Solving the Mystery of Giant Intrathoracic Mass

Karmacharya RM, Singh A, Devbhandari M, Napit D, Ghimire K, Mahat C, Vaidya S

Department of Surgery (CardioThoracic and Vascular),

Dhulikhel Hospital, Kathmandu University Hospital,

Dhulikhek, Kavre, Nepal.

Corresponding Author

Robin Man Karmacharya

Department of Surgery (CardioThoracic and Vascular),

Dhulikhel Hospital, Kathmandu University Hospital,

Dhulikhek, Kavre, Nepal.

E-mail: reachrobin773@hotmail.com

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ABSTRACT

Intrathoracic schwannoma are highly vascular nerve sheath benign tumors arising from neural crest derived schwann cells of the intercostal nerves. Common clinical presentation is palpable mass but in our case patient presented with shortness of breath which is rare presentation in Schwannoma. Imaging studies of the patient showed the lesion in left lung, however surgical finding showed mass to arise from chest wall and it was confirmed to be schwannoma by histopathological examination.

KEY WORDS

Chest wall mass, Intrathoracic mass, Schwannoma

INTRODUCTION

Chest wall tumors are tumor present around the thoracic cavity and can originate from any soft tissue or bony structure.[1] They comprise about 5% of all the thoracic malignancies.[1] Primary chest wall tumors accounts only for 1-2% with schwannomas being a subset.[2] Although the patient can be asymptomatic in about 20% cases, they can present with painful, rapidly growing, palpable masses.[1] In some cases there is obstructive symptoms due to tumor compressing the lungs, trachea or bronchi.[1] Chest wall tumors can be primary or metastatic. Among them about 50% of chest wall tumors are malignant.[1] Primary chest wall tumors can be categorized into muscular, vascular, fibrous and fibrohistiocytic, peripheral nerve, osseous and cartilaginous, adipose, hematologic and cutaneous types.[3] A complete history, proper clinical examination and a plain chest x-ray, aided by other radiographic diagnostic modalities like computed tomography (CT), magnetic resonance imaging (MRI) and positron emission tomography (PET), leads to the diagonis of suspected chest wall tumors. In majority of cases, histological examination is also required for complete diagnosis.[1]

Surgical resection of tumor has been the demonstrated as the most effective method of treatment for chest wall tumors.[4] Furthermore, combination of surgery, radiation therapy and chemotherapy have improved five year survival for localized disease to about 80%.[5] Complete surgical resection with proper reconstruction helps to bring the best possible outcome for patients with malignant and metastatic tumors.[1]

If multiple ribs are to be resected, it can be reconstructed with the help of single or double layered polypropylene mesh contoured and strengthened with bone cement.[6]

CASE REPORT

A 55 years female presented to surgery OPD with the complains of shortness of breath for one month which was insidious on onset, gradually progressive and was present even at rest but not associated with orthopnea and paroxysmal nocturnal dyspnea. She also had loss of weight and loss of appetite for 1 month. However she had no fever, chest pain, cough, hemoptysis. General physical

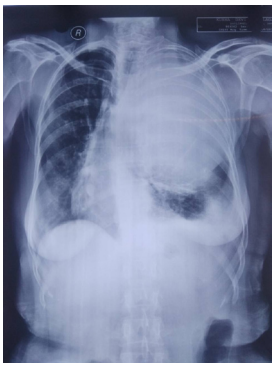

Figure 1. Giant intrathoracic mass

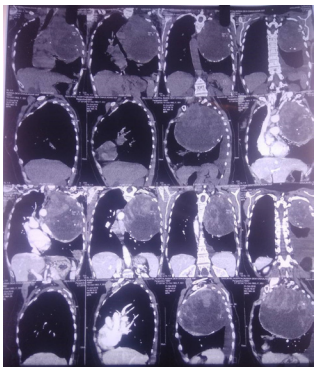

Figure 2. Giant intrathoracic mass

examination was apparently normal. On auscultation of chest, decreased air entry was noted over the left side of chest. Her chest X-ray showed a homogenous opacity occupying almost whole left hemithorax with normally located trachea (Fig. 1) CECT chest was done which revealed a well defined heterogeneously enhancing mass lesion in the left lung (containing enhancing solid component, non-enhancing component, likely necrosis, with peripheral calcification), likely to be neoplastic lesion shown in figure 2. Another well defined enhancing lesion was also present in the left lower lobe, lateral basal segment, likely to be metastatic lesion with associated left sided mild pleural effusion, collapse of the left lung and enlargement of left hilar and mediastinal lymph nodes

She underwent left thoracotomy where a mass 25*25*20 cm with partly cystic content arising from left fifth intercostal foramina related with intercostal nerve space occupying almost the whole of left hemithorax with dense adhesion with adjacent lung, subclavian artery, mediastinum and chest wall was found and the mass was excised and was sent for histopathological examination which later came out to be schwannomaHer post operative stay was uneventful and was discharged on 7th postoperative day.

DISCUSSION

Neurogenic tumors constitute the most common type of mediastinal tumor and account for the majority of neoplasms of the posterior mediastinum.[7] These originate from the cells of the nerve sheath or from the ganglionic cells of the spinal ganglia and of the autonomic, paraganglionic and parasympathetic systems.[8]

Among intrathoracic neurogenic tumors almost 90% are found in mediastinum with only 10% found in peripheral nerve fibres, either from intercostal or pleural nerve.[7]

In a study performed in New york hospital among 149 patients with intrathoracic neurogenic tumors, 72 had benign schwannomas, 10 malignant schwannomas, 17 neurofibromas, 24 ganglioneuromas, 9 ganglioneuroblastomas, 4 neuroblastomas, 9 primitive neuroectodermal tumors, and 4 paragangliomas.[12]

Schwannomas (also known as neurilemmomas or neurinomas) are highly vascular nerve sheath benign tumors arising from neural crest derived schwann cells.[7]

Histologically, schwann cells are arranged in a highly cellular distribution (Antoni A) or in a loose myxoid component (Antoni B) or both types in a single schwannoma. Epineurium contains both schwannoma and affected nerve in the form of true capsule with schwannoma being eccentric to the involved nerve. Thoracic schwannoma are usually marginated, spherical and lobulated or dumbbell shaped, grossly.[9]

In 98% of the cases, these tumors are benign, encapsulated and well-delimited, however cases have been described in which these tumors were aggressive and locally invasive. It has also been reported that cases of malignant schwannomas has tendency to relapse and metastasize.[10]

Schwannomas can be categorized microscopically as conventional, cellular, plexiform, and melanotic. IHC which is strongly positive for S-100 should be done for the diagnostic test.[11] However, IHC was not done in our case as it was proven by FNAC.

Whenever chest wall tumor is suspected, a proper history, clinical examination and a plain chest X-ray, should be taken which is further aided by computed tomography (CT), magnetic resonance imaging (MRI) or positron emission tomography (PET) scan.[13] Histological evaluation must be done as radiological findings may not be adequate for complete diagnosis.[1] Most of the chest wall tumors, including schwannoma present as palpable mass, however in our study patient presented with shortness of breath, weight loss and loss of appetite.[14] Contrast enhanced CT scans of schwannoma usually demonstrate homogenous mass if lesion is small but if lesion is large with cystic or necrotic changes then heterogenous mass is demonstrated, arising from extrapulmonary site.[9,11] The CT scan of our patient also showed similar heterogenous mass, however mass is reported to present in left lung but operative findings demonstrated mass to arise from chest wall.

Complete resection of the tumor is the treatment of choice for benign and malignant thoracic neurogenic tumors. Resection can avoid local invasion, facilitate differential histopathological diagnosis to determine other treatment options, and also help to prevent malignant degeneration.[12] Resection can be done either by videothoracoscopy or open thoracotomy. Thoracoscopic surgery are less invasive and hence, the preferred method of resection.[10] Thoracotomy is preferred for masses in middle and posterior mediastinum and it is also viable for mass in anterior mediastinum which does not cross the midline being contained within one hemithorax whereas indication for Video-assisted thoracoscopic surgery (VATS) are : biopsy to exclude malignancy, relief of compressive symptoms, prevent extension of a tumor into the spinal foramen, and to prevent malignancy.[15]

Postoperative radiation therapy or occasionally postoperative chemotherapy is required in most of the malignant tumors where complete resection cannot be done.[16]

In our case she underwent thoracotomy with resection of the tumor as suggested the treatment of choice by previous literatures. From imaging studies and after surgical findings the mass was suspected to be schwannoma which is finally confirmed by histopathology.

A chest wall schwannoma is a peripheral nerve sheath tumor arising from schwann cells. Clinical findings presenting as palpable mass, aided by radiological investigations helps to suspect the tumor, however histological examination provides final diagnosis. Complete resection of tumor is the treatment of choice.

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