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Solitary Pulmonary tumour associated with clubbing: A case report

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Abstract
Digital clubbing is an antiquated and significant clinical sign in medicine. Despite the fact that clubbed fingers are generally asymptomatic, it frequently predicts the presence of some feared basic illnesses**.** Among the different types of thoracic malignancy, lung cancer is responsible for 80% cases of clubbing, whereas pleural tumors and other intrathoracic and mediastinal growth contribute to 10% and 5% cases, respectively. On the other hand, the prevalence of clubbing in lung cancer patient ranges from 5% to 15%.
Solitary fibrous tumours (SFTs), are relatively rare and most of SFTs originated from pleura, especially the visceral pleura, a few of which occurred outside pleura and rarely in the lung. The incidence of SFTP was less than 5% of all pleural tumours. The WHO classification defines a solitary fibrous tumour as a ubiquitous mesenchymal tumour probably derived from fibroblasts with a hemangiopericytoma-like branching vascular pattern. In initial stage they are mostly asymptomatic and picked up incidentally. Once the tumour enlarges, it causes pressure effects on surrounding structures and the patient becomes symptomatic. En-bloc surgical resection is treatment of choice.
We report a relatively rare case of a 57 year old female patient presents with symptoms of cough and clubbing and detailed history revealed that the clubbing was an acquired sign and was the main reason why she approached the physician and was diagnosed to have a solitary fibrous tumour of the pleura. The patient successfully underwent pre op embolization to decrease blood loss intra operatively followed by an en bloc surgical resection of the tumour via a right posterolateral thoracotomy. On follow up after 6 months she was asymptomatic with no recurrence had a fair resolution of her clubbing.

Keywords: Solitary fibrous tumour, clubbing, pre-op embolization, en-bloc surgical resection

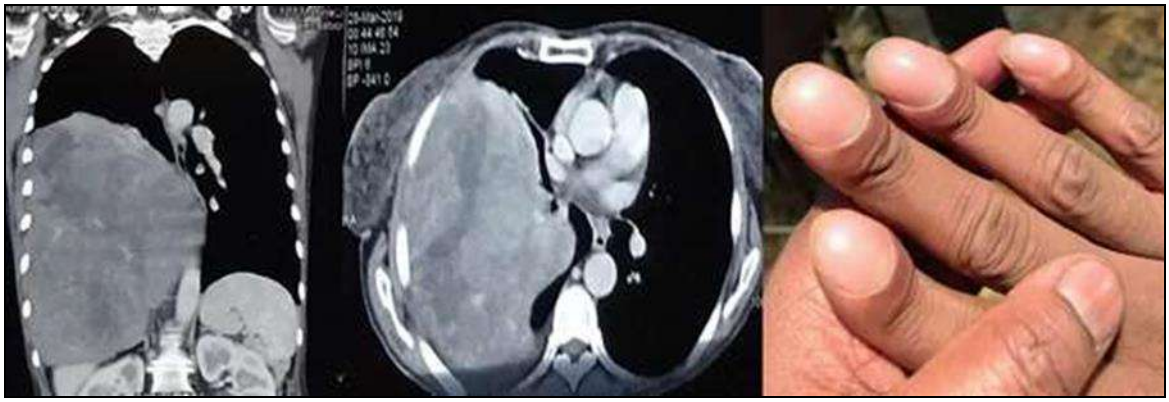
Introduction
Solitary fibrous tumours (SFTs) are relatively rare neoplasms. They account for < 5 % of all pleural tumours. Pleural and intrapulmonary SFTs affect men and women equally. Mean age at diagnosis is the sixth decade of life. Just more than half of patients are symptomatic with presenting complaints that include chest pain (25%), shortness of breath (15%), and/or cough (12%), attesting to the often large size of these tumours. Digital clubbing and hypertrophic pulmonary osteoarthropathy (Pierre-Marie-Bamberg syndrome) have been described in 10% to 20% of patients with either benign or malignant SFT of Pleura [1]. These clinical features usually resolve within 2 to 5 months or sometimes longer after removal of the tumor, but they may reappear with recurrence of the tumor [1]. The causes of digital clubbing and of hypertrophic pulmonary osteoarthropathy could be, respectively, an abnormal production of hepatocyte growth factor or an excessive release of hyaluronic acid by the tumor [2]. In less than 5% of patients, SFTP can also presents with paraneoplastic syndromes and can secrete insulin-like growth factor II, which causes refractory hypoglycemia (Doege-Potter syndrome) [3]. A high serum level of insulin-like growth factor II is typically associated with low levels of insulin and insulin-like growth factor I, which return to normal values within 3 to 4 days after resection of the tumor [4]. We report a relatively rare case of a 57 year old female patient presents with symptoms of coughing and clubbing since 2 months. Detailed history revealed that the clubbing was an acquired sign and was the main reason why she approached the physician. A X-ray chest and contrast CT scan of the thorax showed a large mass occupying almost the 70 % of the right hemithorax. A CT guided core biopsy was consistent with solitary fibrous tumour of the pleura. The patient successfully underwent embolization followed by an en bloc surgical resection of the tumour via a right-posterolateral thoracotomy.

The final histopathology was consistent with solitary fibrous tumour of the pleura.
On follow up, after 6 months, she was asymptomatic with no recurrence and had a fair resolution of her clubbing.

Case report
57 year old Female, with no co morbidities, presented with history of grade IV clubbing since 2 months and subsequent cough since 6 months. She had progressive shortness of breath, which worsened over the last few months. Her X-ray chest and subsequent CT scan thorax revealed a large enhancing lobulated pleural based mass in the right lower hemithorax measuring 17.3 cms x 16.1 cms x 22.5 cms. It was closely abutting right posterior hemi diaphragm.

The lesion was crossing the midline in the retro cardiac region and causing mass effect with loss of fat planes with the posterolateral pericardium.

Pre-operative embolization of tumour was done to reduce its vascularity and decrease blood loss intra operatively. Aortogram showed multiple arterial supply to the right chest wall tumour from the right internal mammary, D11 and D12 intercostal, branch from the right hepatic artery as well as a branch from the celiac artery. Super-selective cannulation of the feeder vessels and embolization done with 300um PVA particles, until stasis achieved. Post embolization aortogram showed complete absence of vascularity in the lesion.



(Large Solitary Fibrous tumour occupying nearly the entire right Hemithorax)

Fig Ia CT scan Thorax Coronal **Fig Ib** CT scan Thorax Axial **Fig Ic** Finger clubbing

Next day after post embolization she underwent right posterolateral thoracotomy with sixth rib resection and en bloc excision of the lesion.
A portion of the right lower lobe of the lung was firmly adherent to the mass and was inseparable from it. A wedge resection of right lower lobe lung was performed along with mass. After removal of the mass, the middle and right lower lobe of the lung showed good expansion. Patient was discharged on eighth post-operative day.

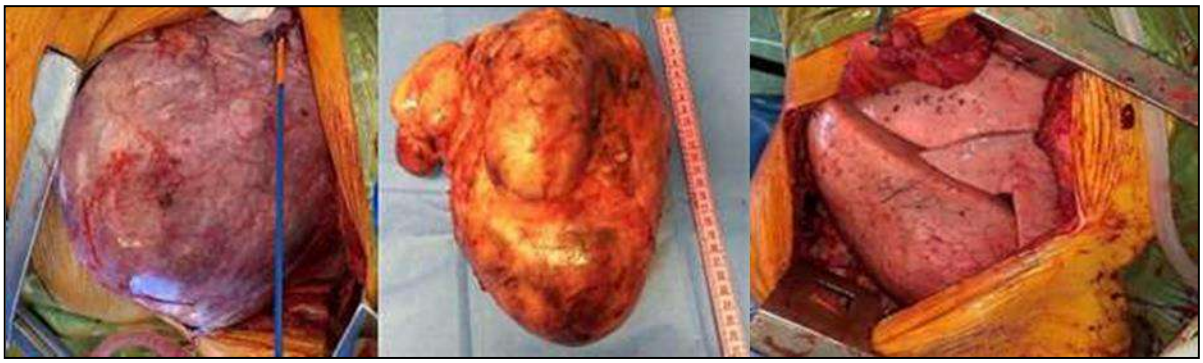


Fig IIa: Intraoperative view - Thoracotomy with Large Solitary fibrous tumour **Fig IIb:** Gross Specimen 22cm x17.5 cm x 13 cm **Fig IIc:** Complete Expansion Right Lung after removal of the mass

Final Histopathology report was consistent with solitary fibrous tumour, low to intermediate risk of malignant behaviour. The large encapsulated tumour mass measures 22 x 17.5 x 13cm. and weighs 2838gms.

Pre-operative embolization of tumour done followed by right standard posterolateral exploratory thoracotomy and 6th rib resection with wedge resection of lower lobe of right lung done.

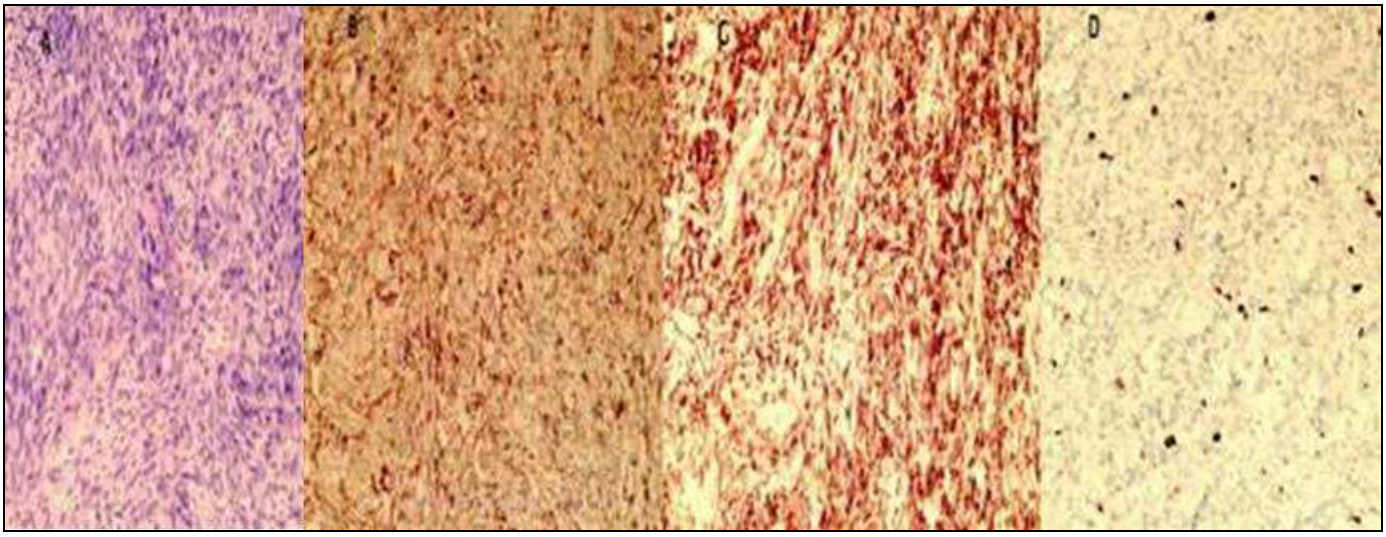


Fig III: IHC Tumour cells express (a) Bcl2, (b) B-catenin, (c) CD34 and (d) CD99 Ki-67 nuclear labelling index is 10%.

Discussion

Solitary fibrous tumor of pleura is symptomatic in about half of patients, including most commonly cough, chest pain, dyspnoea, and pulmonary osteoarthropathy. Digital clubbing and hypertrophic pulmonary osteoarthropathy (Pierre-Marie-Bamberg syndrome) have been described in 10% to 20% of patients with either benign or malignant SFT of Pleura [1]. Hypoglycaemia is an uncommon manifestation of solitary fibrous tumour(Doege-Potter syndrome) resulting from secretion of insulin-like growth factor II by neoplastic cells and is resolved with complete excision. On rare occasions, it is found together with a microscopically similar tumour in the peritoneum or retroperitoneum [5].

Grossly, the lesion is well circumscribed, firm, lobulated, and grey–white to yellow–white, with frequent whirling and fasciculation. The mean diameter is 6 cm, but some are very large, completely opacifying a hemithorax as we saw in our case. The majority are attached to the visceral (80%) or parietal pleura, within an interlobar fissure, or sometimes within the lung parenchyma without a pleural connection (intrapulmonary solitary fibrous tumour).

In the typical benign case, there is a tangled network of fibroblast-like cells, squeezed in between abundant collagen fibres, many of which have a keloid-like quality, resulting in what some have referred to as the pattern-less pattern of Stout. The proliferation of mesenchymal spindle cells separated by thick bands of keloid-type collagen is characteristic of this entity. Malignant solitary fibrous tumours are characterized by increased cellularity, atypia, mitotic activity, and necrosis.

It arises from non-committed mesenchymal cells present in the areolar tissue subjacent to the mesothelial lining. Ultra structural examination of these cells show fibroblast-like rather than mesothelial-like features, and by immunohistochemistry their filament profile includes vimentin and sometimes desmin but not keratin [6].

The tumour cells of this lesion stain strongly and consistently for CD34 and BCL2.Recurrent intrachromosomal rearrangements resulting in fusion of NAB2 and STAT6 on chromosome 12q have been identified as a unique driver mutation in solitary fibrous tumours. The fusion product is overexpressed in nearly all cases and can be detected using a commercially available antibody directed against STAT6 characterized by diffuse nuclear staining.

The vast majority of solitary fibrous tumours of the pleura are cured with complete surgical excision and tumour-free margins [7]. Pre-operative embolization for the very large tumours have an advantage. Besides decreasing the blood loss, the ischemia-induced perilesional oedema is another advantage of preoperative embolization, facilitating dissection of the tumour from adjacent structures [8].

A complete en bloc resection reduces the risk of recurrence. Relapse can present as recurrent intrathoracic or widely metastatic disease. In a scoring system intended to predict risk of recurrence, Tapias et al. proposed parietal (vs. visceral) origin, gross morphology, size (≥10 cm), hyper cellularity (i.e. crowded tumour cells with overlapping nuclei), necrosis or haemorrhage, and a high (≥4/10 hpf) mitotic rate **[9]**. However, long term follow up is necessary because there is a small but persistent risk of local recurrence.

Conclusion

Solitary fibrous tumour (SFT) is a rare mesenchymal tumour of the pleura and association with clubbing is even rarer. Digital clubbing and hypertrophic pulmonary osteoarthropathy have been described in 10% to 20% of patients with either benign or malignant SFT of Pleura .SFT arises from visceral pleural and projects into the pleural cavity. We report a rare case of SFT of the lung associated with clubbing which underwent an en bloc surgical excision of tumour. In the immediate post-operative period there was complete re expansion of the lung alleviating the patient symptoms completely. At six months follow up, the patient was completely asymptomatic and there was complete resolution of clubbing.

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