Pulmonary sclerosing hemangioma mimicking bronchial malignancy: A case report
Bhuiyan AKMMR, Hasan MN, Islam MT, Ferdous SMH, Hossain A

Abstract
Pulmonary sclerosing hemangioma is a rare benign neoplasm of the lung and predominantly occurs in female patients. Commonly, it presents as asymptomatic solitary pulmonary nodule on chest radiograph. We report a case of pulmonary sclerosing hemangioma who presented with fever and chest x-ray and computed tomography revealed cavitary lesion and mediastinal lymphadenopathy. The chest imaging findings were mimicking bronchial carcinoma. On histopathology and immune-cyto-histochemistry, we found her as having pulmonary sclerosing hemangioma. The patient is now on follow up without any progression of the disease for last two months.

Key words: bronchial carcinoma, cavitary lesion of lung, pulmonary sclerosing hemangioma.

Introduction
Pulmonary sclerosing hemangioma (PSH) is an uncommon benign neoplasm, which was identified by Liebow and Hubbel in 1956.1 PSH is prevalent in females, with a female to male ratio of 5:1 and is common in middle aged people over the age 50 years.2,3 The origin of the tumor is debatable but recent theory suggests it arises from primitive pulmonary endothelium most likely type-II alveolar pneumocyte. But the nomenclature of the tumor came from the fact that it arises from both endothelial and vascular cells.2 PSH remains clinically asymptomatic and found in chest radiograph as peripheral solitary pulmonary nodule.3 The tumor size varies from 0.3 cm to 8 cm.2,5 In this report, a PSH case that presented with cough and cavitary lesion with mediastinal lymphadenopathy on chest imaging mimicking lung malignancy was studied.

Case report
A 50-year-old male presented with nonproductive cough for 3 months. He had no history of fever and weight loss. He was a diagnosed case of hypertension and diabetes mellitus. He was non-smoker and non-alcoholic.
On examination, pulse was 84 beats/min, blood pressure was 110/70 mm of Hg and there was no lymphadenopathy and clubbing. Systemic examination findings were normal. Investigation reports revealed haemoglobin 10.9 gm/dl, erythrocyte sedimentation rate 65 mm in first hour, fasting blood glucose 6.7 mmol/l and renal and liver function tests were normal. Chest x-ray postero-anterior view showed left sided pulmonary cavitory lesion with hilar lymphadenopathy (Figure 1).

Figure 1 Chest x-ray postero-anterior view showing left sided cavitory lesion with hiliar lymphadenopathy
Computed tomography (CT) scan of chest with contrast showed cavitary lesion with consolidation with mediastinal lymphadenopathy suggestive of neoplastic lesion (Figure 2).

The core biopsy from cavitary lesion of lung followed by histopathology with haematoxyllin and eosin staining showed that surface cell lining papillae and round cell within papillary cores and round sheets. The surface cells were cuboidal, resembling type-II pneumocytes and were showing foamy cytoplasm, nuclear pseudo-inclusions. It had sclerotic stroma. No malignant cells were seen (Figure 3).

Immuno-cytocohistochemistry report revealed that CD34, TTF1 and CD31 were positive (Figure 4-A,B,C). All the reports were suggestive of PSH. After consultation with pulmonology and thoracic surgeon decision was made to follow up the patient as he was asymptomatic. For the last two months he was asymptomatic without any progression of the disease in the chest x-ray.

Figure 2 Contrast enhanced computed tomography (CT) scan of chest showing a thick walled cavity in the left side of the lung field

Figure 3 Histopathological findings of the tissue taken from the cavitary lesion of lung with H&E staining showing surface cells lining papillae and round cells within papillary cores and solid sheets. The surface cells are cuboidal resembling type-II pneumocytes.

Figure 4 Immuno-cyto-histochemistry of the tissue taken from the cavitary lesion of right lung showing: A) CD34 positive, B) TTF-1 positive and C) CD31 positive
Discussion

PSH is a rare disease entity which is more often misdiagnosed as lung tumor either benign or malignant. Moreover pathologists can give correct diagnosis only in fifty percent of the cases. In this case, patient presented with chronic cough. PSH typically presents as peripheral, solitary, well defined, homogenous nodule or mass lesion, rarely with cystic component and mediastinal lymphadenopathy on chest x-ray. Contrast enhanced CT scan of chest demonstrates rounded mass with regular boundary but this is not diagnostic. Signs in the CT scan may include pseudo-capsule, overlying vessels, air gap and halo sign. Most of the PSH show increased fluoro-deoxyglucose uptake and lesion above 2 cm can be frequently misinterpreted as malignant tumor. Our patient had cavitary lesion on the chest x-ray and CT scan which is an uncommon manifestation according to the literature. Moreover there was mediastinal lymphadenopathy on chest radiograph that is also a rare feature.

Histologically, PSH comprises of cuboidal surface cell and polygonal stromal cell that has potential to differentiate into other type of cells. Immuno-cytohistochemical findings are required for confirming the diagnosis. The stromal cells are positive for epithelial membrane antigen (EMA) and thyroid transcription factor-1 (TTF-1), while the surface cells are positive for CD31, EMA, TTF-1 and cytokeratin-7 (CK7). The stromal cell can also be positive for neuroendocrine markers like chromogranin-A, neuron-specific enolase (NSE) and synaptophysin. In addition, some cells are positive for carcinoembryonic antigen (CEA) and vimentin. In this particular case, immuno-cytohistochemistry report revealed TTF-1 and CD31 were positive which was consistent with the diagnosis of PSH.

In conclusion, PSH may present with radiological features suggestive of bronchial malignancy, but care should be taken not to miss PSH in appropriate scenarios.

Conflicts of interest: Nothing to declare.

References

1. Liebow AA, Hubbell DS. Sclerosing hemangioma (histiocytoma, xanthoma) of the lung. Cancer 1956; 9: 53-75.
2. Devouassoux-Shisheboran M, Hayashi T, Linnoila RI, Koss MN, Travis WD. A clinicopathologic study of 100 cases of pulmonary sclerosing hemangioma with immune-ohistochemical studies: TTF-1 is expressed in both round and surface cells, suggesting an origin from primitive respiratory epithelium. Am J Surg Pathol 2000; 24: 906–16.
3. Illei PB, Rosai J, Klimstra DS. Expression of thyroid transcription factor-1 and other markers in sclerosing hemangioma of the lung. Arch Pathol Lab Med 2001; 125: 1335–39.
4. Katzenstein AL, Gmelich JT, Carrington CB. Sclerosing hemangioma of the lung: a clinicopathologic study of 51 cases. Am J Surg Pathol 1980; 4(4): 343-56.
5. Kuo KT, Hsu WH, Wu YC, Huang MH, Li WY. Sclerosing hemangioma of the lung: an analysis of 44 cases. J Chin Med Assoc 2003; 66:33–38.
6. Kim KH, Sul HJ, Kang DY. Sclerosing hemangioma with lymph node metastasis. Yonsei Med J 2003; 44: 150–54.
7. Shin SY, Kim MY, Oh SY, Lee HJ, Hong SA, Jang SJ, et.al. Pulmonary sclerosing pneumocytoma of the lung: CT characteristics in a large series of a tertiary referral center. Medicine (Baltimore) 2015; 94: 98.
8. Chan AC, Chan JK. Pulmonary sclerosing hemangioma consistently expresses thyroid transcription factor-1 (TTF-1): a new clue to its histogenesis. Am J Surg Pathol 2000; 24(11): 1531-36.
9. Bejarano PA, Baughman RP, Biddinger PW, Miller MA, Fenoglio-Preiser C, al-Kafaji B, et al. Surfactant proteins and thyroid transcription factor-1 in pulmonary and breast carcinomas. Mod Pathol 1996; 9(4): 445-52.
10. Lee E, Park CM, Kang KW, Goo JM, Kim MA, Paeng JC, et al. 18F-FDG PET/CT features of pulmonary sclerosing hemangioma. Acta Radiol 2013; 54:24–29.