

## CASE REPORT

# Resection and primary reconstruction of a massive chest wall chondrosarcoma

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## Abstract

Primary malignant tumours of the chest wall are among the rarest cartilaginous tumours. Chondrosarcomas present a difficult clinical problem due to its high resistance to conventional chemotherapy and radiotherapy. Complete surgical resection has been the cornerstone for treatment. It has been associated with better prognosis, survival and less recurrence in contrast to other methods of therapy. Patients with chondrosarcomas generally have a good prognosis when surgical resection is performed. Improvement in outcomes is seen when patients are evaluated in a multidisciplinary care facility. We present the case of a 66-year-old male patient that exhibits a chest wall mass diagnosed as a chondrosarcoma. After chemotherapy failure, our patient was successfully treated with complete surgical excision of the mass. Final tissue biopsy report was remarkable for a p53 gene mutation, which is known to be associated with tumour progression and loss of growth control.

## INTRODUCTION

Primary chest wall tumours are uncommon [1]. Chondrosarcomas are the most common tumours arising from the chest wall [1]. This neoplasm is more common in men than woman and occurs more often during the third and fourth decade of life [1]. Some of the histopathologic variants are conventional, clear cell, mesenchymal and dedifferentiated chondrosarcomas [2].

These tumours tend to present as a slowly growing mass. In rare cases, a p53 gene mutation may induce tumour progression and loss of growth control. Chondrosarcomas are largely considered to be resistant to conventional chemotherapy and radiotherapy [2]. For this reason, wide margin surgical excision remains the best available surgical approach [3]. When chondrosarcomas involve the chest wall, surgical excision may result in chest wall defects that require reconstruction to obliterate dead space,

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Figure 1: Massive anterior chest wall mass that measured approximately 25 cm in length.

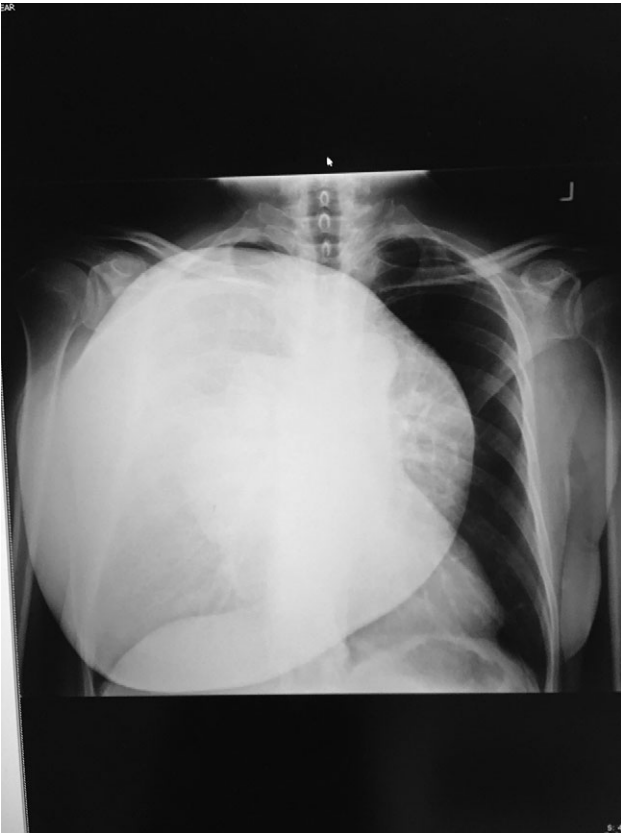


Figure 2: Chest X-ray showing a large lobulated soft tissue mass overlying the right chest.

restore chest wall rigidity, preserve respiratory mechanics, maintain pulmonary function, protect intrathoracic organs, provide soft tissue coverage and minimize deformity [4, 5].

## CASE REPORT

A 66-year-old male patient with medical history significant for Hypertension, Gout and GERD presented to his primary care physician with a slow-growing chest mass over a 10-year period, associated with chest pain during the latter 5 years. Home medications included lisinopril, allopurinol and ranitidine. Patient denied alcohol consumption, illegal drugs, Tb exposure and previous history of malignancy or tobacco. A needle biopsy and a subsequent minimally invasive incisional biopsy, performed at a local hospital, were nondiagnostic. A later biopsy, performed at a tertiary care hospital and evaluated by the University Hospital Pathology service reported a high-grade chondrosarcoma. The patient initially refused surgery which prompted initial treatment with a Doxorubicin and Cisplatin chemotherapy regimen. Chemotherapy was discontinued after 10 months due to an accelerated mass growth.

At that point, physical examination was remarkable for a round, firm, non-tender chest wall mass, confirmed by chest radiography (Figs 1 and 2). A magnetic resonance imaging (MRI) revealed a large right chest wall mass originating posteriorly to the right pectoralis major muscle, infiltrating the right anterior rib cage and extending into the right anterior thoracic cavity and the superior mediastinum. The mass measured 17.0 cm on the AP diameter, 20.6 cm on the transverse measurement and 18.3 cm in the cranio caudal diameter. The mass was adjacent to the Superior Vena Cava, and the right anterolateral surface of the ascending thoracic Aorta (Fig. 3). The mass itself had multiple septa, cystic and solid components. It was producing complete destruction and encasement of the right anterior chest wall including the anterior right ribs and the right surface of the sternum. The patient was then evaluated by the Cardiothoracic and Plastic Surgery services, which

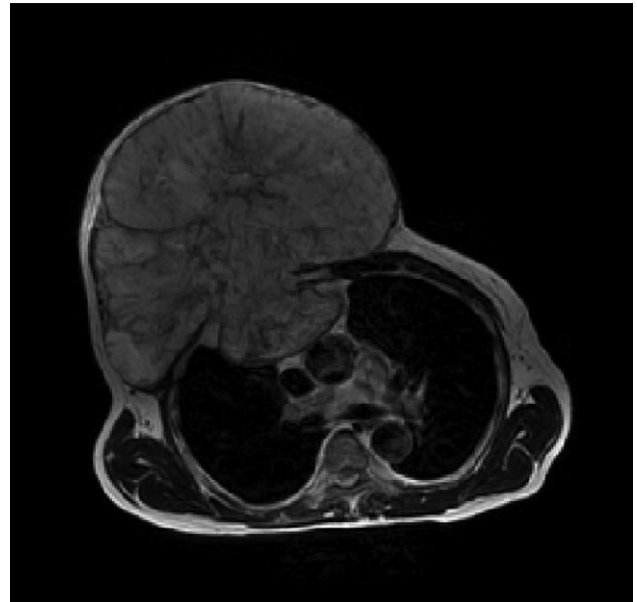


Figure 3: Thoracic MRI axial T2 image demonstrates a large, infiltrative heterogeneous soft tissue mass centred in the right chest wall with associated cortical destruction of the rib/lateral border of the sternum and invasion into the anterior mediastinum.



Figure 4: Intraoperative chest wall images, showing mass exposed.

recommended and together performed a complete resection with chest wall reconstruction.

The surgery entailed elevation of a lateral skin-muscle flap, elevation of a medial skin-fascia flap, and exposure of the mass, while being careful to maintain margins free of malignancy: a layer of normal muscle tissue along its anterior surface and margins of normal bone or cartilage at the chest wall (Fig. 4). En-bloc excision of the mass, with pectoralis major tissue, the right pectoralis minor muscle and the anterior chest wall, from the right nipple line to the left margin of the sternum, was performed, with a final specimen measurement of  $27 \times 24 \times 17$  cm, and a weight of 4.5 Kg (Fig. 5).

The excision resulted in resection of five ribs (second to sixth), near total resection of the sternal body, and exposure of the mediastinum and right lung (Fig. 6). The chest wall was reconstructed with a synthetic bony cement (methyl methacrylate) construct, sandwiched between two embedded layers of polypropylene mesh. The prosthesis was designed with a paper template of the chest wall defect, constructed on a back table with the methyl methacrylate and shaped during the cement hardening reaction. The cement construct was inset in the chest wall defect and the fringe of mesh beyond the cement edge was fixed to the bony margins of the defect with polypropylene sutures, and to the surrounding soft tissues with polyglactin sutures. Wound closure was completed by advancing, tailoring, and inserting the aforementioned flaps, over the reconstructed chest wall. The patient was extubated in the operating room, had an uneventful postoperative course, was discharged home on postoperative day 11, and recovered well from his surgery (Figs 7 and 8). Subsequently a P53 mutation was identified in tumour. Pre- and post-surgery pulmonary



Figure 5: Post-surgical mass specimen that weighted approximately 10 pounds.

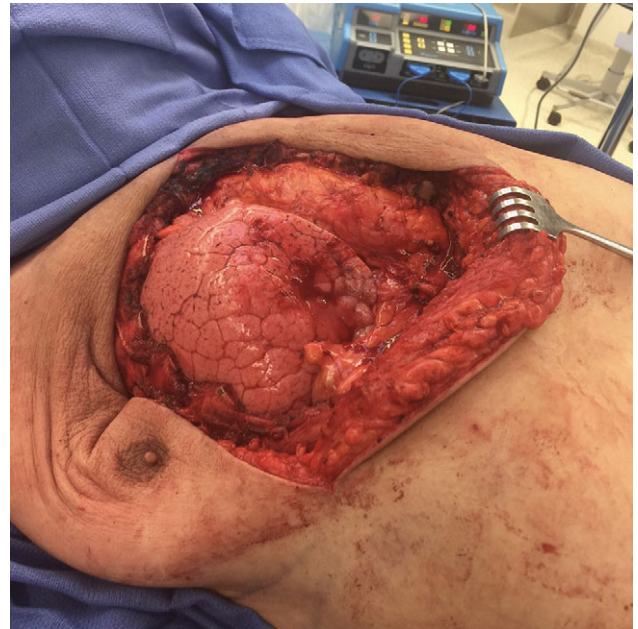


Figure 6: Intraoperative chest wall images, showing anterior thoracic cage with several incised ribs.

function tests revealed a normal spirometry, lung volumes and no abnormality in diffusing capacity (Table 1).

## DISCUSSION

Primary malignant tumours of the chest wall are very uncommon in clinical practice, accounting for 8% of all chest wall tumours [6]. The annual incidence of chest wall chondrosarcoma is less than 0.5 per million [7]. Our patient presented the

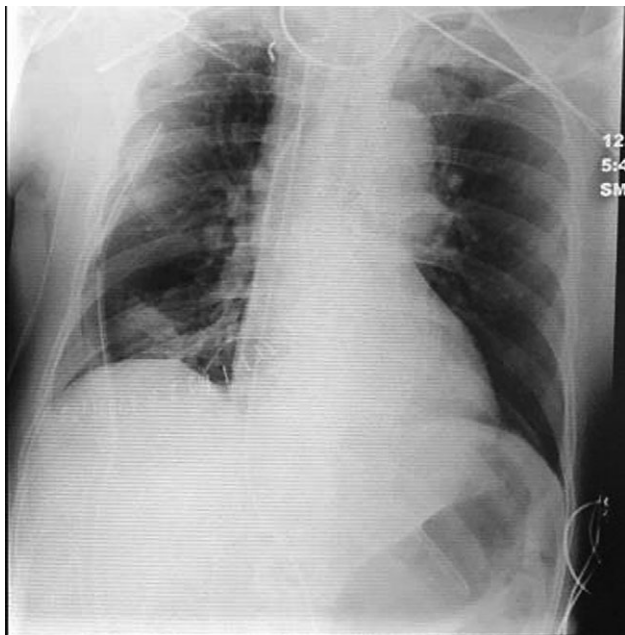


Figure 7: Post-surgical chest X-ray without thoracic mass.

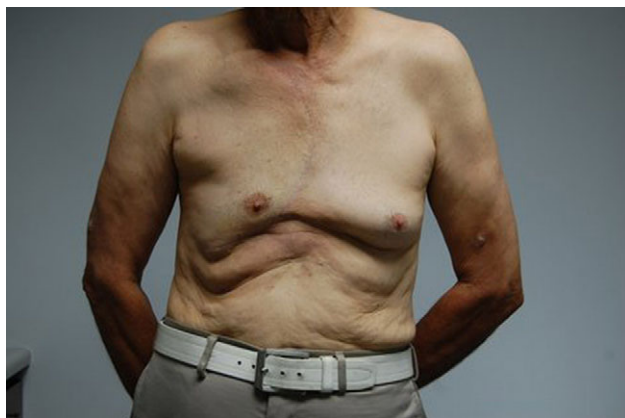


Figure 8: Post-surgical and reconstructive surgery.

most common manifestation; a palpable mass associated with chest pain, which occurs in approximately 80% and 60% of cases, respectively [1, 8]. Chondrosarcomas constitute a group of neoplasms typical for the production of cartilage matrix by the tumour cells [9]. Conventional chondrosarcoma is the most common type, but several rare subtypes exist, such as Dedifferentiated chondrosarcoma, Mesenchymal chondrosarcoma and clear cell chondrosarcoma [3]. Primary chondrosarcomas arise *de novo* and secondary chondrosarcomas emerge from pre-existing benign cartilaginous neoplasms [9]. The natural history and prognosis of chondrosarcomas is extremely variable [9]. There have been few long-term studies on the treatment outcomes for chondrosarcomas [9].

Computed tomography and MRI are useful to characterize the tumour and its extension [7]. Different techniques are available to obtain a tissue diagnosis, including minimally invasive incisional or needle biopsies, however, excisional biopsies are preferred [1, 6, 9]. Management strategies are diverse and depend strongly on the pathological diagnosis and

Table 1: Patient pulmonary function test—pre- and post-surgery.

Lung function test	Pre-surgery	Post-surgery
FEV1	2.77 L (97%)	2.95 L (105%)
FVC	3.26 L (89%)	3.27 L (90%)
FEV1/FVC	85%	90%
TLC	5.67 L	5.61 L
DLCO	19.4 ml/mmHg/min	19.3 ml/mmHg/min
RV/TLC	42%	41%

the extent of disease. For grade I chondrosarcomas with intact cortex and absence of soft tissue mass, intraregional procedure such as curettage with adjunctive ablation can be considered [9]. However, if there are aggressive imaging features such as cortical breakthrough, soft tissue mass or the tumour is grade II or higher, wide surgical excision is required [9]. Wide, en-bloc surgical excision remains the best available treatment for intermediate to high-grade chondrosarcomas [3]. The 5-year survival rate after resection with adequate surgical margins (4 cm on each side) was 100% compared with 50% in patients with inadequate surgical margins [1]. Inadequate margin of resection is associated with worse overall survival and a higher chance of having local recurrence [1]. Our patient initially refused surgery and had a poor response to chemotherapy. It is generally believed that this is due to penetration through the extracellular matrix, low percentage of dividing cells, and poor vascularity, chondrosarcomas are relatively chemo-radiotherapy resistant with the exception of mesenchymal chondrosarcoma with a limited number of cases reporting adequate response [3].

In conclusion, patients with chondrosarcomas generally have a good prognosis when optimally diagnosed and treated [3]. Our case reports interesting and unique findings due to the large size of the chondrosarcoma. One of the largest encountered when compared to reported literature, and its unusual growth and tumour progression after initiation of chemotherapy, which was related and has been associated to the presence of a p53 gene mutation [10–12]. Complete resection with wide surgical margin excision remains the best available treatment [1]. It is highly recommended that the patient be referred as soon as possible to a multidisciplinary care centre with an experienced team to improve patient outcomes [7].

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## CONFLICT OF INTEREST STATEMENT

There is no conflict of interest that could be perceived as a prejudice to the impartiality of this case.

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## PATIENT CONSENT

A written informed consent was obtained from the patient for the documentation and publication of this case.

**GUARANTOR**

Michael Cruz Caliz

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