

# Symptomatic pulmonary sclerosing hemangioma: a rare case of a solitary pulmonary nodule in a woman of advanced age

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## Abstract

**Background:** Pulmonary sclerosing hemangioma (PSH) is a rare tumor that usually develops in middle-aged Asian women. PSH has four histological types (hemorrhagic, sclerotic, solid, and papillary) and often grows slowly in a lower lobe of the lung. Preoperative misdiagnosis frequently occurs because of the absence of specific clinical manifestations and imaging findings. Few reports have described PSH in women of advanced age.

**Case presentation:** A 75-year-old woman presented to our hospital in China with a 5-day history of productive cough and intermittent hemoptysis. Computed tomography indicated bronchiectasis and a large mass in the left inferior lobe of the lung. Treatment of the bronchiectasis provided no symptom relief. The hemoptysis resolved following left lower pulmonary lobectomy, and PSH was pathologically diagnosed following surgery. At the time of this writing (after 6 months of follow-up), the tumor had not recurred, no metastases had been detected, and close follow-up was ongoing.

**Conclusions:** Both bronchiectasis and PSH can cause hemoptysis. This case demonstrates that PSH should be included as a differential diagnosis of hemoptysis in women of advanced age. For patients with chronic hemoptysis, the diagnosis of PSH should be considered if the therapeutic effect of bronchiectasis is poor.

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## Keywords

Pulmonary sclerosing hemangioma, pathological diagnosis, hemoptysis, bronchiectasis, pulmonary lobectomy, misdiagnosis

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## Background

Pulmonary sclerosing hemangioma (PSH) was first described by Liebow and Hubbell in 1956.<sup>1</sup> It is a rare neoplasm that is most often encountered in middle-aged women.<sup>2</sup> PSH has four histological types (hemorrhagic, sclerotic, solid, and papillary)<sup>3</sup> and often grows slowly in a lower lobe of the lung.<sup>4</sup> Many reviews and reports have described PSH, but very few have described PSH in women of advanced age. The present case involved a 75-year-old woman in China who presented with a 5-day history of cough and sputum production with intermittent hemoptysis. The diagnosis of PSH was pathologically confirmed after surgical resection. This case is being reported to inform clinicians of the need to include PSH as a differential diagnosis of hemoptysis, particularly in older women, and to pursue pathological biopsy for a definitive diagnosis.

## Case presentation

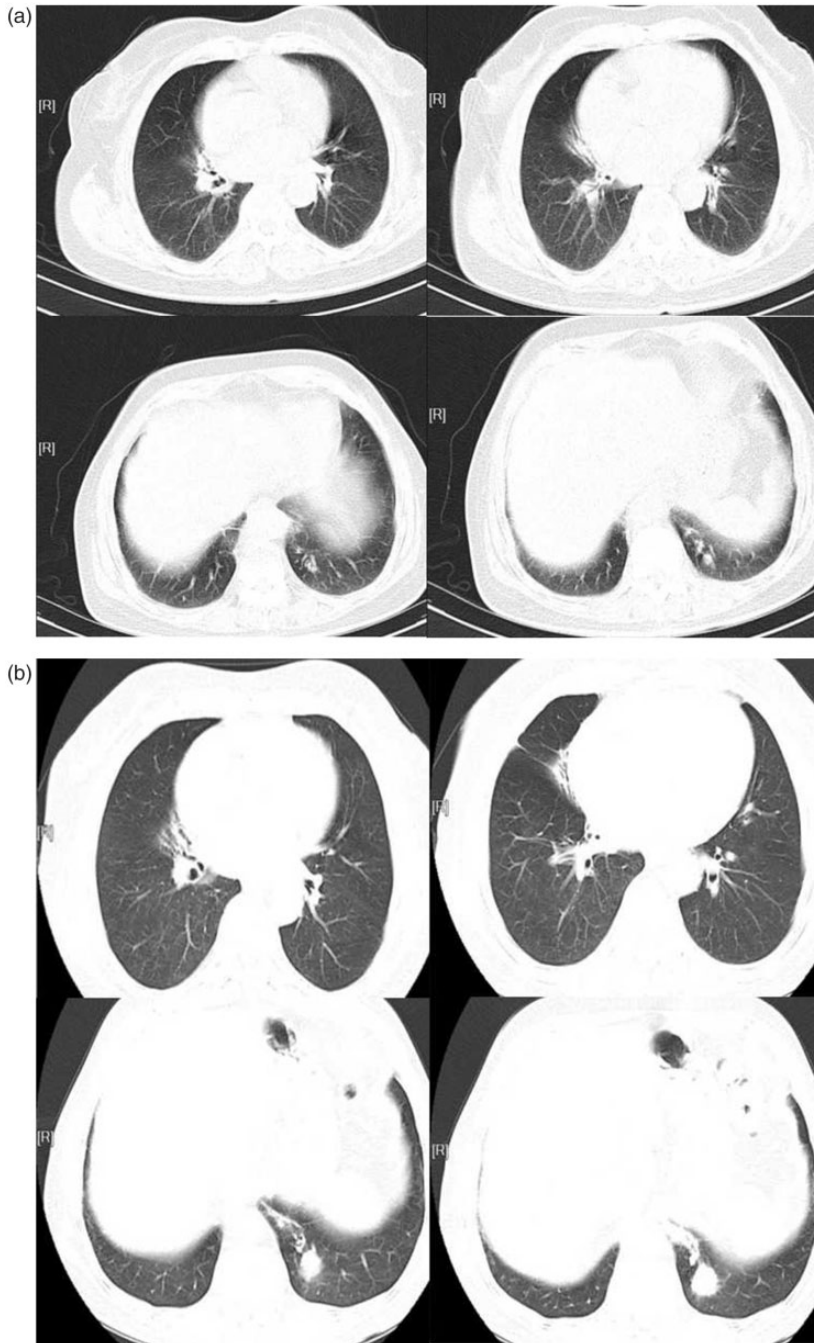
A 75-year-old woman presented to our hospital because of a 5-day history of cough and sputum production followed by intermittent hemoptysis. She was a nonsmoker. In 2015 (approximately 2 years before the current presentation), a 1-cm-diameter nodule had been detected in the left lower lung at our hospital (Figure 1(a)). The patient refused to undergo further diagnostic tests and treatment at that time. At the current presentation, a computed tomography (CT) scan

showed that the nodule had grown to 3 cm in diameter (Figure 1(b)). The nodule in the left lower lobe had become a well-circumscribed mass. However, the blood levels of tumor markers were not elevated. A CT scan also indicated bronchiectasis. Treatment for the bronchiectasis was given but provided no relief.

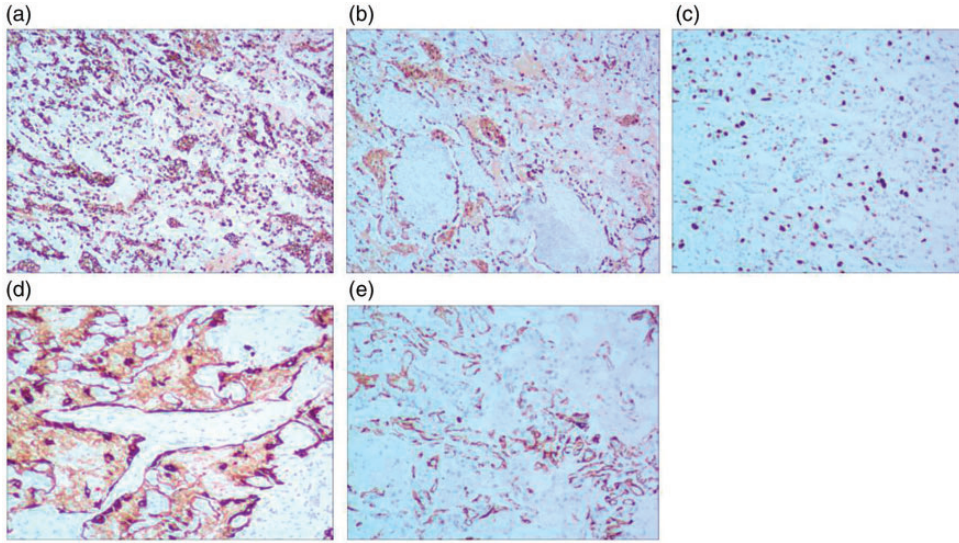
Because the patient was not cured by medical treatment, lobectomy was proposed by a surgeon. After surgical resection, the disease was pathologically diagnosed as PSH, which is a rare tumor, especially in older women. The findings in this case suggested that the hemoptysis had originated from this benign tumor.

The tumor was of the sclerosing type and included both cuboidal and polygonal cells. Immunohistochemical staining assisted in the diagnosis. The resected tissue was positive for cytokeratin 7 (Figure 2(d)). Additionally, thyroid transcription factor 1 (TTF-1) (Figure 2(a)) staining was observed in both superficial and polygonal cells. The superficial cells were also positive for napsin A (Figure 2(b)). The tumor was positive for Ki-67 (Figure 2(c)). The Ki-67 index was <1% in the superficial cells but higher in the polygonal cells. Some perivascular tumor cells were CD34-positive (Figure 2(e)). The final diagnosis was PSH.

The patient recovered fully after surgery and returned home with no symptoms. At the time of this writing (after 6 months of follow-up), the tumor had not recurred, no



**Figure I.** Comparison of computed tomography (CT) examinations. (a) Contrast-enhanced CT examination of the chest in 2015. (b) CT examination of the patient in 2017.



**Figure 2.** Pulmonary sclerosing hemangioma was confirmed by surgical resection and postoperative pathological diagnosis. (a) Both the cuboidal surface cells and polygonal cells showed thyroid transcription factor 1 staining. (b) Epithelial membrane antigen highlighted both the cuboidal cells and polygonal cells. (c) The polygonal cells showed higher Ki-67 expression than the surface-lining cells. (d) Cytokeratin staining was positive only in the surface cells, not in the stromal cells. (e) CD34 was only focally positive in the perivascular tumor cells.

metastases had been detected, and close follow-up was ongoing.

## Discussion

PSH was first described by Liebow and Hubbell in 1956.<sup>1</sup> It is a rare neoplasm that most often occurs in middle-aged women.<sup>2</sup> The male:female ratio is 1:5.<sup>2</sup> PSH is usually asymptomatic, but a few patients present with common respiratory symptoms.<sup>5</sup> Preoperative misdiagnosis frequently occurs because of the absence of specific clinical manifestations and imaging findings. A few reports have described PSH in women of advanced age. We have drawn the following conclusions from the few medical records of older women diagnosed with PSH.<sup>6,7</sup> First, because of the diversity of symptoms and lack of obvious nodules, PSH is readily misdiagnosed as a malignant tumor.<sup>8,9</sup> Second, in some patients, the

nodules become smaller and the surrounding cavities disappear after anti-infection treatment; thus, the tumor is readily misdiagnosed as a pulmonary abscess.<sup>10</sup> Finally, some articles have pointed out that dynamic enhanced CT showed significant early enhancement of histologically hemangiomatous and papillary lesions, and this enhancement has become a major cause of misdiagnosis.<sup>11,12</sup> The patient in the present case was atypical. She was a 75-year-old Chinese woman who presented with cough and sputum production with intermittent hemoptysis of 5 days' duration.

In two previous reports, 96 of 100 lesions were well-circumscribed nodules<sup>13,14</sup> ranging from 3 to 11 cm in the greatest dimension (mean of approximately 4.3 cm). In our patient, CT showed an unremarkable nodule. Differences in growth have been reported,<sup>2,3</sup> with sizes ranging from 1 to 3 cm in diameter.<sup>15</sup> Surprisingly, our patient

did not experience discomfort for 2 years. Some studies have described PSH as a tumor with a low malignant potential.<sup>16,17</sup>

No consensus has been reached regarding the origin of PSH, but immunohistochemistry can be helpful. PSH is characterized by the presence of cuboidal and polygonal cells and four histological types: papillary, sclerotic, solid, and hemorrhagic.<sup>3,18</sup> The tumor cells in the present case were immunopositive for TTF-1 and epithelial membrane antigen. The two cell types coincidentally expressed both antigens,<sup>19,20</sup> which are the same markers as previously reported by other researchers.<sup>21</sup> Because TTF-1, epithelial membrane antigen, phosphoenolpyruvate carboxykinase, and cytokeratin 7 are epithelial markers, the immunopositive staining pattern was consistent with an endothelial origin.

The Ki-67 nuclear antigen is a marker of cell proliferation, and high Ki-67 expression indicates tumor activity.<sup>22</sup> In this patient, the Ki-67 index was 10%, indicating that the PSH was a benign tumor that had enlarged.

PSH is frequently treated by or diagnosed after surgical resection, but no previous reports have described complications of surgical resection or patient survival following surgery.<sup>4,23</sup> Some reports have indicated that preoperatively diagnosed PSH had no effect on respiratory function or survival.<sup>24,25</sup> In the present case, surgical resection was probably the best treatment for the following reasons. First, preoperative and intraoperative diagnosis of PSH is difficult because its pathological features are similar to those of atypical adenomatous hyperplasia and bronchioloalveolar carcinoma. Second, conservative treatment for PSH may be indicated if the patient has no symptoms. We determined that surgery was indicated for this patient.

## Conclusions

In conclusion, PSH is a rare lung neoplasm that is considered to have very low potential

for malignancy. Great care should be taken during resection and patient follow-up because of the potential for malignancy. Both bronchiectasis and PSH can cause hemoptysis. This case supports the inclusion of PSH as a differential diagnosis of hemoptysis in older women. When other common lung diseases respond poorly to treatment, PSH should be considered and pathological biopsy should be performed if necessary.

## Abbreviations

PSH, pulmonary sclerosing hemangioma; CT, computed tomography; TTF-1, thyroid transcription factor 1.

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## Availability of supporting data

All data are fully available without restriction.

## Declaration of conflicting interest

The authors declare that there is no conflict of interest.

## Ethical approval and consent to participate

This study was approved by the institutional research ethics committee of the Wuhan Third Hospital (Tongren Hospital of Wuhan University), and written informed consent was obtained from the patient.

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