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Can Costal Chondrosarcoma Be Cured?

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Abstract **Case Report**

Primary chest wall tumours are rare tumours which account for <5% of thoracic malignancies. 50-80% primary chest wall tumours are malignant tumours. Thoracic chondrosarcoma is the commonest malignant primary chest wall tumour which occurred in 30% of all malignant primary chest wall tumours. We report a case of a 62-year-old gentleman presented with left chest wall mass associated with pain. CT scan showed left 9th rib mass with intraperitoneal extension. Histopathological examination showed costal conventional chondrosarcoma, grade 2. Patient underwent tumour resection with a safe margin of 4cm, and chest wall reconstruction. Subsequently, he had radiotherapy due to close surgical margins. After 6 months' follow-up, patient's ECOG is 0 and no evidence of recurrence.

Keywords: Costal chondrosarcoma, chest wall tumour.

INTRODUCTION

Primary chest wall tumours are rare tumours which account for <5% of thoracic malignancies [1]. 50-80% primary chest wall tumours are malignant tumours [2]. Thoracic chondrosarcoma is the commonest malignant primary chest wall tumour which occurred in 30% of all malignant primary chest wall tumours [3]. Chondrosarcoma is a malignant tumour with cartilaginous differentiation, contains foci of tumour-induced or endochondral ossification with definitive diagnosis based on histology. CT imaging is required for diagnosis, for assessment of tumour invasion to nearby structure or metastasis, and for operative planning. Surgical excision with wide safe margin is the definite treatment since costal chondrosarcoma is relatively insensitive to chemotherapy and radiotherapy [4].

CASE PRESENTATION

A 62-year old gentleman with underlying hypertension and dyslipidemia presented with left chest wall mass which increased in size progressively over 6 months' period and associated with pain [Figure 1]. He had no constitutional symptom, no underlying bone disease, and no history of chemotherapy or radiotherapy. Chest X-ray showed vague left chest wall mass (red circle area) without lung abnormality [Figure 2]. CT thorax and abdomen showed left 9th rib mass with intraperitoneal extension but no organ invasion [Figure 3]. Fine needle aspiration showed chondrosarcoma.

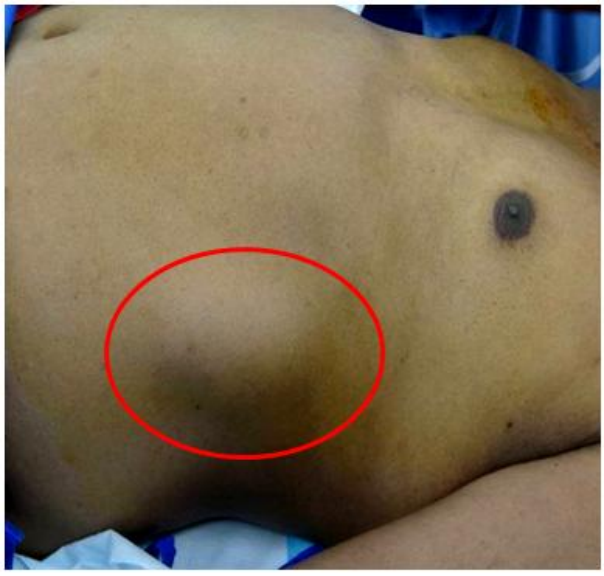

Fig-1: Left chest wall mass (red circle area)

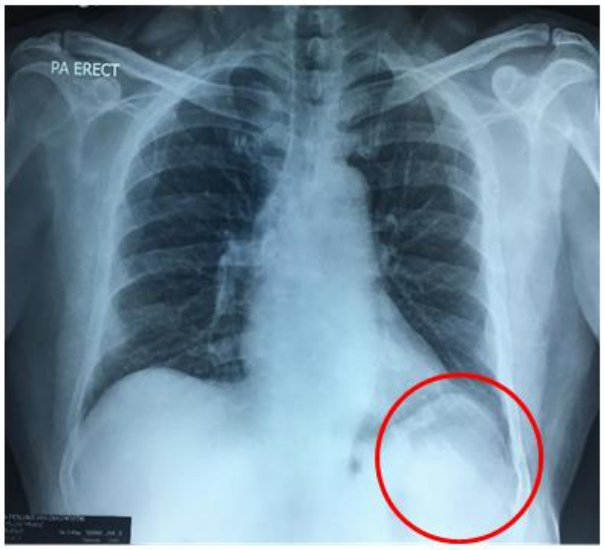

Fig-2: Chest X-ray showed vague left chest wall mass (red circle area) without lung abnormality

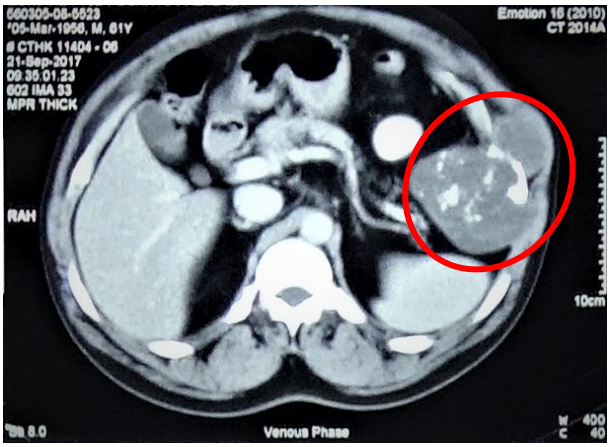

Fig-3: CT thorax and abdomen showed left 9th rib mass with intraperitoneal extension (red circle area) but no organ invasion

After case discussion in multi-disciplinary meeting which includes cardiothoracic surgeons, plastic surgeons and orthopedic surgeons, patient was arranged for surgery. Intra-operative findings showed left 9th rib mass measured 8x10cm externally with involvement of overlying muscle, and intraperitoneal extension measured 10x15cm with diaphragm involvement measured 5x7cm [Figure 4]. Otherwise, no intraabdominal organs invasion noted. Hence, enbloc resection of diaphragm, rib mass, overlying muscle with a safe margin of 4cm were performed, followed by repair of diaphragm (direct suture of diaphragm to chest wall) and chest wall reconstruction with rectus abdominis muscle flap to cover 12x12cm chest wall defect resulting from tumour resection [Figure 6].

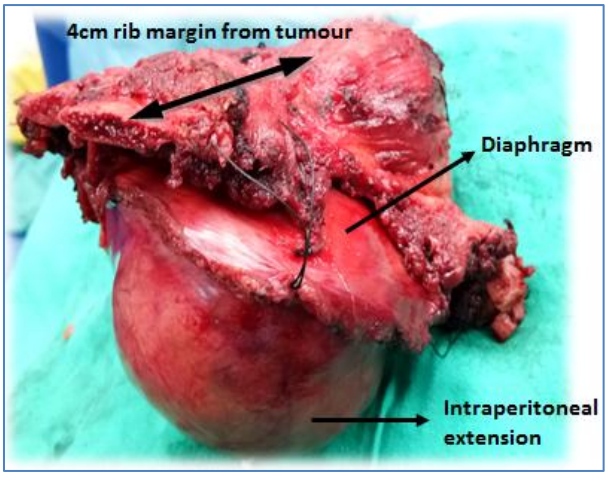

Fig-4: Left 9th rib mass measured 8x10cm externally with involvement of overlying muscle, and intraperitoneal extension measured 10x15cm with diaphragm involvement measured 5x7cm. Surgical resection with safe margin of 4cm

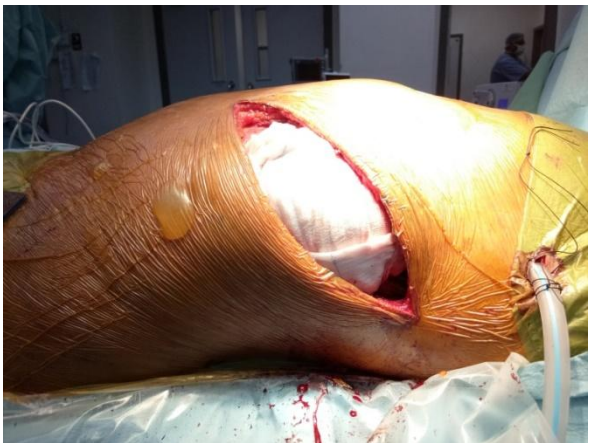

Fig-5: 12x12cm chest wall defect after rib tumour resection

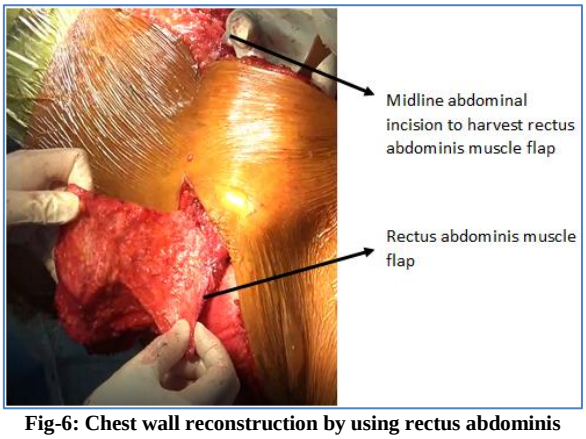

Fig-6: Chest wall reconstruction by using rectus abdominis muscle flap

Histopathological examination showed conventional chondrosarcoma, grade 2, Stage IIb (pT2N0M0). Subsequently, he had radiotherapy due to close surgical margins. After 6 months' follow-up, patient's ECOG is 0 and no evidence of recurrence.

DISCUSSION

Primary chest wall tumours are rare tumours which account for <5% of thoracic malignancies [1]. 50-80% primary chest wall tumours are malignant tumours [2]. Thoracic chondrosarcoma is the commonest malignant primary chest wall tumour which occurred in 30% of all malignant primary chest wall tumours [3]. Chondrosarcoma is a malignant tumour with cartilaginous differentiation, contains foci of tumour-induced or endochondral ossification. It commonly arises de novo bone (primary or central) within the medullary cavity of but can result from malignant transformation of the cartilage cap of a preexisting benign cartilaginous tumour such as enchondroma or osteochondroma (secondary or peripheral) or can arise in patient with previous radiation [5]. Majority of the patients present with an enlarging painful anterior chest wall tumour. It occurs predominantly in males and usually presents above 50 years old.

Every patient with chest wall mass will have chest X-ray as initial investigation. It may occasionally reveal the features of costal chondrosarcoma include "popcorn" calcification, endosteal scalloping and lytic lesion [6]. However, the chest X-ray of this patient did not show any of these features. Apart from X-ray, contrasted CT scan of thorax and upper abdomen should be performed in every patient with chest wall mass for diagnosis since it is more sensitive than X-ray to reveal the salient feature of chondrosarcoma which is a well-defined mass with foci of chondroid matrix calcification or "popcorn" calcification [7]. Besides, CT scan is important to detect the extent of invasion, pulmonary metastasis and for operative planning. Rarely, MRI is required for costal chondrosarcoma with mediastinal, paravertebral, or thoracic outlet involvement to define vascular or neural involvement. PET scan is sometimes performed to differentiate benign cartilaginous tumours from chondrosarcoma and to rule out extrapulmonary metastases. Histopathology is the gold standard of diagnosis either through percutaneous biopsy or open biopsy [8]. In view of possible tumour seeding along the biopsy tract, the site of biopsy should be decided with consideration given for eventual removal at definitive resection later[9].

First line therapy of costal chondrosarcoma is surgical resection in view of costal chondrosarcoma is relatively resistant to chemotherapy or radiotherapy [10]. Since costal chondrosarcoma has high potential for locoregional invasion and recurrence after treatment, wide resection should be performed by removing rib en bloc with a safety margin of at least 4cm and, two intercostal spaces above and below. If overlying skin, subcutaneous tissues, muscle and internal organ are involved, they should be resected en bloc with an appropriate margin. If uninvolved by tumour, they may be spared. In view of large chest wall defect after surgical resection, chest wall reconstruction is crucial to maintain chest stability, respiratory functions and protect internal organs. These reconstruction techniques include pedicled skin and muscle flaps, free skin grafts, autologous bone transplants or prosthetic material depending on the extent and localization of the defect. In our case, chest wall reconstruction was performed by using rectus abdominis muscle flap for inferior anterolateral chest wall defect.

Costal chondrosarcoma is a slow growing tumour with low fraction of dividing cells. Hence, chondrosarcoma is relatively resistant to radiotherapy or chemotherapy. Radiotherapy can be considered in situations where resection is incomplete or resection is not feasible. While chemotherapy should be considered in patients with mesenchymal chondrosarcoma or in the case of metastatic disease as palliative chemotherapy [11].

Costal chondrosarcoma has high possibility of recurrence despite of treatment especially in patient with high tumour grade or inadequate margins at initial resection. Hence, lifelong surveillance is mandatory. Surveillance consists of physical examination and thoracic imaging with either postero-anterior/lateral radiograph or CT scan every 3–6 months for the first 5-years and annually thereafter for a minimum of 10-years.

CONCLUSION

Wide surgical excision is the first line therapy for chondrosarcoma and chest wall reconstruction is often needed to maintain chest stability, respiratory functions and protect internal organs. Due to the possibility of local and systemic recurrence, patient should undergo routine lifelong surveillance after surgery.

Lists of abbreviations used

ECOG - Eastern Cooperative Oncology Group performance status
CT - Computed tomography
MRI - Magnetic resonance imaging
PET - Positron emission tomography

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