

Analysis of Surgical Resection and Prognostic Factors of Primary Chest Wall Chondrosarcomas

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ABSTRACT

Introduction: Primary chest wall tumours are very rare. Chondrosarcoma represents 40% of all chest wall tumours. Wide local excision with tumour free margins has been the gold standard therapy. We evaluated this therapy in relation to various prognostic factors for anticipating the recurrence of the tumour.

Material and methods: 22 cases of Primary chest wall chondrosarcoma was operated upon from 2009 to 2019 with wide local excision with adequate margins. Male constituted 77.27% and females 22.73% with the tumour size ranging from 5 to 30 cm in size (median 7cm).

Results: 19 patients were subjected to lateral chest wall resection. 3 cases underwent partial sternectomies. Resection was extended to lungs in 7 cases, diaphragm in 2 cases, vertebral body in 2 cases and clavicle in 2 cases. Reconstruction was done with polypropylene or titanium mesh and a muscle flap coverage. There was no perioperative mortality. Post-operative complications occurred in 13.6% (n=3). There was recurrence in 4 cases within 5 years. 5 year disease free survival rate was 81.81%.

Conclusions: Wide local excision with tumours free margins still remains the standard form of therapy. The reconstructive procedures have evolved largely to cover the huge defects following surgery. Post operative adjuvant therapy is guided by various prognostic factors.

Keywords: Primary Chest Wall Chondrosarcomas, Wide Local Excision, Chest Wall Reconstruction, Prognostic Factors

INTRODUCTION

Primary chest wall tumours are uncommon.¹ 15% of chondrosarcomas are located in the thoracic cage, making it to be the most frequent primary malignant chest wall tumour.² 80% originate from the cartilaginous and bony structures of ribs and 20% originate from the sternum.³⁻⁵ It is a slow growing tumour. Males are affected more commonly. It usually occurs during the third and fourth decade of life. Median survival for these tumours is 2.5% and overall survival is 46%.⁶ Because of its resistance to radiotherapy and most of chemotherapy drugs, wide local excision with adequate tumor free margin is the only curative option for primary chest wall chondrosarcomas.^{2,7} Often a large defect is created in the chest wall following surgery which can lead to chest wall instability and respiratory dysfunction. This necessitates complex reconstructive procedures which have been made possible by advances in surgical technique and in various prosthetic and biological materials to provide chest

stability and to reduce respiratory dysfunction.⁸⁻¹⁰ The aim of our study was to evaluate various surgical techniques along with reconstructive procedures for covering the huge chest wall defects and also to evaluate various prognostic and clinical risk factors.

MATERIAL AND METHODS

A total of 22 cases of primary chest wall chondrosarcoma was operated upon from 2009 to 2019 in the Department of Cardiothoracic Surgery of Medical College Kolkata. Males constituted 77.27% and females 22.73% with the tumour size ranging from 5 to 30 cm in size (median 7cm) (Fig 1). Data of these patients were reviewed with respect to gender, age, tumour size, localization of tumour, histological grade, type of resection and reconstruction, tumour free surgical margins, adjuvant chemotherapy, recurrence and disease free survival. These patients presented with chest wall mass of varying size and locations. Diagnostic work up included a standard chest x-ray and CT Scan (Fig 2) chest to delineate the extent of tumour. A tissue diagnosis was obtained either with FNAC or a tru cut biopsy. Evidence of distant metastasis was obtained by PET scan. In cases of the tumours extending posteriorly near the vertebrae, MRI was added to the diagnostic spectrum to rule out spinal cord involvement. Surgical excision was performed with skin incision including the site of previous biopsy along with excision of the involved ribs and adjacent lungs. An average of 3 to 7 ribs were excised. Partial sternectomy was done in 3 cases. Adequate resections with approximately atleast 3 cm margins around the tumour was done. Small tumours in the posterior aspect did not required any reconstructions. Tumours extending near the vertebral body required neurosurgical expertise for stabilisation of vertebral bodies

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with titanium plates and screws. All the specimen was subjected to histopathological examination and the degree of differentiation recorded. The defects in the lateral chest wall was reconstructed with polypropylene mesh(Fig 3) and reinforced by latissimus dorsi flap mostly(Fig 4). Few cases required reinforcement with titanium mesh and additional reinforcement with stainless steel wires to maintain the chest stability(Fig 5). Anterior defects in addition was reconstructed with pectoralis major flap. After the surgical procedures the patient was extubated and shifted to intensive care unit for a routine post-operative care. 2 patients with large sized tumours were shifted to intensive care unit with endotracheal tube and extubated the next day. After an average of 7 to 10 days hospital stay, the patient was discharged and there was no peri-operative mortality.

Any postoperative complications were noted and recorded and appropriate measures were taken. Tumors with higher grades and inadequate tumour free margins were subjected to post-operative chemotherapy. These cases were routinely followed up in the out patient department and were evaluated for appearance of any local recurrences or distant metastasis.

RESULTS

We had performed surgical resection with reconstruction in 22 cases of PCWC between 2009 to 2019. There were 77.27% males (n=17) and 22.73% females(n=5). The tumour size ranged from 5-30 cm with a median of 7cm. 3 cases with huge anterolateral tumors with partial involvement of sternum underwent partial sternotomies with excision of lower and middle body of sternum. 19 cases underwent lateral chest wall resections without sternal involvement. The median number of ribs resected was 2 with a range from 2 to 7 ribs. Resection was extended to lungs in 7 cases(31.82%), to the diaphragm in 2 cases (9.09%), to the vertebral body in cases (9.09%) and to the clavicle in 2 cases(9.09%). Reconstruction of the anterolateral defects was carried out with polypropylene mesh which was covered with pectoralis major flap or latissimus dorsi flap in 59.09% cases(n=13). Lateral chest wall reconstruction was not required in small and posterior defects less than 7cm in 40.09% cases (n=9). Tumor free margins were obtained in 86.36%(n=19). Post operative histopathological examination revealed a G1 tumor



Figure-1: Showing a huge primary chest wall chondrosarcomas

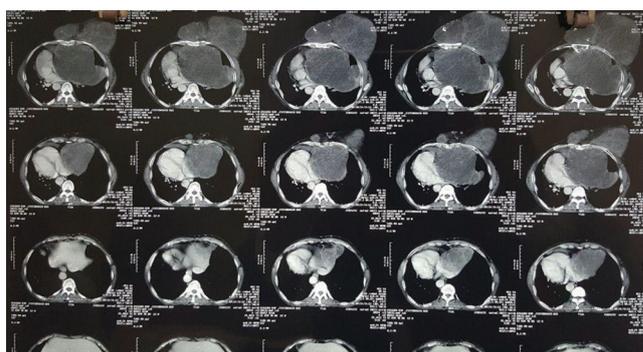


Figure-2: CT Scan showing the extension of chondrosarcoma

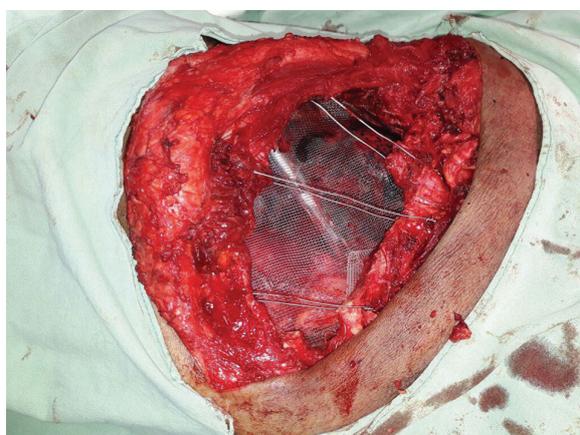


Figure-3: Reconstruction with polypropylene mesh

Serial no.	Parameters	Recurrence/no. of patients	percentage
1	Gender		
	Male	4/17	23.52%
	Female	0/5	0%
2	Age - >55 years	4/17	23.52%
3	Negative margins		
	yes	1/19	5.26%
	no	3/3	100%
4	Histological grade		
	G1	0/9	0%
	G2	2/10	20%
	G3	2/3	66.68%
5	Adjuvant treatment	4/5	80%
6	Dimensions > 8cm	4/8	50%

Table-1: Prognostic factors for recurrences



Figure-4: Reconstruction with Latissimus dorsi flap



Figure-5: Reinforcement with stainless steel wire and titanium mesh

in 9 cases (40.91%), G2 tumors in 10 cases (45.45%), and G3 tumors in 3 cases (13.64%). There was no perioperative mortality, however a prolonged endotracheal intubation was required in 2 cases with a huge anterolateral tumor. 2 cases had prolonged fever which resolved with appropriate antibiotics and 1 case experienced post operative pneumonia. These post operative complications in 3 patients accounted for 13.6%. 5 patients (22.73%) were subjected to adjuvant chemotherapy. Indications for adjuvant chemotherapy was incomplete resection or inability to obtain tumour free margins after surgical resection and a high grade G3 tumours. There were 4 cases of recurrences in 5 years follow up with 1 case having distant metastasis. 5 year disease free survival was 81.80%. There were recurrence in another 2 cases beyond 5 years of follow up.

Recurrences in 5 years follow up was seen in males, age >55 year, in tumour size >8cm, in surgical resection without adequate tumour free margins, in high grade tumours and in patients receiving adjuvant chemotherapy (table - 1)

DISCUSSION

Chondrosarcoma is the most frequent primary malignant tumour of the thoracic cage. Though it represents 40% of all chest wall tumours^{2,11}, the incidence is <0.5 cases per million persons per year. They are lobulated tumours of varying sizes. It may grow to massive proportions. They may extend internally to the pleural space and intra-thoracic cavities or

externally invading muscle, adipose tissue and skin of the thoracic wall. The patients present with slow-growing mass for several months or years. Palpable mass in thorax is the main symptom in approximately 80% of the patients. Of these, 60% present with associated pain.³

The diagnostic tools like Computed tomography (CT) and magnetic resonance imaging (MRI) are important to delineate the tumour and its extension. CT is superior to MRI in demonstrating calcification. MRI is the imaging modality of choice to evaluate the tumour extension and its relationship with adjacent structures.¹²⁻¹⁶

As compared to other forms of malignancy, the surgical management of PCWC have not evolved over time except for the reconstructive surgery to provide chest stability and improved respiratory hemodynamics. There is still no effective induction or adjuvant therapy for chondrosarcoma as it is relatively radio- and chemoresistant. There are few studies on the mechanisms underlying this resistance. Expression of the multidrug-resistance-1 gene, P-glycoprotein, has been reported in most chondrosarcomas^{18,19} leading to resistance to doxorubicin in vitro.²⁰ The presence of large amount of extracellular matrix may prevent the access of anticancer agents. Chemotherapeutic agents are usually ineffective in chondrosarcoma because of their slow growth in nature. In this regard, surgery is the only effective form of treatment for PCWC, regardless of the size or histological grade of the tumour, as long as it is resectable.^{2,7,21} The standard form of surgical resection is wide local excision with adequate margin (at least 3cm of tumour free margins)^{2,7} to prevent local recurrence. The recurrence rate for patients with adequate surgical margins was 10%, compared with 75% for patients with inadequate margins.¹⁷ In our study, 5 year recurrence rate was 5.26%, and 15.79% recurrence beyond 5 years in cases where adequate tumour free surgical margins were obtained and almost all cases with inadequate tumour free margins recurred within 5 years. An inadequate margin of resection was associated with a significantly worse overall survival and a higher chance of having local recurrence develop.¹⁷ The 5-year survival rate in our study for patients with adequate surgical margins was 100%, compared with 50% in patients with inadequate surgical margins. Local recurrence-free survival is around 64%.⁶ In our study, 5 year disease free survival was 81.80%.

The extent to which a radical resection is feasible depends on several factors: the localization of the tumour (difficult in the paravertebral area), the involvement of mediastinal structures and the dimension. In our study the median tumour diameter was 7 cm, comparable with 8 cm reported by Widhe et al.²¹ The relative large median diameter of the tumour requires more complex and extended resections. Early diagnosis is of utmost importance in order to allow radical resection and reduce the risk of recurrence. The results depend also on the biological aggressiveness of the disease as depicted by histological grades. According to cell differentiation, Campanacci et al.²² grouped malignant progression into three grades: Grade 1, consisting of well-differentiated cartilaginous cells with rare and doubtful

aspects of atypia; Grade 2, presenting with some atypical cells; and Grade 3, comprising scarce cartilaginous cells in a context of predominant atypia. In our study, G1 comprised 36.36%(n=9), G2 comprised 45.45%(n=10) and G3 comprised 18.18%(n=3). The 5-year survival rates for Grades 1, 2 and 3 were 100, 80, and 33.33%, respectively, in our study and the grading was a significant predictor of survival.

To achieve this goal of radical surgery and to reduce the risk of postoperative complications, resection and reconstruction techniques have to follow some principles: maintenance of chest stability, adequate respiratory dynamics, absence of harmful paradox movements, and an acceptable cosmetic result.²³ A healthy soft-tissue coverage is required to seal the pleural space, to protect the viscera and great vessels and to prevent infection.^{23,24,25} The most commonly used non rigid prosthetic materials are Marlex mesh and ePTFE.²³⁻²⁷ The choice of prosthetic material is usually based on the surgeon's preference. Deschamps et al.²⁶ have shown no significant difference in the postoperative outcome or in complication rate because of this choice. In the case of large anterolateral chest wall defects with or without involvement of sternum, a reconstruction with rigid material is mandatory to restore chest-wall stability. This maintains the geometry of the thoracic cage and offers adequate protection of the surrounding structures.^{28,29} There are four ideal characteristics of prosthetic material, according to LeRoux and Shama³⁰: rigidity, malleability, radiolucency and inertness. Marlex-methacrylate prostheses satisfies the first 3 criterias. The fourth ideal criteria, inertness, was still lacking, because rigid shields prevents ingrowth of fibrous tissue and limits integration of muscle flaps. This leads to increased dead spaces and the likelihood of infection. A large prosthesis used for large defects increases the rigidity of the chest and patient discomfort. Different centres worked on different techniques or used new materials to expand the possibility of rigid reconstruction, to improve biocompatibility and overcome problems like respiratory failure and local infection. The 'rib-like' reconstruction, as used by the Instituto Nazionale Tumori in Milan⁸ and the cryopreserved allograft of cadaveric sternum with attached costal cartilage¹⁰ were technical evolutions developed to obtain a biocompatible prosthesis. These new evolutions protected the mediastinum and reproduced the tridimensional sternocostal anatomy. This also allowed favourable respiratory dynamics. After chest wall stabilization, a soft-tissue coverage is used to complete the reconstruction. This controlled infections, obliterate dead space, and cover the synthetic material. Advancements in reconstructive surgery with the use of various muscles or musculocutaneous flaps have been made possible. The selection of the type of flaps was discussed preoperatively with our plastic surgeon. Pectoralis major flap was preferred after partial sternectomies, because of its proximity, reliability and versatility. Because of its length and bulk of muscles it provides to cover extensive defects in chest wall, Latissimus dorsi flap transposed on thoracodorsal vessels was preferred for anterolateral chest wall defects.

CONCLUSIONS

Considering the various advancements and progress in multimodality treatment of different malignancy, the management of PCWC solely relies on wide local excision with adequate tumour free margins and developments in reconstructive procedures and biocompatible prosthesis. Wide surgical resection with tumour free margins is necessary to reduce the incidence of local recurrence. Our experience found that the age of the patient, tumour size, grades of tumour, ability to obtain tumour free margins and adding adjuvant therapy were independent prognostic factors for recurrence and disease free survival.

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