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CASE REPORT Open Access

Solitary fibrous tumor of pleura presenting as respiratory failure: unusual presentation of a rare tumor

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Abstract

Solitary fibrous tumors of the pleura (SFTPs) are rare primary pleural tumors derived from sub-mesothelial mesenchymal tissue that are mostly incidentally diagnosed in asymptomatic subjects. Symptomatic SFTPs usually present with dry cough and chest pain. We report the case of a 55-year-old lady with a 9-month history of shortness of breath, right-sided chest pain, and heaviness, who presented to our institute with hypercapnic respiratory failure and later required invasive mechanical ventilation. Subsequent radiological and histopathological investigations led to the diagnosis of a benign SFTP occupying almost the entire right hemithorax. She was treated with complete surgical resection by thoracotomy resulting in immediate resumption of adequate ventilation in the postoperative period. We emphasize the importance of large pleural tumors as a rare but reversible cause of hypercapnic respiratory failure.

Introduction

SFTPs are rare primary pleural tumors arising from mesenchymal tissue beneath the mesothelial layer of the pleura. They were first described in 1931 by Klemperer and Rabin as a distinct pathological entity, and only about 2000 cases have been described in the medical literature [1–3]. SFTPs are mostly benign but sometimes they behave aggressively and recur after complete resection; however, no definitive criteria exist to differentiate the two subtypes [4]. These tumors usually grow to a large size before causing symptoms and are rarely reported to cause respiratory failure [5].

Case description

A 55-year-old non-smoker, homemaker woman presented with acutely worsening breathlessness for 3 days. She had a history of progressive shortness of breath and dull, poorly localized right-sided chest pain for the past 9 months. She also reported a loss of appetite and an undocumented weight loss. There was no history of any seasonal symptoms, hemoptysis, chronic cough or expectoration, syncope, and joint pains. General examination revealed a tachypnoeic patient in severe respiratory distress for which non-invasive ventilation (NIV) was initiated. Pallor, icterus, cyanosis, clubbing, and edema were absent. Vital parameters at presentation were pulse rate of 106/min, respiratory rate of 30/min, blood pressure 110/60 mmHg and room air oxygen saturation of 87%, with a GCS (Glasgow coma scale) of 15/15. Her BMI was recorded to be 19.6 kg/m^2. Respiratory examination showed fullness of right hemithorax and left-ward shift of mediastinum. Respiratory movements were reduced on the entire right hemithorax compared to the left. There was reduced vocal fremitus, dull note on percussion and absent breath sounds, on the entire right hemithorax. Normal vesicular breath sounds were heard on the left side without any adventitious sounds.

Routine laboratory parameters were normal, but chest X-ray (CXR) showed a large homogenous opacity

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occupying almost the entire right hemithorax (Fig. 1A) with contralateral mediastinal shift. Subsequent ultrasonography (USG) of the thorax revealed a large, heterogeneous, hypoechoic, vascular mass lesion, raising the suspicion of a pleural based tumor. Arterial blood gas (ABG) at presentation was suggestive of an acute-on-chronic hypercapnic respiratory failure (pH-7.20, $PaCO_2$-108 mmHg, PaO_2-58 mmHg, HCO_3-40 mmol/L).

In view of worsening hypercapnia despite the NIV trial, the patient was intubated. She was ventilated using a low tidal volume and high respiratory rate strategy and was extubated 36 h later. Post-extubation, she required intermittent NIV support for persistent hypercapnia and increased work of breathing. Contrast-enhanced computed tomography (CECT) of the thorax done prior to extubation revealed a 14 × 22 × 26 cm mildly enhancing lesion with tissue radiodensity of 30–35 HU suggesting a soft tissue mass, with foci of coarse calcification in the right hemithorax, associated with diaphragmatic flattening and contralateral mediastinal shift (Fig. 1B). Scans of the abdomen and brain were unremarkable. Infective work-up of the endotracheal aspirate inclusive of gram stain, aerobic culture, acid fast bacilli smear, and KOH mount for fungal elements was negative, and there was no clinical or microbiological evidence of any other infective process elsewhere. A USG-guided core biopsy was done from the mass and sent for histopathological examination (HPE) and immunohistochemistry (IHC) study, which indicated a mesenchymal tumor, likely benign SFTP (Fig. 2A). IHC stains revealed CD34, diffuse strong positivity; SMA, patchy cytoplasmic positivity; WT-1, negative; and Ki67, < 1%. Preoperative pulmonary function tests could not be performed due to persistent NIV requirements of the patient post-extubation.

The patient subsequently underwent a right posterolateral thoracotomy under the cardio-thoracic-vascular surgery (CTVS) team. An on-table check bronchoscopy was performed immediately prior to thoracotomy and it revealed only extrinsic compression of segmental openings of the right bronchial tree. At thoracotomy, a large, vascular encapsulated tumor was visualized upon entering the pleural cavity and it was resected en bloc. The mass was approximately 26 × 24 x 12 cm in size and weighed 3.6 kg (Fig. 2B). Following surgery, the patient was hemodynamically stable and was extubated the next day. She did not require any form of assisted ventilation postoperatively. CXR also showed significantly improved aeration on the right side. Detailed pathological evaluation of the resected specimen re-affirmed the diagnosis of benign SFTP. She was discharged on post-op day five with a normal ABG [pH-7.44, $PaCO_2$-42.2 mmHg, PaO_2-65.8 mmHg, HCO_3-28.1 mmol/L].

The patient initially followed up 2 weeks post-discharge and reported significant improvement in her symptoms. CXR at this time was equivocal (Fig. 2C). She remained asymptomatic at 3- and 6-month follow-up visits, had resumed her daily activities, and was advised to remain in yearly follow-up or when required.

Discussion

According to the WHO classification, SFTP is classified as a fibroblast/myofibroblast tumor. There are no known environmental or genetic predispositions [6]. The first description of SFTP was in 1767 by Lietaud et al. [1, 5]. Common age of presentation is in the sixth decade of life with no gender predilection [7]. Approximately half the patients are diagnosed incidentally [8]. Symptoms appear when the tumor has grown considerably

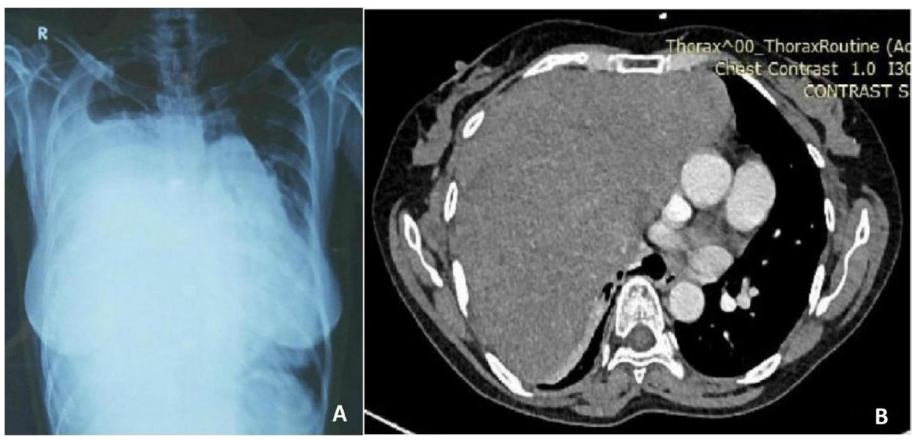

Fig. 1 **A** Preoperative CXR posteroanterior view showing a homogenous opacity occupying all lung zones of the right hemithorax with gross contralateral mediastinal shift. **B** Axial section of the CECT-thorax showing a large, well-defined, mildly enhancing soft tissue density lesion [14 × 22 × 26 cm] in the right hemithorax. Contralateral mediastinal shift due to the tumor is appreciable

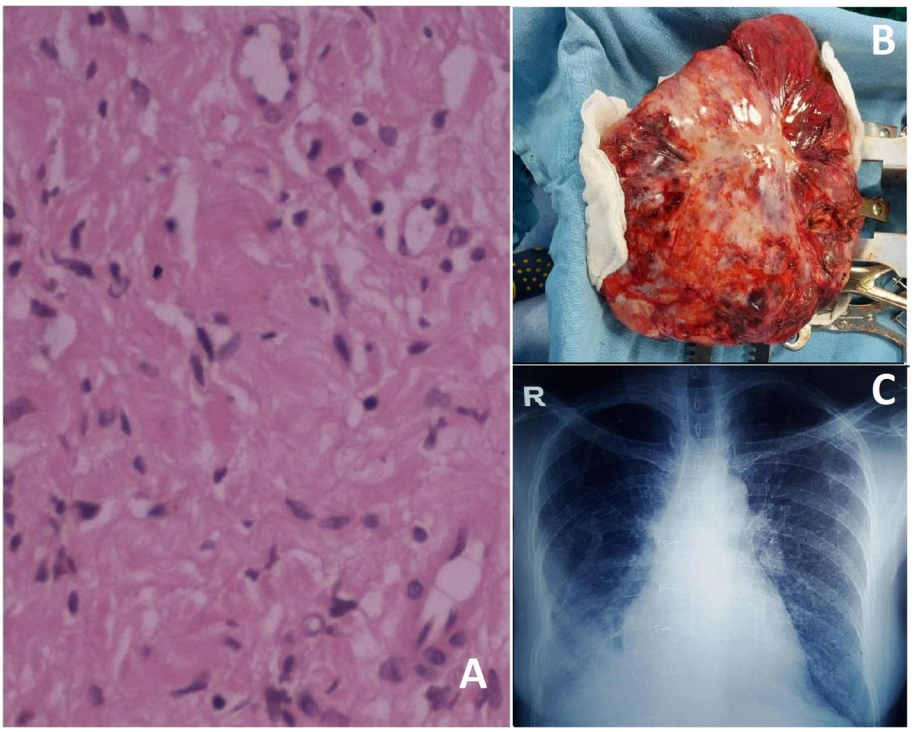

Fig. 2 **A** Photomicrograph showing tumor cells that are spindled along with thick bands of collagen and prominent hyalinised vessels (H&E × 400). **B** Gross appearance of the large 3.6 kg encapsulated mass measuring 26 × 24 x 12 cm removed by thoracotomy. **C** CXR posteroanterior view done at 2-weeks' follow-up post-discharge showing improved right-sided aeration and a centralized mediastinum with blunting of right costophrenic angle due to minimal reactionary fluid collection

and include dry cough, chest pain, chest heaviness, and rarely dyspnea [6].

A rare entity associated with SFTP is respiratory failure, especially the hypercapnic variety [6]. Abe et al. in their case series of three SFTP patients with respiratory failure reported that one of the cases required invasive mechanical ventilation; however, the type of respiratory failure was not indicated. In all three cases, SFTPs occupied almost the entire affected hemithorax [4]. Such large tumors result in significant alterations in respiratory mechanics and compliance. The decrease in functional lung volume which is proportional to the tumor size results in increased airway resistance. Thus, the patients need an increased driving pressure to generate a given tidal volume owing to the altered physiological circumstances, leading to dyspnea. The large-sized SFTPs also lead to a decreased transmural transpulmonary pressure gradient causing compromise of the ventilation/perfusion ratio and gas exchange, further contributing to dyspnea and respiratory failure [9].

The mainstay of treatment for SFTP is en bloc surgical resection. Recurrences have been documented to occur with the malignant subtype and require re-excision. SFTPs have a favorable prognosis with 5-year overall survival rates above 79% [3].

The dramatic improvement in symptoms and resolution of hypercapnia post-surgery implied that our patient's respiratory compromise was attributable to the extra-parenchymal restrictive ventilatory defect due to the SFTP. The normal-appearing lung parenchyma in the preoperative CT scan, normal neurological examination and brain imaging findings; the absence of any history of airway disease; absence of obesity; and patient not being on any chronic medications that might decrease respiratory drive and euthyroid status were used as supportive evidence to rule out any contributing factors to her hypercapnic state.

To conclude, we would like to emphasize the importance of bedside ultrasound prior to all pleural procedures as the presenting radiograph (Fig. 1A) of our patient mimicked that of a massive pleural effusion. We would also like to highlight that while the usual suspects of hypercapnic respiratory failure encountered by respiratory physicians are chronic and per se incurable pulmonary disorders, large pleural tumors like SFTPs may stand out as a rare but potentially correctable cause.

Conclusions

- Respiratory failure can occasionally result from worsening respiratory distress due to giant intrathoracic tumors.
- SFTPs, one such kind of a tumor, can be a rare but potentially reversible cause of hypercapnic respiratory failure.
- Extensive clinical and pathological workup is essential to facilitate their early and accurate diagnosis.
- Definitive treatment by complete surgical resection can offer symptom-free long-term survival in these patients.

Abbreviations

SFTP	Solitary fibrous tumor of pleura
NIV	Noninvasive ventilation
GCS	Glasgow coma score
CXR	Chest X-ray
USG	Ultrasonography
ABG	Arterial blood gas
CE-CT	Contrast-enhanced computed tomography
HPE	Histopathological examination
IHC	Immunohistochemistry
CTVS	Cardiothoracic vascular surgery

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Authors' contributions
Concept/design of the case report and acquisition of the data: Dr Avishek Layek. Revising the article critically for important intellectual content: Dr Michael Leonard Anthony; The case was operated and surgically managed by Dr Anshuman Darbari. Final approval of the version to be published and guarantor: Dr Mayank Mishra. The authors read and approved the final manuscript.

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Competing interests
The authors declare that they have no competing interests.

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