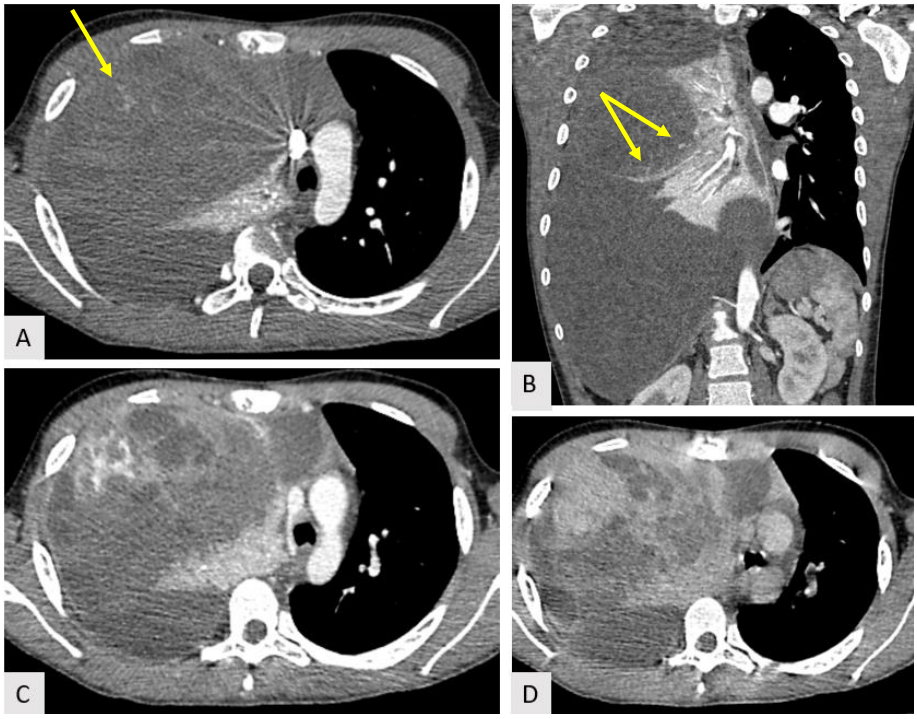

Figure 3: The post-biopsy contrast-enhanced CT thorax in axial (A) and coronal (B) views demonstrate enlarging right upper lobe mass with hyperdensities seen within it and at the perivascular region (arrows) during arterial phase. Massive right pleural effusion is also noted. The late arterial-early portal venous phase (C) shows increasing hypderdensities within the mass with pooling of contrast on delay phase (D) suggestive of active right tumoral haemorrhage

He subsequently underwent a thoracotomy for tumour resection and the extensive peritumoral haematoma (amounting to approximately three litres) was evacuated. Intraoperative finding also revealed another small hard nodule (1.0 x 0.8 x 0.2 cm) at the base of right upper lobe. The bleeding at the biopsy site was secured and repaired. The histopathological examination confirmed osteoid forming tumour cells consistent with metastatic osteosarcoma. The patient recovered post-surgery and was discharged two weeks later with clinic follow up.

Discussion

One of the most frequent types of primary malignancy of the bone is osteosarcoma, which accounts for approximately 3% of all paediatric malignancy and has the second-highest mortality rate among paediatric cancers (1). At presentation, 80% of the patients with osteosarcoma have localized disease, and 10% have distant metastasis (8), with the lung being its most common site (9). Distant metastasis usually occurs within 18-24 months of the primary disease, with 8% of patients with osteosarcoma already have metastatic pulmonary lesions which may not be apparent on initial the CXR (10). Therefore, close follow-up with imaging is mandatory.

CT scan is superior to the CXR in the identification of metastatic pulmonary nodules. The typical appearance of the pulmonary metastasis in osteosarcoma are calcified lung nodules and lymph nodes which are sometimes associated with spontaneous pneumothorax (11, 12). The metastatic pulmonary nodules are usually multiple, well-defined, peripherally located and are of varying sizes. Some nodules may have cavitations while the hypervascular metastatic nodules may complicate with peritumoral haemorrhage, depending on the histology of the primary tumour. In daily practice, about 14% of metastatic nodules may have atypical features on CT with unusual presentation (13).

In hypervascular metastatic pulmonary nodules, the CT usually shows abundant intra or peritumoral blood vessels (14). If peritumoral haemorrhage is suspected, the lung nodules will have a halo of ground-glass opacity at its surrounding giving a CT halo sign. However, the halo sign is not a specific feature for peritumoral haemorrhage, since it is also present in other diseases such as invasive aspergillosis or candidiasis (15). The bleeding tumour can also be appreciated through enhancing areas surrounding the feeding vessels suggestive of contrast extravasation or leakage. However, these findings can be very subtle and quite challenging to appreciate, especially in the presence of large and heterogeneous enhancing mass**.** Therefore, imaging findings must always be confirmed with a histological examination and biochemical correlation to establish a definitive diagnosis.

Hypervascular pulmonary metastatic nodules are not commonly seen in osteosarcoma. Few case reports have been published on pulmonary metastasis in extremity osteosarcoma. However, our patient is the first case with such an unusual clinical course. Despite its rarity, the new and rapidly growing right lung mass with intralesional and peripheral vascularity as seen in our case has raised the suspicion of hypervascular pulmonary metastasis. The imaging appearance of progressively enlarging lung mass with worsening pleural effusion in several-day duration, associated with gradual Hb drop and intraoperative evidence of massive peritumoral haemorrhage hinted that slow haemorrhage has occurred in our patient. A biopsy is not a procedure of choice in this kind of presentation. However, due to the unawareness of the gradual Hb drop and the lung mass was large abutting the pleura, ultrasound guided thru-cut biopsy was decided instead of CT guided procedure in order to avoid repeated exposure to ionising radiation. In general, the biopsy for such condition should be performed with high precautions and close monitoring since hypervascular metastatic pulmonary nodules bleed easily and may potentially lead to fatal complications.

Pulmonary metastasis usually indicates poor prognosis among osteosarcoma patients. Management for osteosarcoma with pulmonary metastasis includes combination of chemotherapy (neoadjuvant and adjuvant regimens), surgical removal of primary tumour and pulmonary metastasectomy and radiation therapy (16). Proper imaging and adequate management at initial presentation may give an excellent clinical result and improve patient survival.

Conclusion

Osteosarcoma presenting with rapidly enlarging hypervascular pulmonary metastasis is an extremely rare case, and it may occur even post resection of the primary tumour. Regular follow up with imaging is required as part of the surveillance in order to detect new lung lesion, especially in the presence of new chest symptoms. Familiarity with characteristic CT feature of hypervascular lung metastasis and its complications is of paramount importance since proper management will reduce the risk of mortality from haemorrhage.

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Conflict of interests

The authors declare that they have no conflicts of interest.

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Informed consent

Verbal informed consent was obtained from the patient's next of kin before the manuscript was prepared.

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