Cent. Eur. J. Med. • 5(4) • 2010 • 447-451
DOI: 10.2478/s11536-009-0090-x

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Central European Journal of **Medicine**

Pulmonary sclerosing hemangioma with atypical radiologic findings

Case Report

Ozgur Oztekin[1*], Recep Savas[2], Duygu Gurel[3], Nezih Ozdemir[4], Mustafa Akcaoglu[5]

[1] *Izmir Education and Research Hospital, Radiology Department, 35360 Izmir, Turkey*

[2] *Ege University Faculty of Medicine, Department of Radiology, 35100 Izmir, Turkey*

[3] *Dokuz Eylül University Faculty of Medicine, Department of Pathology, 35340 Izmir, Turkey*

[4] *Dokuz Eylül University Faculty of Medicine, Department of Thoracic Surgery, 35340 Izmir, Turkey*

[5] *Tepecik Education and Research Hospital, Radiology Department, 35120 Izmir, Turkey*

Received 12 June 2009; Accepted 13 June 2009

Abstract: Sclerosing hemangioma of the lung is an uncommon benign tumour that usually presents as an asymptomatic solitary nodule and affects middle age women. Because there are few findings on radiologic studies that are characteristic of sclerosing hemangioma, it is difficult to diagnose on the basis of imaging and biopsy remains the definitive diagnostic test. We report a case of pulmonary sclerosing hemangioma with extremely unexpected imaging findings and review the literature.

Keywords: *Sclerosing hemangioma • Lung • Computed Tomography • Magnetic Resonance Imaging • Positron Emission Tomography*

1. Introduction

Sclerosing hemangioma of the lung is a rare benign neoplasm affecting predominantly females over 50 years of age [1,2]. Affected patients are mostly asymptomatic, but some present with hemopthysis, coughing, chest pain, dyspnea and pleurisy [2]. Sclerosing hemangioma of the lung was first reported by Liebow and Hubbell [2] in 1956. Although initially regarded as a variant of hemangioma on the basis of results of immunohistochemical and electron microscopic studies, sclerosing hemangioma is now considered an epithelial tumor. Most of the investigators have mentioned relatively characteristic plain film, CT and MRI findings, but there is little information regarding the positron emission tomography (PET) scan findings. Here we report a patient with atypical radiological presentations.

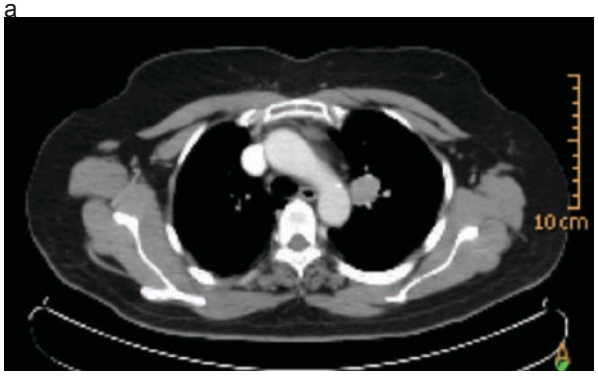
2. Case Report

A 58-year-old female with hemopthysis and insistent coughing over two months was admitted to our hospital. Her medical history, physical examination, and laboratory findings were normal. Then the patient underwent chest radiography, which revealed a solitary well circumscribed left suprahilar mass. On CT the medial border of the suprahilar mass was sharply marginated and the aortic margin was intact (Figure 1). However the inferior border of the lesion was in close contact with the left main pulmonary artery, which was demostrated with the multiplanar imaging capability of MRI. MRI revealed that the tumor was in contact with the pulmonary artery without an invasion. Transthoracic biopsy was performed, but finding for a definitive diagnosis was not obtained. Although the clinical and

* *E-mail: oztekinozgur@gmail.com*

Figure 1. (a) Contrast-enhanced CT image of the thorax obtained at the level of aortic arcus shows well circumscribed noduler mass with a very low contrast enhancement (8 HU). (b) Contrast-enhanced CT at lung window settings shows a sharply demarcated mass with a diameter of 2.8cm located at the left apicoposterior segment of the upper lobe.

a



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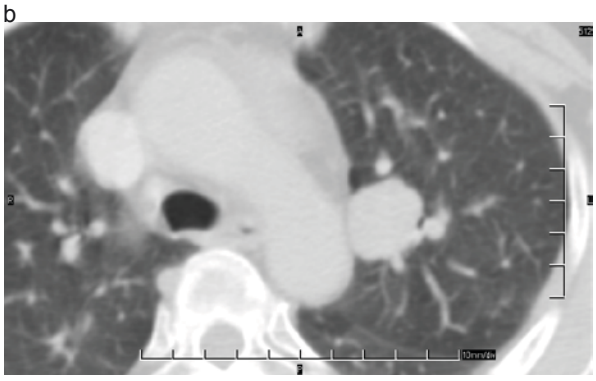


laboratory findings were unremarkable, CT and MRI findings could not rule out the possibility of malignancy. Then PET scan was planned. The mass exhibited no uptake of ^{18}F-fluorodeoxyglucose (FDG), with a maximum standardized uptake value of 1.3 SUV (Figure 2).

Because a malignant lung tumor could not be excluded, the patient underwent mass resection for the possibility of lung cancer. Surgical exploration revealed a well-demarcated round mass without invasion to the surrounding tissue with the exception of the inferior border. In the inferior border the mass was in close contact with the pulmonary artery. After learning the result of frozen biopsy the mass was resected, leaving only small pieces of the mass that were adherent to the pulmonary artery due to the risk of pulmonary vasculature damage. The mass was resected without serious damage to the pulmonary paranchyma and vasculature. Macroscopic pathologic examination revealed a solitary grey-brown tumor, measuring 3x5cm, that was located in the lung paranchyma. Microscopically, the histopathologic specimen showed features of SH, with variable proportions of solid patterns and focal papillary component. There were also hemorrhagic areas scattered throughout the mass. The tumor was composed of solid sheets of round to polygonal cells punctuated by papillea, and the cleft was lined by cuboidal cells. The tumor cells stained positively for epithelial membrane antigen, CAM 5.2, thyroid transcription factor-1, and cytokeratin fragment 7; they stained negatively for factor VIII and CD34. These immunohistochemical results suggested that the tumor originated from the epithelium, especially from type II pneumocytes. Therefore, this tumor was diagnosed as pulmonary sclerosing hemangioma.

3. Discussion

SH is also referred to as benign sclerosing pneumocytoma, benign pulmonary histiocytoma and sclerosing angioma xanthomatous pseudotumor, and it is a rare disease. It was first described by Liebow and Hubbell in 1956 [2]. It occurs predominantly in young or middle-aged women. Pathologically, it is composed of four major histologic components: solid, papillary, sclerotic, and hemangiomatous components.

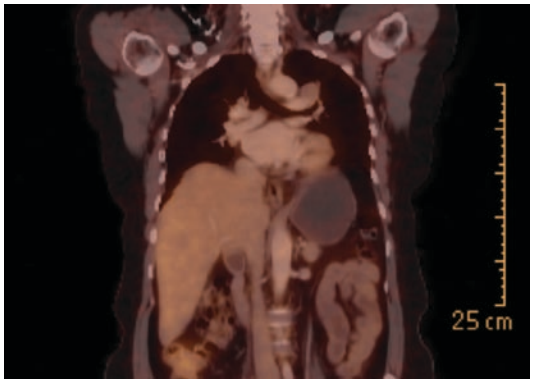
SH of the lung is almost always benign, with only approximately 2-4% having nodal metastasis [3]. As of 2008, there were 10 reported patients with lymph node metastases; in a report with the largest series of nodal metastases, who all underwent lobectomy and regional lymph node dissection (hilar and peribronchial), all four were alive at a mean follow-up of 4.7 years, without evidence of residual or recurrent disease [4,5]. Devouassoux-Shisheboran et al. observed several unusual presentations of SH, which included multifocal lesions, hilar lymph nodes, and pleural and mediastinal locations [6]. Daan B. de Koning et al. described a case with an association of familial adenomatous polyposis (FAP) and SH, suggesting that SH and FAP share the same pathophysiology [7]. However, series with large number of patients with FAP will need to be screened with modern imaging modalities to support this estimation.

There have been many suggestions as to the origin of the tumor. More recent studies have offered a variety of proposed etiologies regarding the cell line of origin. These include mesothelial, mesenchymal, neuroendocrine and epithelial origins [3,5]. Various diagnostic modalities including immunohistochemistry and electron microscopy have been developed, and this disease is now accepted as an epithelial neoplasm derived from primitive respiratory epithelium, incompletely differentiated type II pneumocytes, or clara cells [6,8].

Figure 2. (a) Axial PET image reveal a mass with a maximal SUV of 1.3. (b) Coronal and (c) axial PET-CT images corresponding to (a) reveal no uptake in lesion.

a

b



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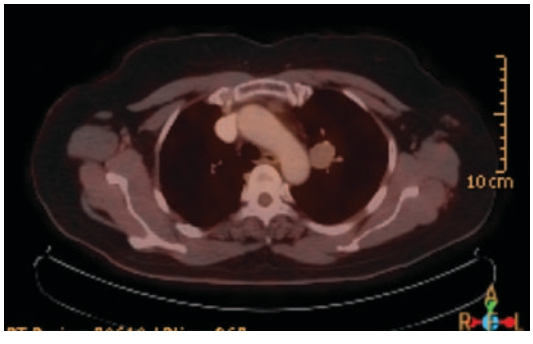
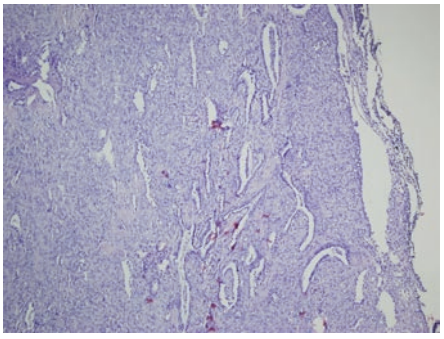


Macroscopically, SH were found as well circumscribed and often hemorrhagic tumours. A thin fibrous pseudocapsule separates it from the surrounding pulmonary paranchyme. Microscopically, SH is composed of two types of cells: cuboidal cells that line papillary structures and round to polygonal cells that form solid sheets [5-7,9]. The neoplasm has four possible histologic components: papillary, sclerotic, solid, and hemorrhagic (Figure 3). Most tumors have at least three of these components, and a minority have only two. In the large series reported to date, no tumor has shown a single component [6,7,9].

Figure 3. E(a) Low-power image shows typical sclerotic and papillary pattern seen in SH of the lung. (b) Papillary pattern shows uniform cuboidal cells lining the papillae (H & E, X40), (c) and spaces filled with a hemmorhage between the papillea (H & E, X40).

a



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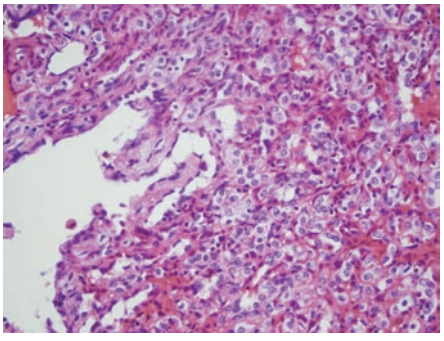

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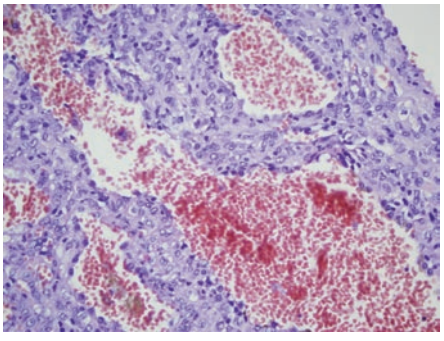

Although not specific, CT characteristics of SH are relatively characteristic. According to a report on the CT findings in ten cases, this tumor presents as a smooth marginated (100% of cases), homogenously attenuating (100%) round or ovoid mass (100%), sometimes including calcifications (30%) [10]. Dynamic CT studies have shown in this report that nine of the ten tumors had more than 60 hansfield unit (HU) net enhancement. Only one compact solid type of tumor in this study had slow and persistant enhancement (33 HU) with little washout. Interestingly in the present study, the mass in our patient had only little contrast enhancement (8 HU) after IV contrast administration. Until now this was the lowest amount of contrast enhancement, which may

Table 1. In the literature there were only five case reports of sclerosing hemangioma of the lung concerning FDG uptake.

Source, Year	FDG uptake	SUV
Longo R. et al. 2005	Moderate uptake	1.8
Hara M. et al. 2001	Moderate uptake	1.8
Neuman J. et al. 2006	Increased uptake	3.1
de Koning DB. et al. 2007	Moderate-low uptake	1.6
Hosaka N. et al. 2004	Moderate uptake	1.8
Oztekin O. et al. 2009	Low uptake	1.3

be due to difference in histologic composition of the tumor. However, studies comparing different histologic types and amount of net contrast enhancement in these different types need to be studied to prove this hypothesis. The air meniscus sign or air-trapping zone around sclerosing hemangioma has been reported to be a specific CT sign for sclerosing hemangioma [2]. The mechanism has been suggested as being peritumoral hemorrhage followed by clearance through the airway, creating a peritumoral air space [2]. This sign may be helpful to differentiate sclerosing hemangioma from other benign tumours, but is observed seldomly.

MR seems to be also useful in suggesting the diagnosis; as reported by Fujiyoshi et al. [10], the T1-weighted high signal intensity areas correspond to those including abundant clear cells, and the T2-weighted low signal intensity areas correspond to the fibrotic or hemorrhagic regions within the tumours. The hemangiomatous component of the tumours shows high signal intensities on T2-weighted images and marked signal elevation after contrast administration. However, Guibaud et al. [11] reported the MR appearance of a young girl with SH as being homogeneous and similar to muscle on T1-weighted images, hyperintense on T2-weighted images, and mildly enhancing with gadolinium. In addition, Hara et al. [12] noted: mixed low and intermediate signal intensities on T1-weighted images; mixed low, intermediate, and high (the last representing cystic change) signal intensities on T2-weighted images; and marked gadolinium enhancement of a portion of the tumor in a 54-year-old asymptomatic woman. Further evaluation in a large number of cases would be necessary to confirm the usefulness of MRI for differential diagnosis.

To our knowledge, there have been only five case reports that have presented the PET findings in SH of the lung (Table 1). Hara et al. [12] presented a women with moderate increased uptake of radiolabeled fluorodeoxyglucose (FDG) on PET who had incidental left lower lobe mass on chest radiography. The second report [13] concerning a young women with a left upper lobe SH and the third report [7] concerning a 53-year-old female with left lower lobe mass with a known FAP disease both noted moderate uptake of FDG by the lesion. Neuman J. et al. [5] presented a middle aged women with right lower lobe SH and substantially increased FDG uptake. Hosaka N. et al. also presented a 45-year-old woman with an asymptomatic solid tumor in the lower right lobe of the lung [14] with moderate increased uptake of FDG. In all of these cases PET scan simulated that of a malignancy. Our case has the lowest amount of FDG uptake on PET, which can speculate variable FDG uptake for SH.

FDG PET provides quantitative information in the form of the standardized uptake value (SUV). The SUV is obtained by putting the circular region of interest over the portion of the lesion with the greatest accumulation of FDG. In spite of the controversial views, SUVs of 2.5 or greater have been used as a cutoff value indicative of malignancy.

In conclusion, although SH has some relatively characteristic radiological appearences, cases such as the present case with atypical imaging findings remind us of the importances of biopsy for definitive diagnosis.

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