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Dedifferentiated Chondrosarcoma of the Rib Masquerading as a Giant Chest Wall Tumor in a Teenage Girl: An Unusual Presentation

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Chondrosarcoma of the chest wall is a rare primary neoplasm found to occur in elderly men. Patients present with an enlarging, painful, anterior chest wall mass arising from either the vicinity of the costochondral junction or the sternum. Treatment includes wide resection with appropriate chest wall reconstruction. We report an unusual presentation of this uncommon tumor occurring as a huge chest wall mass in a young teenage girl.

Key words: 1. Dedifferentiated chondrosarcoma
2. Ribs
3. Chest wall

CASE REPORT

A 17-year-old girl presented with complaints of a progressively increasing swelling over the front of her left chest over the previous 5 months. The swelling was not painful, situated just adjacent to her left breast, with no associated shortness of breath. There was no history of trauma to the chest wall. She did not complain of an engorgement of the left breast, nipple discharge, or retraction. There was no family history of similar complaints. On examination, her vital signs were stable. There was a large, hard, non-tender mass, 25×30 cm in size, extending from just below the left clavicle to the 6th intercostal space and from the lateral border of the sternum to the mid-axillary line (Fig. 1). The mass itself was adherent to the chest wall although the overlying skin was not. The left breast had been lifted up by the mass and was not adherent to it. There was no palpable breast nodule, ulceration, nipple discharge, or retraction. There were no palpable lymph nodes. Breath sounds were equal and normal bilaterally. The systemic examination was unremarkable. Chest X-ray showed a diffuse opacification over the mid-lung

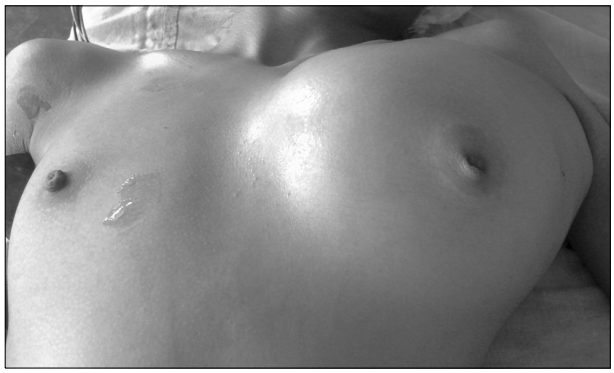

Fig. 1. Preoperative image showing the extent of the chest wall mass.

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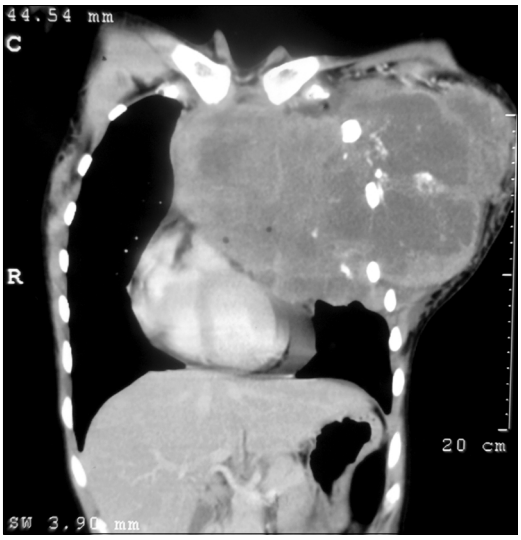

Fig. 2. Computed tomography image showing the extent of the large destructive tumor.

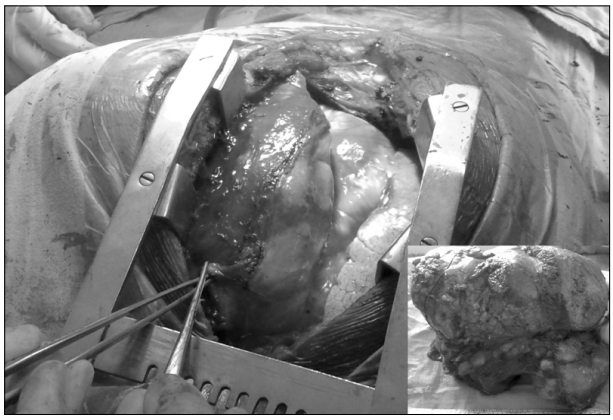

Fig. 3. Intraoperative image showing the tumor (forceps) accessed via a median sternotomy and the completely dissected tumor (inset).

field extending over to the lateral chest wall. Bronchovascular markings in the periphery were normal. A blood investigation revealed a raised lactate dehydrogenase and alkaline phosphatase, positive C-reactive protein, and normal total counts. Contrast-enhanced computed tomography (CT) of the thorax showed evidence of irregular destruction and periosteal reaction involving the left 3rd rib (Fig. 2). There was a large soft tissue component extending through the skin and the subcutaneous region, anterior mediastinum, and in the left upper lobe. This was causing compression of the left upper and

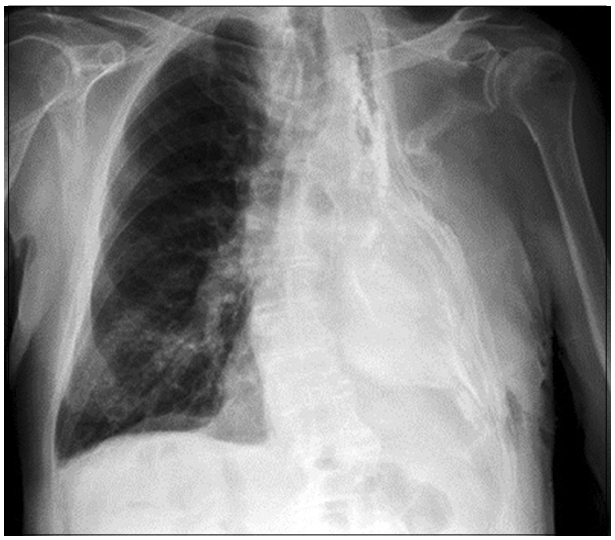

Fig. 4. Post-thoracoplasty chest X-ray.

lower lobe bronchus with a partial collapse of the underlying lung field. The lesion was also abutting the arch of the aorta and its branches and was found to be displacing the left brachiocephalic vein laterally. Magnetic resonance imaging (MRI) findings were similar with the lesion appearing hyperintense in the T2-weighted images and hypointense in the T1-weighted images. Multiple necrotic areas were seen in the lesion. Fine-needle aspiration cytology from the mass was inconclusive, and a core-needle biopsy was taken, which was suggestive of chondrosarcoma. The patient was subsequently taken up for the exploration and excision of the mass. The chest was opened via a median sternotomy and the tumor freed from its anterior mediastinal attachment (Fig. 3). The superior, inferior, and lateral extents of the tumor were accessed via a left thoracotomy, dissecting under the submammary plane, and the required resection of the 2nd to the 5th ribs. Sternal involvement required the resection of the left half of the sternum. Extensive involvement of the left lung parenchyma warranted a left pneumonectomy and a completion thoracoplasty to obliterate the potential space (Fig. 4). Major vascular structures and the left main bronchus were uninvolved. The biopsy report revealed features of a dedifferentiated chondrosarcoma with a highly pleomorphic sarcomatous component. Large foci of necrosis were present, and the tumor was found to infiltrate the lung parenchyma. All margins were found free of tumor involvement, thus signifying a

complete resection. The patient had an uneventful postoperative course and was discharged on the 6th postoperative day.

DISCUSSION

A wide variety of benign and malignant conditions are classified under tumors of the chest wall. The most common entities are rib metastases via a hematogenous spread and direct chest wall invasion from contiguous lung and breast carcinoma. Primary chest wall tumors, which are quite rare, may arise from any of the soft-tissue, bony, or cartilaginous constituents of the chest wall, the latter comprising only one-third of the total number of cases. Chondrosarcoma is the most common malignant primary tumor of both the bony thorax and, in fact, the entire chest wall [1]. It accounts for nearly one-third of all primary chest wall tumors. Ten percent of chondrosarcomas are radiation-induced. Chest wall chondrosarcoma occurs more frequently in men and in an older age group. The occurrence of a large chondrosarcoma in a teenage girl is extremely rare.

Chondrosarcomas are malignant tumors that produce a chondroid matrix. Primary chondrosarcomas arise *de novo*. Secondary chondrosarcomas occur in pre-existing benign cartilaginous neoplasms, such as the complication of a pre-existing enchondroma or osteochondroma. Chondrosarcoma most commonly arises de novo within the medullary cavity of the bone (primary or central) but can result from a malignant transformation of the cartilage cap of a pre-existing benign cartilaginous tumor such as enchondroma or osteochondroma [2,3].

A majority of the patients with thoracic chondrosarcoma present with an enlarging, painful, anterior chest wall mass arising from either the vicinity of the costochondral junction or the sternum. Asymptomatic tumors detected incidentally by thoracic imaging are more likely to be benign. In a retrospective series of patients seen at the Mayo Clinic over 73 years, only 6.5% of the 96 patients were without physical findings or symptoms [1]. A history of trauma associated with the tumor location has been implicated although its direct causation has not been established [3].

A basic posteroanterior and lateral chest radiograph consists of the initial imaging work-up of these patients. A classic radiographic appearance of chondrosarcoma is a lobulated, mixed lytic and sclerotic lesion. The sclerotic regions on the radiographs correspond to the chondroid matrix mineralization. CT is optimal for the detection and characterization of the chondroid matrix, which classically has well-formed rings and arcs of mineralization. The characteristic flocculent or 'popcorn' pattern of calcification has been described for chondrosarcoma. MRI is particularly useful for defining the vascular or neural involvement and, therefore, provides complementary information to the CT scan. Positron emission tomography with fluorine-18 fluorodeoxyglucose is performed to rule out extrapulmonary metastases [4].

Percutaneous image-guided core-needle biopsy and sophisticated cytopathology techniques should yield a diagnosis in a majority of the cases. Permeation of bone, size, and periosteal invasion are specific findings of chondrosarcoma. Cellularity, nuclear atypia (including multinucleated cells), and increased proliferation ($>10\%$) are all indicators of an increasing grade and aggressive behavior. The less-differentiated component of dedifferentiated chondrosarcoma usually shows the multi-lineage histological features of malignant fibrous histiocytoma and fibrosarcoma.

There is no indication of neoadjuvant radiation or chemotherapy in a suitable surgical candidate with a resectable thoracic chondrosarcoma. A wide resection of all thoracic diseases with appropriate margins, reconstruction of the bony chest wall to ensure preservation of respiratory mechanics, and soft-tissue coverage of reconstructive prostheses with healthy, vascularized tissue is the treatment of choice. The chondrosarcoma of the sternum should also be resected with at least a 4-cm margin. This typically requires the resection of the entire body of the sternum. The ipsilateral lung should be carefully palpated for occult metastases, which should be resected prior to the commencement of reconstruction [5,6]. In this patient, due to the extensive involvement of the chest wall and the lung parenchyma, pneumonectomy with thoracoplasty had to be done. Thoracoplasty is the surgical removal of the skeletal support of a portion of the chest. This is accomplished by the subperiosteal removal of a varying number of rib segments to approximate the chest wall to the underlying lung or mediastinum to effect lung collapse or

pleural space obliteration. In spite of mutilating surgery, thoracoplasty is justified in developing nations where issues such as poor patient compliance, post pneumonectomy empyema, drug resistance, poor healthcare delivery systems, and a certain amount of mismanagement at the primary and secondary levels of healthcare need to be addressed.

Chondrosarcoma is relatively radioresistant, and conventional radiation therapy has demonstrated some efficacy and should be administered at the sites of positive pathologic margins (if a wider surgical margin cannot be achieved) or in cases of unresectability. Although considered a rare entity, chondrosarcoma of the chest wall is the most common malignant primary neoplasm. Its occurrence in a young female and with such a dimension as that described above is extremely rare. A careful work-up as well as histological correlation is vital for the adequate management of these patients, who require a wide resection of the tumor supplemented with chest wall reconstruction, if possible. Lung parenchymal infiltration by the tumor is an indication for resection. Overall survival is good if the tumor has been adequately resected, respiratory mechanics preserved, and the margins are found to be free of involvement during a histopathological examination [7,8].

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

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