

## Case Report

# Pulmonary Schistosomiasis Presenting as a Solitary Fibrous Tumor

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**Abstract:** A 37 years old man presented with shortness of breath on mild exertion. Chest X-ray and CT-scan showed a large mass in the right chest. CT-guided fine needle biopsy was suggestive of solitary fibrous tumor of the pleura. Excision through right thoracotomy was done, and histopathology reported a solitary fibrous tumor with *Schistosoma* eggs. This case report may be the first in the English literature of a patient with pulmonary schistosomiasis presenting as a solitary fibrous tumor.

**Keywords:** Schistosomiasis, Pulmonary Schistosomiasis, Solitary Fibrous Tumor

## 1. Case Report

An Egyptian man, 37 years old, presented to thoracic surgery clinic complaining of shortness of breath on moderate exertion for 2 months. There was no history of cough, fever, or night sweating. He is an office worker, ex-smoker (stopped 9 years ago), with no other significant past medical history.

The vital signs were within normal except for diminished air entry over the right lower zone of the chest posteriorly, and all blood lab tests were normal, including WBCs count, which was normal except for significant eosinophilia (11.2%).

Chest X-ray and CT scan (Figures 1, 2) showed a large mass in the right lower chest attached to the chest wall. CT-guided fine needle biopsy was suggestive of solitary fibrous tumor of the pleura (SFTP). Immune-histochemistry (IHC) of the tumor was strongly positive for CD34, CD99 and *bc12*, and negative for Cytokeratins, TTF-1 and Calretinin, therefore the morphologic pattern and IHC profile was consistent with solitary fibrous tumor. The patient refused larger True-Cut biopsy. Thoracotomy and excision of the right chest mass was recommended based on the suggested diagnosis. However, the unusually high eosinophilia was unexplained pre-operatively.

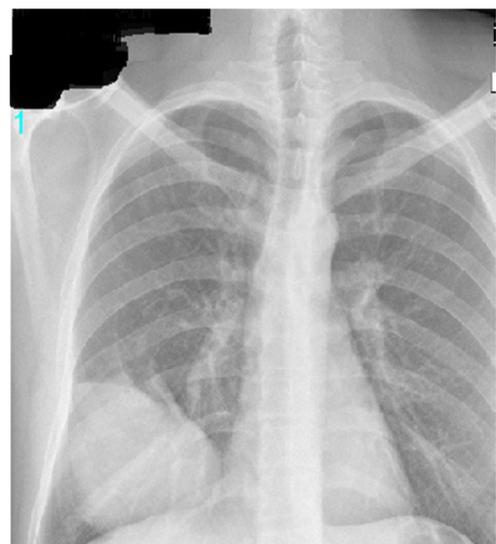
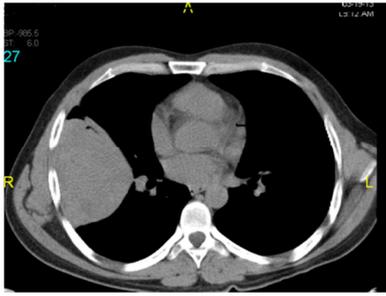


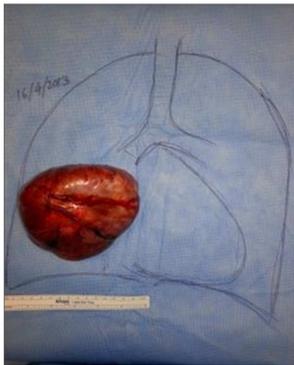
Figure 1. "Chest X-ray showing a large mass in the right lower zone".



**Figure 2.** "CT scan of the chest showing a mass about 12\*10 cm attached to the right chest wall".

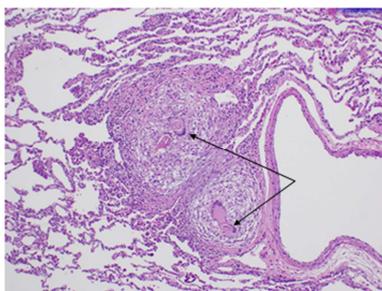
## 2. Hospital Course

Under general anesthesia, right posterolateral thoracotomy incision through the fifth intercostal space was done; there was no pleural effusion, or adhesions. A very large (12\*12\*10 cm) smooth but firm mass was readily identified. The mass appeared vascular and attached to the edge of the right upper lobe of the lung without any other adhesions or attachment to the parietal pleura or chest wall. The attachment of the mass to the lung was divided with GIA surgical stapler and the mass was completely removed (Figure 3).



**Figure 3.** "Composite photography showing the resected mass size and location".

The postoperative course was smooth and the patient was discharged from the hospital 2 days after surgery. Histopathology showed solid fibrous tumor with foci of granulomatous inflammation in the mass as well as in the surrounding lung parenchyma. *Schistosoma* ova were identified in the mass, and the pulmonary resection margin was free of the neoplasm (Figure 4).



**Figure 4.** Histopathology slide showing granulomatous reaction around *Schistosoma* ova (arrows) in the lung tissue.

In the follow up visits to the outpatient clinic, lab studies showed positive *Schistosoma* antibody titer at 1:256, and eosinophilia gradually decreased 3 months later to 0.7%. No ova of schistosoma were detected in the stool or the urine. He was given Praziquantel tablets 1200mg for one day in two divided doses.

## 3. Discussion

Solitary fibrous tumors of the pleura (SFTP) are ubiquitous rare spindle cell neoplasms, most commonly arising from the pleura. In 1931, Klemperer and Rabin first documented the occurrence of a distinctive localized pleural-based tumor, and proposed a submesothelial cell origin. Later on, based on tissue culture experiments, Stout and Murray claimed its derivation from mesothelial cells. This controversy is reflected in the variety of synonyms used for solitary fibrous tumors in the past including: localized fibrous tumor, localized fibrous mesothelioma, solitary fibrous mesothelioma, fibrous mesothelioma, subserosal fibroma and submesothelial fibroma. With the advent of immunohistochemistry, a fibroblastic origin, occasionally with myofibroblastic differentiation, is firmly established. This is further reinforced by the description of solitary fibrous tumors in extrathoracic sites devoid of mesothelial cells [1].

About 80% of SFTPs originate in the visceral pleura, while 20% arise from the parietal pleura. Although they are often very large tumors (up to 40 cm. in diameter), over half are asymptomatic at diagnosis. The size of the tumors can vary greatly between 1 - 36 cm (mean, 6 cm) in diameter. Many large tumors are pedunculated and pleural-based [2].

Non-specific radiologic features and inconclusive fine needle aspiration (FNA/FNB) make it difficult to reach preoperative diagnosis of SFTP of the pleura and lung. Diagnosis of SFTP is usually established with certainty only after surgery, as we found in our patient. Resection with clear margins is the most important prognostic factor [3].

Schistosomiasis is an endemic parasitic disease in more than 70 countries, including Egypt. It is estimated to infect 200 million worldwide [4]. It is documented that *Schistosoma haematobium* was endemic in Ancient Egypt; Ruffer in 1910 was the first to diagnose *S. haematobium* infections in mummies. He recovered calcified *Schistosoma* eggs from two Egyptian mummies of the 20<sup>th</sup> Dynasty [5].

The Egyptian Ministry of Health reported that from 1982 to 1992 the prevalence of *S. haematobium* declined from 15% to 1% in the Nile Delta, and from 13% to 3% in Upper Egypt, while the prevalence of *S. mansoni* declined from 40% to 20% in the Nile Delta region [6].

Pulmonary involvement in schistosomiasis is not very rare. There may be two forms: acute and chronic. The acute form usually occurs about 6 weeks after infection (Katayama syndrome), and seems to be due to an allergic manifestation to the presence of *Schistosoma* sp. worm or eggs. The chronic form is more commonly seen in endemic areas, and may cause pulmonary hypertension, cor pulmonale, granulomatous schistosomiasis and pulmonary arteriovenous fistulas.

Globalization and increased international travel makes it necessary for clinicians around the world to be aware of some "old" diseases from endemic areas of the globe [7].

Acute pulmonary schistosomiasis is thought to be an immune-mediated disease similar to other forms of eosinophilic pneumonia. This postulation is supported by the presence of eosinophilia, immune complexes and elevated IgE levels [4].

Chronic pulmonary schistosomiasis is a granulomatous reaction to eggs trapped in the pulmonary vasculature, which leads to pulmonary fibrosis, pulmonary hypertension and eventually cor pulmonale. Histologically it is a delayed-type hypersensitivity reaction. Patients with chronic pulmonary schistosomiasis have non-specific clinical and radiographic findings; the latter being more related to pulmonary hypertension than to infection. The majority of reported chronic pulmonary compromises are due to *S. mansoni*, with few case reports due to *S. hematobium* or *S. japonicum*. We could not determine the *Schistosoma* species in our patient. The type of chronic disease and typical end-organ involvement depends on the schistosoma species involved, with *S. mansoni* and *S. japonicum* classically affecting the hepatic portal system, and *S. hematobium* affecting the genitourinary system [8].

To date, Praziquantel is the only available effective treatment against all forms of schistosomiasis with few transient side effects. The use of this drug in the treatment of our patient was not associated with significant side effects. Prognosis depends on infesting species and end organs involved. In acute pulmonary schistosomiasis, complete recovery is expected. Although chronic lung involvement is usually associated with pulmonary fibrosis, pulmonary hypertension and eventually cor pulmonale [8], our patient did not have clinical signs of such complications.

After searching for: "Solitary Fibrous Tumor of the Pleura" and/or "Pulmonary Schistosomiasis" in PubMed database, we did not find any previously reported case showing a relationship between SFTP and pulmonary schistosomiasis.

#### 4. Conclusion

This case report is probably the first documented case in the English literature of a patient with pulmonary schistosomiasis presenting as solitary fibrous tumor. In this patient, *Schistosoma* ova were unexpectedly found histopathologically.

Pre-operative chest X-ray, CT scan and CT-guided FNA did not lead to the definite diagnosis.

Our patient is from Egypt, which is known to be an endemic area of schistosomiasis. He was followed up in outpatient clinic until his eosinophilia returned to normal range. We believe that Eosinophilia can be a good marker for the response to the treatment. *Schistosoma* antibody titer is not considered a good marker for treatment response as it remains high for long period of time even after surgical excision and pharmaceutical therapy [7]. In addition, we believe that surgical removal and anti-bilharzia medications are essential in the management plan.

N.B.: Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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