

Reconstruction of the chest wall after excision of a giant malignant peripheral nerve sheath tumor

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Abstract

Primary chest wall tumors are uncommon and constitute 0.2-2% of all tumors. Metastatic tumors and tumors of local extension are more common. Malignant peripheral nerve sheath tumor (MPNST) of the chest wall is even rarer and its incidence on the chest wall not stated in the literature. The incidence in the general population is 0.0001% while the risk is approximately 4600 times higher in patients with type I neurofibromatosis and 3-13% of them will finally develop into MPNST, usually after latent periods of 10-20 years. Clinically, these tumors are aggressive, locally invasive, and highly metastatic. Excision of giant chest wall tumor leaves a defect that is reconstructed using musculocutaneous flaps with or without a mesh. We report the case of a 24-year-old man who presented at the surgical outpatient clinic with 7 months history of persistent left sided chest pain minimally relieved by analgesics, 5 months of cough and worsening dyspnoea, and 3 months history of anterior chest swelling on the left side of the manubrium. Following evaluation and investigations, the tumor was excised and the residual defect closed with methylmetacrylate sandwiched between two prolene meshes and overlaid with both pectoralis major muscles. The histology of the excised mass revealed MPNST. He made an uneventful postoperative recovery, but died barely 3 months later from widespread pulmonary metastases. A review of the literature revealed that such tumors hardly ever reach such large-size as in our case.

Key words: Chest wall, reconstruction, tumor

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Introduction

Primary chest wall tumors are uncommon and constitute 0.2-2% of all tumors.^[1] Metastatic tumors and tumors of local extension are more common. The actual incidence of malignant peripheral nerve sheath tumor (MPNST) of the chest wall is not stated in the literature, but the incidence in the general population is 0.0001%.^[2] The risk is approximately 4600 times higher in patients with type I neurofibromatosis and 3-13% of them will finally develop MPNST, usually after latent periods of 10-20 years.^[2] This tumor previously known as malignant schwannoma, neurogenic sarcoma, and neurofibrosarcoma arise from embryonic neural crest cells, which normally constitute ganglia, paraganglionic, and parasympathetic systems. Clinically, these tumors are aggressive, locally invasive, and highly metastatic.^[3] Excision of giant chest wall tumor leaves a defect that is reconstructed

using musculocutaneous flaps with or without a mesh. A review of the literature revealed that such tumors hardly ever reach such large-size as in our case.

Case Report

Our patient was a 24-year-old man who presented at the surgical outpatient clinic with 7 months history of persistent left sided chest pain minimally relieved by analgesics and, which greatly interfered with his sleep. He also presented with 5 months history of cough and worsening dyspnoea and 3 months history of anterior chest swelling on the left side of the manubrium. There was no preceding history of trauma, fever,

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drenching night sweats, contact with a patient with chronic cough, hemoptysis, orthopnoea, and paroxysmal nocturnal dyspnoea or body swellings. He had no significant premorbid medical history. At initial presentation, he had a performance score of 90% (Karnofsky). The essential finding was that of a manubrial lump more to the left with normal overlying skin [Figure 1]. It was neither tender nor warm and was hard in consistency. It measured 8 cm × 12 cm in widest dimensions. The regional and other peripheral lymph nodes were not enlarged. The intensity of the breath sound was only slightly diminished on the left side. An initial impression of a chest wall tumor possibly a chondrosarcoma was entertained. Having visited several hospitals, he came with a chest X-ray which showed a large radio-opaque mass in the antero-superior mediastinum. It took him 2 months to get a chest computed tomography scan [Figure 2] and another 4 months to get money for surgery by which time the tumor was exceedingly large and he was now dyspnoeic at rest. Because an initial Tru-cut needle biopsy was not representative, we decided to go ahead with surgical excision. At surgery, Via a T-shaped sternal incision [Figure 6] the tumor was found to have invaded the whole of the upper third of the manubrio-sternum and having a dumb-bell shape. The outer and smaller portion (manubriosternal) measured 8 cm × 12 cm × 14 cm while the larger half wholly intrathoracic portion (left side) measured 18 cm × 24 cm × 30 cm in widest dimensions and weighing 5.4 kg [Figure 3]. The tumor was excised in continuum with the medial ends of the clavicles, the upper half of the manubrio-sternum and the upper four costal cartilages. Total left parietal and visceral pleurectomy was achieved. The pericardium was not invaded and mediastinal lymph nodes were not enlarged. The excision was thought to be complete as no macroscopic tumor was left behind. The anterior sternal defect created was covered with methylmetacrylate sandwiched between two 15 cm × 15 cm prolene meshes [Figure 4]. Both pectoralis major muscles were mobilized and used to cover the methylmetacrylate/prolene sandwich. He made an uneventful recovery postoperatively. The histology report revealed MPNST with incomplete resection margin [Figure 5]. He was discharged 2 weeks after surgery, was seen in the outpatient clinic 2 weeks thereafter discharge. By the second visit which was 8 weeks after surgery he had a recurrence at the right root of the neck. The chest X-ray then revealed widespread pulmonary metastases. He was readmitted 2 weeks later, barely 3 months postoperative and died within 4 days of readmission.

Discussion

The treatment of large chest wall tumors was limited for many years until 1898 when Parham described the first thoracic resection of a chest neoplasm.^[4] Initially, the risks of surgery were related to pneumothorax and respiratory failures.^[5] Advances in anesthesia and controlled airway ventilation with positive pressure allowed chest wall resections to be done safely. Early attempts at reconstruction included the use



Figure 1: Preoperative view showing tumor penetrating the chest wall (manubrio-sternum)



Figure 2: Computed tomography scanogram of chest showing tumor occupying most of the left hemithorax



Figure 3: Excised tumor measuring 18 cm × 24 cm × 30 cm

of fascia lata and rib grafts.^[6] As early as 1960, Graham and Usher introduced the use of prosthetic materials when they described the use of marlex mesh to repair defects of the chest wall.^[5] A combination of prosthetic materials and rotational

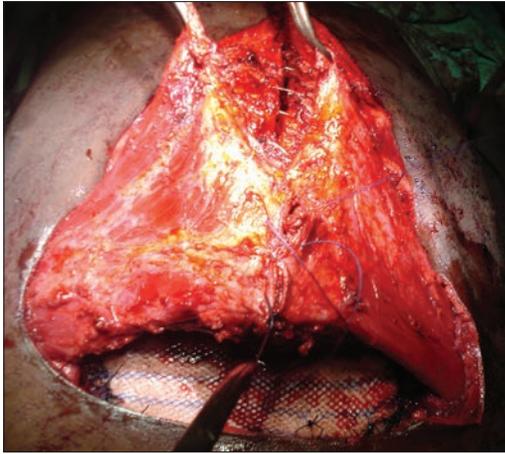


Figure 4: Both pectoralis major muscles been sewn together over the sandwiched mesh (white with blue stripes)



Figure 6: Patient sitting up on bed in intensive care unit postoperative revealing T-shaped incision used to excise tumor

flaps, with improvement in mechanical ventilation provide good cosmetic and functional results and short hospital stays. Latissimus dorsi myocutaneous flap is the most used method for extensive soft-tissue defects in chest wall reconstruction, but it is best suited for lateral defects.^[5] For our patient, we used both pectoralis muscles because of their proximity to the midline.^[7] Chest wall reconstruction in our environment is a challenge because of late presentation as in our patient. Other challenges include availability of the prostheses (mesh and methylmetacrylate). The methylmetacrylate restores the rigidity of the chest wall while the mesh provides a scaffold for fibrous tissue infiltration during healing. Another challenge is affordability of these prostheses even where there is the expertise; this featured prominently in this patient as it took him over 6 months to source funds for his surgery (enough time for the tumor to more than double in size and metastasize). The sum of these challenges serves to limit the amount of work done by thoracic surgeons and therefore, the availability of literature in this regard (particularly in our locality). To achieve a cure as much as possible, about 5 cm (King *et al* had recommended 4 cm for aggressive tumors)^[8] margin was given during resection of the tumor. Total left parietal pleurectomy (as the tumor was

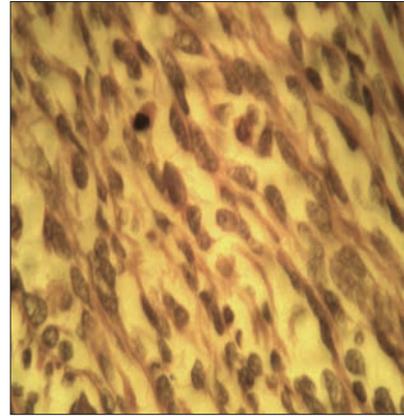


Figure 5: Excised tumor measuring 18 cm × 24 cm × 30 cm Hematoxyline and Eosin (x 400). Sections show fascicles of spindle cell with serpentine dark nuclei, fibrillary cytoplasm and mitotic figures. The tumour is collagen rich on special stain (Mason's Trichrome)

adhered to the upper lateral and anterior left parietal pleura), and bilateral medial third claviclectomy was performed. This unfortunately proved to be inadequate because the tumor disseminates hematogenously like most sarcomas. Therefore, while the patient had an excellent postoperative recovery, he died barely 3 months later.

Conclusion

Chest wall reconstruction poses a formidable challenge in our environment due to late presentation and therefore, very advance diseases. Affordability and availability of the prostheses also serve to limit the extent of work done by even the very experienced thoracic surgeons.

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