Primary Chondroblastic Osteogenic Sarcoma of the Rib in an Adult

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Abstract
Primary osteosarcoma of rib, especially in adults, is extremely rare in literature. We present an unusual case of a 24-year-old female with an osteolytic, infiltrative mass originating from the third rib. It showed mediastinal and paraspinal extension with consequent pleural effusion and compressive myelopathy. It was mistaken for neurofibroma and chondrosarcoma on imaging studies. Microscopy showed coexistent osteoid and chondroid elements consistent with chondroblastic osteogenic sarcoma. Thus, histomorphology remains the diagnostic gold standard.

Introduction
Primary osteogenic sarcoma (OS) shows a predilection for the metaphysis of long bones, while only 1-2% cases involve the flat bones of ribs. Pediatric population is affected more frequently than adults. We present a rare case of adult-onset chondroblastic osteogenic sarcoma of the rib presenting as unilateral pleural effusion and compressive myelopathy due to mediastinal and paraspinal infiltration respectively. Thus, unusual location, unusual age of onset and unusual clinical presentations should be borne in mind to prevent misdiagnosis.

Case History
A 24-year-old female presented with bilateral lower limb weakness for two months. She did not have fever, breathlessness or weight loss. She did not have any history of irradiation, Paget’s disease or chemotherapy. Her vital parameters were stable while complete hemogram, liver and renal function tests were within normal limits. Chest X-ray showed a calcified mass extending from left supraclavicular region to the posterior mediastinum with associated left sided pleural effusion. Magnetic Resonance Imaging (MRI) showed compressive myelopathy of T1-T5 region due to paraspinal expansion of the mass (Figures 1a and 1b). The MRI diagnosis was giant neurofibroma.

Computed Tomography (CT) of thorax showed a 10.7 cm x 10 cm x 9.5 cm, well-defined, lobulated, densely calcified mass arising from the posterior surface of the third rib. The mass was osteolytic and heterogeneously enhancing. The great vessels, lungs and abdominal organs were uninvolved by the tumor. The CT scan diagnosis was primary rib chondrosarcoma.

Partial resection of the tumor was done. Histomorphology showed an osteoid-forming malignant tumor coexistent with a prominent chondroid element, thus confirming chondroblastic osteogenic sarcoma (Figures 2 and 3). The patient was treated with adjuvant radiotherapy and chemotherapy. She is disease-free and asymptomatic for fourteen months after therapy.

Discussion
Primary osteogenic sarcoma (OS) shows a predilection for the metaphysis of long bones, these being the sites of greatest bone growth. Approximately 10% cases are situated in flat bones, especially of the pelvis while only 1-2% involve the thoracic bones such as ribs, sternum and clavicle. Osteogenic sarcoma of ribs is extremely rare. Children and adolescents are affected more frequently than adults. Rib OS can be clinically asymptomatic or may present as a thoracic mass with resultant dyspnea, as an exophytic mass of the chest wall, as an intrapulmonary mass or as hemorrhagic pleural effusion. In our patient, large size of the rib tumor along with mediastinal and paraspinal invasion resulted in unilateral pleural effusion along with compressive myelopathy.

CT scan is useful to identify the location, origin and extent of osteoid-forming tumors while MRI is useful to evaluate extension into...
underlying medulla or adjacent soft tissues. On CT scan, rib OS usually present as lobulated, calcified masses within the bone while parosteal tumors present as exophytic, broad based superficial masses. Calcified masses of the rib can be chondrosarcoma, pleural osteosarcoma, metastatic tumor or myositis ossificans.

In our patient, the tumor was mistaken for neurofibroma on MRI and for chondrosarcoma on CT scan. Osteosarcoma can have osteoblastic, chondroblastic and fibroblastic variants. On radiology, chondroblastic OS is more likely to show aggressive periosteal reactions such as sunburst appearance, Codman’s triangle and onion peel appearance as compared to chondrosarcoma. However, as in our case, the characteristic periosteal reaction of long bone OS may not be obvious in the flat rib bones.

Morphological variants of OS as per their matrix production include osteoblastic OS in 50% cases while chondroblastic OS and fibroblastic OS form 25% cases each. Chondroblastic OS shows spindled or epithelioid tumor cells along with chondroid lobules and osteoid matrix. In addition, surface OS like periosteal OS and parosteal OS can show chondroid elements. The chondroid element raises the differential diagnosis of chondrosarcoma. The latter affects adults with a mean age of 56.7 years while that of chondroblastic OS is 24.7 years in appendicular skeleton. Chondrosarcoma is morphologically characterized by absence of osteoid matrix, presence of chondroid lobules and tumor cells within lacunae. The osteoid matrix is echoed on radiology as fluffy, cloudy opacity while chondroid matrix as stippled calcification. Our case showed classic chondroblastic differentiation coexisting with osteoid matrix on microscopy, thus consistent with chondroblastic OS.

Treatment of rib OS requires wide resection with adjuvant chemotherapy or radiotherapy. Disease free survival of rib OS ranges from 6 to 52 months. Invasion of rib OS into intercostal vessels can cause a fatal hemorrhagic shock. Our patient was treated with subtotal resection and neoadjuvant radiotherapy and chemotherapy. Till date, she did not show recurrence or metastases for fourteen months after therapy.

Conclusion

Location within the rib, symptoms of pleural effusion with compressive myelopathy and adult onset disease are extremely rare features for a primary chondroblastic OS. Histomorphological evidence of osteoid and chondroid matrix formation by tumor cells is the diagnostic gold standard, since radiology poses many diagnostic challenges. Wide excision with adjuvant chemotherapy and radiotherapy is recommended for rib OS.

References