CASE REPORT

Thoracic sarcoma case series in Hospital Kuala Lumpur, Malaysia

Yurkdes Aran Sittampalam, MBBS, Narasimman Sathiamurthy, MMED (Surgery)

Thoracic Surgical Unit, Department of General Surgery, Hospital Kuala Lumpur, Kuala Lumpur, Malaysia

SUMMARY

Thoracic sarcomas (TS) are rare neoplasms that may present with non-specific pulmonary symptoms. Chest radiographs and computed tomography of the thorax are the investigations of choice, while pre-operative minimally invasive biopsies may not be entirely helpful with the diagnosis. The commonly identified variants of thoracic sarcomas include liposarcomas, leiomyosarcomas and synovial sarcomas. Complete excision of the tumour with clear margins remain the sole definite form of management and there is limited role with adjuvant chemotherapy and radiotherapy. In the present case series, we explore our experience with three patients who presented with TS and the management decisions that was made.

INTRODUCTION

Thoracic sarcomas (TS) are seldom encountered even in the busiest thoracic centres. Sarcomas affecting the mediastinum, lung, pleura and even the thoracic wall are rare. The more commonly identified types of TS include leiomyosarcomas, fibrosarcomas and liposarcomas.[1] With regards to the broad spectrum of malignant thoracic tumours, they make up less than 1%.[1]

In this series of three cases, we discuss the presentation, management and outcome of patients who came to us predominantly with respiratory symptoms in which thoracic sarcomas were diagnosed.

Table I: Description of the three cases of thoracic sarcomas

	Patient 1	Patient 2	Patient 3
AGE	48	56	32
SYMPTOMS	Loss of appetite and loss of weight (lost 10kg). Cough, loss of appetite, loss of chest pain.	Cough, loss of appetite, loss of weight shortness of breath for 2 weeks.	Productive cough, loss of weight On and off haemoptysis, low grade fever for 2 months shortness of breath.
PHYSICAL EXAMINATIONS	Comfortable under room air, not tachypnoeic. Lungs: reduced air entry over left mid-zone to lower zone	Mildly tachypnoeic requiring nasal prong. Lungs: Reduced air entry over right lower zone with crepitations and increased vocal resonance.	Comfortable under room air, not tachypnoeic Lungs: Reduced air entry on left, dull on percussion
RADIOLOGY	Chest radiograph (Figure 1a): no cardiomegaly with left lower lobe collapse CT Thorax (Figure 1d): large posterior mediastinal mass with fatty content. (11.5cmx12.5cmx13.6cm).	Chest radiograph (Figure 1b): Right lower lobe collapse CT Thorax (Figure 1e): Large mediastinal mass with multiple echogenic areas (suggesting bleeding from mass).	Chest radiograph (Figure 1c): left hemithorax mass with tracheal and mediastinal shift CT Thorax (Figure 1f): Large heterogeneous mass occupying left hemithorax with mediastinal displacement.
INTERVENTION	Left thoracotomy and excision of mass.	Right mini thoracotomy and open biopsy.	Left thoracotomy, excision of mass and left pneumonectomy.
HISTOLOGY	HPE: Mixed type liposarcoma with dedifferentiation into leiomyosarcoma, clear margins.	HPE: monophasic synovial sarcoma.	HPE: biphasic synovial sarcoma, clear margins.
OUTCOME	Post op 3 years, no adjuvant treatment. No recurrence.	Patient died post-operatively.	Post op 2 years. No adjuvant treatment.

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Corresponding Author: Dr Narasimman Sathiamurthy
Email: drnara@hotmail.com

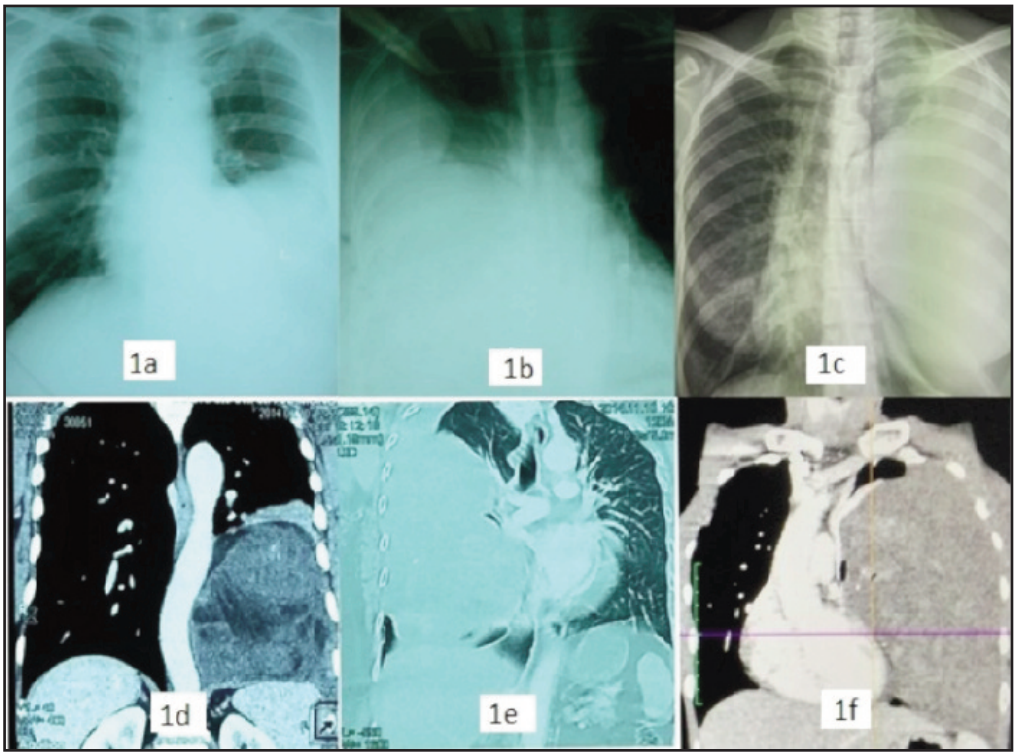

Fig. 1: Chest X-ray (1a, 1b, 1c) and Contrast Enhanced Computed Tomography (1d, 1e, 1f) of the thorax in sagittal view of the three patients with thoracic sarcoma.

DISCUSSION

The complexity that comes with diagnosing a thoracic sarcoma is further compounded by the fact that the presenting symptoms and clinical features can be vague and non-specific. In general, symptoms are related to the size of the sarcoma as well as its relations to adjacent structures. Patients often present with dyspnoea, arrhythmias, gastroesophageal reflux disease, dysphagia, chest discomfort or persistent cough.[1] While there are no specific signs to look out for, reduced air entry on auscultation or dullness on percussion provides clues as to the site of pathology that warrant further investigation.

Chest radiograph is a useful first-line of investigation in identifying an intrathoracic mass and this provides a rough idea regarding the size, location as well as effects on other structures subsequently visualised on the radiograph. However, computed tomography scans are invaluable in helping with diagnosis and preoperative planning. Among which, a clinician should strongly suspect a thoracic sarcoma when faced with a large mass (measuring more than 7cm), and at the same time illustrating well-defined, smooth or lobulated margins.[1] In addition, vessels may be identified within the mass, with an associated pleural effusion in the absence of significant lymphadenopathy.[1]

Establishing a diagnosis by tissue biopsy may be attempted preoperatively, however may be inaccurate.[2] Core needle biopsies, while it is superior to fine needle aspiration cytology, may still provide inadequate tissue samples for a detailed pathological evaluation when compared to an excision biopsy.[2] Furthermore, the biopsy tract must be carefully planned in such a way that it is included with the tissues that will later be involved with the excision of the tumour and prevent biopsy tract recurrence.[2]

Among the variety of soft tissue sarcomas, liposarcomas are unusually found in the intrathoracic region, accounting for only 2.7%.[3] In contrast, primary pulmonary leiomyosarcomas are far less common, making up less than 0.5% of all pulmonary and thoracic neoplasms.[4] Thoracic synovial-type sarcomas are also rarely encountered and are made up of two types of cells, epithelial cells and fibrosarcoma-like spindle cells which gives rise to the classification of monophasic, biphasic or the poorly differentiated types.[5]

Overall, the long term survival of patients with liposarcomas are generally more favourable compared to other subtypes.[1] Synovial sarcomas in other areas have a 5-year survival rate of about 63-75%, however, this situation changes when considering patients with advanced synovial sarcomas, with one study demonstrating a median survival rate of 22 months, similar to what was encountered in our cases.[5]

A surgical approach excising the lesion with clean margins forms the primary aim of treatment.[1] Neoadjuvant or adjuvant chemoradiotherapy is of little value in sarcoma with poor tumour response.[3] Research into the prevention of local recurrence with adjuvant radiotherapy is ongoing, and few studies have established that although adjuvant external-beam radiotherapy has a limited role in preventing

recurrence, it may not be necessary in patients with established low risk of recurrence, and this needing further research.[3]

CONCLUSION

TS are rarely seen in clinical practice, and as for most rare conditions in medicine, not much evidence is available to provide beneficial clinical guidelines for management. However, it is worth noting that upon identification of a suspicious mass on imaging, a percutaneously obtained biopsy should not be attempted prior to a referral to a thoracic surgical unit as it may not yield enough information to assist with the management and at the same time risks upstaging the tumour. As such, with the limited number of reports available, the way forward with resectable lesions remains excision, with or without chemo or radiotherapy.

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