Original Investigation

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Pulmonary Sclerosing Hemangioma (Pneumocytoma): An Analysis of 8 Cases

Gülbanu Horzum Ekinci[1], Osman Hacıömeroğlu[1], Ayşe Ersev[2], Levent Alpay[3], Ayçim Şen[2], Pınar Atagün Güney[1], Adnan Yılmaz[1]

[1]Clinic of Chest Diseases, Süreyyapaşa Chest Diseases and Thoracic Surgery Training and Research Hospital, İstanbul
[2]Clinic of Pathology, Süreyyapaşa Chest Diseases and Thoracic Surgery Training and Research Hospital, İstanbul
[3]Clinic of Chest Surgery, Süreyyapaşa Chest Diseases and Thoracic Surgery Training and Research Hospital, İstanbul

Abstract

Objective: We aimed to evaluate clinical manifestations, radiological findings, treatment, and outcomes of pulmonary sclerosing hemangioma (SH).

Methods: We retrospectively reviewed eight cases of pulmonary sclerosing hemangioma diagnosed at our institution from January 2006 to April 2014. Their demografic findings, symptoms, radiological appearences, diagnostic methods, treatments and surveys were recorded.

Results: There were four female and four male patients. The age at the time of diagnosis ranged from 23 to 79 years, with a mean age of 56.1 years. Two patients were asymptomatic. Among six symptomatic patients, hemoptysis was the most frequent symptom. The radiological feature was a solitary nodule or mass in seven cases. In the remaining case, there were multiple distinct masses and nodules in bilateral lung fields. The tumor involved lung parenchyma in seven cases and endobronchially located in the remaining patient. Seven patients with parenchymal location underwent surgery. Wedge resection was the most common surgical procedure. Diagnosis of SH was established by surgical biopsy in seven cases and by bronchoscopic biopsy in one case. The most common histological pattern was solid pattern. During the follow-up ranging from 2 months to 76 months, seven cases who underwent surgery had an excellent prognosis with no evidence of recurrence. The patient with multiple lesions died one month after diagnosis.

Conclusion: SH of the lung is a rare tumor. Surgical resection usually requires for both diagnosis and treatment of this tumor. The patients had excellent prognosis with no evidence of recurrence following surgery.

Keywords: Benign tumor, lung, sclerosing hemangioma, surgery

INTRODUCTION

Pulmonary sclerosing hemangioma (SH) is a rare tumor, accounting for approximately 0.2%–1% of all primary lung tumors (1). Various alternative terms such as pneumocytoma, sclerosing pneumocytoma, and papillary pneumocytoma have been suggested for this tumor (2). SH can occur at any age, although it is predominant in middle-aged females (3). Most patients remain asymptomatic, and the tumor is usually detected incidentally during routine radiological examinations for other reasons (4, 5). Generally, SH presents as a peripheral, solitary, well-circumscribed, round or oval nodule or mass (2, 4, 5). However, multiple bilateral lesions have also been reported (6, 7). Although this tumor almost always involves the pulmonary parenchyma, endobronchial localization is rare (8). Preoperative diagnosis of the tumor is difficult and precise diagnosis is established by surgical resection in most patients (1, 3, 9). Surgical excision of the lesion is a curable treatment method and the overall prognosis is excellent after surgery (4, 9). Here we present a review of 8 patients who had been pathologically diagnosed with pulmonary SH and evaluate their clinical manifestations, radiological findings, treatment, and outcomes.

METHODS

This retrospective study was conducted at Süreyyapaşa Center for Chest Diseases and Thoracic Surgery Training and Research Hospital, İstanbul, Turkey. This study was approved by the ethics com-

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Corresponding Author
Gülbanu Horzum Ekinci
E-mail: gulbanuh@hotmail.com

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mittee of the institute. A review of the records of the Department of Pathology at the institution from January 2006 to April 2014 revealed 8 patients with pulmonary SH who had histological proof of diagnosis. Pathological examination was by surgical resection of the tumor in 7 patients and by bronchoscopic biopsy in 1 patient. Hematoxylin and eosin staining was the basis for diagnosis. Hematoxylin and eosin-stained slides of all 8 cases were reviewed by 2 experienced pathologists to reconfirm the diagnosis. Immunohistochemical staining was also performed on tumor tissues from 7 of 8 patients using the avidin–biotin method.

Data regarding radiological examinations, diagnostic procedures, treatments, and follow-up were reviewed from the patients' clinical records. All patients underwent routine laboratory studies, electrocardiography, chest X-ray, computed tomography (CT) of the thorax, and flexible bronchoscopy. Pulmonary function tests and positron emission tomography (PET-CT) were performed in 7 patients who underwent surgery for diagnosis and treatment. Mediastinoscopy or lymph node dissection was performed in 4 of 7 patients, because of high maximum standardized uptake value (SUV_{max}) of the lesion on PET-CT.

Table 1. Clinical data of the patients with sclerosing hemangioma

Patient	Sex	Age (years)	Smoking Status	Symptoms
1	F	50	NS	Asymptomatic
2	F	58	NS	Cough, hemoptysis
3	F	41	NS	Asymptomatic
4	M	58	S	Hemoptysis
5	M	68	ES	Chest pain, hemoptysis
6	M	23	NS	Chest pain
7	F	72	NS	Dyspnea, chest pain
8	M	79	S	Dyspnea, cough, hemoptysis, weight loss

ES: Exsmoker; F: female; M: male; NS: non-smoker; S: smoker

RESULTS

Among the 8 patients with pulmonary SH, 4 were female and 4 were male, indicating a female-to-male ratio of 1:1. Age at the time of diagnosis ranged from 23 to 79 years, with a mean age of 56.1 years. Five (62.5%) patients were non-smokers. Two (25%) patients had no symptom and the tumors were detected during routine medical examination. Among the 6 symptomatic patients, hemoptysis was the most frequent symptom. The clinical data of the patients is given in Table 1.

Chest X-ray showed a solitary nodule in 5 patients, solitary mass in 2 patients, and multiple nodules and masses in 1 patient. CT of the thorax revealed multiple mediastinal lymphadenopathies in 3 patients. The size of the tumor on CT ranged from 2.2 cm to 5.1 cm, with a

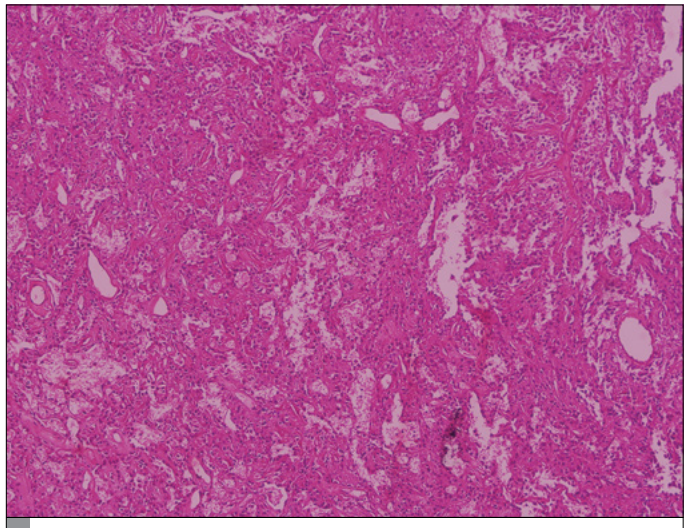

Figure 1. Microscopically, the tumor exhibited a papillary pattern (hematoxylin and eosin staining, ×100)

Table 2. CT, PET-CT, and bronchoscopy findings and pathologic data of the patients

Patient	CT	Size (cm)	Bronchoscopy	PET-CT (SUV_{max})	Location	Diagnostic method	Subtype of SH
1	Solitary mass and mediastinal LAP	3.5	Normal appearance	3.1	Superior segment of LLL	Surgery	Solid and hemorrhagic
2	Solitary nodule	2.2	Normal appearance	3.6	Lateral segment of RML	Surgery	Papillary
3	Solitary nodule	2.9	Normal appearance	4.3	Medial segment of RML	Surgery	Solid
4	Solitary mass and mediastinal LAPs	3.6	Normal appearance	6.3	Anterior segment of RUL	Surgery	Solid and sclerotic
5	Solitary nodule and mediastinal LAPs	2.8	Normal appearance	3.0	Posterior segment of RUL	Surgery	Solid and hemorrhagic
6	Solitary nodule	2.5	Normal appearance	2.7	Anterior segment of RUL	Surgery	Sclerotic
7	Solitary nodule	2.3	Normal appearance	1.6	Apicoposterior segment of LUL	Surgery	Solid
8	Multiple masses and nodules and LUL bronchi	5.1	Masses in the RLL	No	Bilateral	Bronchoscopy	Solid

CT: Computed tomography; LAP: lymphadenopathy; LLL: left lower lobe; LUL: left upper lobe; PET-CT: positron emission tomography-computed tomography; RLL: right lower lobe; RML: right middle lobe; RUL: right upper lobe; SH: sclerosing hemangioma; SUV_{max}: maximum standardized uptake value

Table 3. Treatment and outcomes of the patients

Patient	Surgical procedure	Operation	Size (cm)	Lymph node metastasis	Follow-up (months)	Prognosis
1	Left thoracotomy and mediastinoscopy	Wedge resection	4.0	No	2	Alive
2	Right thoracotomy	Wedge resection	2.2	--	34	Alive
3	Video-assisted thoracoscopic surgery	Enucleation	3.0	--	31	Alive
4	Right thoracotomy and lymph node dissection	Lobectomy	3.0	No	20	Alive
5	Right thoracotomy and lymph node dissection	Lobectomy	3.0	No	24	Alive
6	Right thoracotomy and mediastinoscopy	Wedge resection	2.5	No	36	Alive
7	Left thoracotomy	Wedge resection	2.0	--	76	Alive
8	No	No	--	--	1	Died

mean size of 3.1 cm. Flexible bronchoscopy revealed an endobronchial lesion in only 1 of the 8 patients. The tumor location was peripheral in all but 1 patient. Among the peripheral lesions, 3 were located in the right upper lobe, 2 in the right middle lobe, 1 in the left upper lobe, and 1 in the left lower lobe. The mean SUV_{max} of the lesions on PET-CT was 3.5 (range: 1.6–6.3). Diagnosis of SH was established by surgical biopsy in 7 cases and by bronchoscopic biopsy in 1 case. The most common histological pattern was solid pattern. The papillary pattern was observed in 1 patient (Figure 1). Three patients exhibited 2 patterns (Table 2).

Data regarding the treatment and outcomes of the patients are summarized in Table 3. Surgical procedures were administered for 7 patients. Wedge resection was the most common surgical procedure. Because the lesion showed a high SUV_{max} on PET-CT, mediastinoscopy or lymph node dissection was performed in 3 of these patients, and there was no lymph nodule with pulmonary SH metastasis. Lymph node biopsies revealed chronic inflammation in 2 patients and tuberculous lymphadenitis in 1 patient. An intraoperative frozen section for pathological examination was performed in 5 of 7 patients who underwent a surgical procedure. Among them, 2 patients were diagnosed with SH. Two patients were claimed to have a benign lesion without a definitive diagnosis. Malignancy was suspected in 1 patient. There was no operative mortality and morbidity. None of the patients underwent adjuvant therapy. During the follow-up period ranging from 2 months to 76 months, none of the patients experienced tumor recurrence. The patient with multiple lesions died of respiratory failure a month after diagnosis.

DISCUSSION

Pulmonary SH is a rare tumor, accounting for approximately 1% of all benign pulmonary tumors (10). This tumor was first described by Liebow and Hubbel in 1956 (2, 3). Its histogenesis is uncertain (9). Initially, the name "sclerosing hemangioma" was used for this lesion because the tumor was thought to be a lesion derived from the vascular endothelium. However, recent studies have showed that SH originates from type II pneumocytes. Thus, some investigators have used the term pneumocytoma (3, 11).

Pulmonary SH can occur in any individual in the age group of 4–73 years, with the risk of occurrence being highest in the fifth decade of life (3, 12). This tumor is predominant in females, and the female-to-male ratio ranges from 5:1 to 31:1 (3-5, 12, 13). The mean age of the patients was 56.1 years in the present study. There was no sex predilection in our series. Most patients remain asymptomatic, and the tumor is usually identified incidentally during routine radiological examinations for other reasons. Among symptomatic patients, hemoptysis, chest pain, and cough are the most common symptoms (2, 9, 13, 14). The typical radiological finding of SH is a peripheral, solitary, well-circumscribed, round or oval solid nodule or mass (2, 4, 5). Multiple lesions occur in 4% of all cases (7, 8). Cavitation, calcification, and cystic appearance are rarely seen (2, 15). SH almost always involves the pulmonary parenchyma. Endobronchial localization is rare and seen in only 1% of the patients (8). Seven of our patients presented with a typical radiological appearance. The remaining patient had multiple lesions, and bronchoscopy revealed 2 endobronchial gross tumors in this patient. Experience of using PET in SH is limited. There are no large series defining the PET characteristics of SH. A few case reports noted that there were different SUVs of 18F-FDG in the lesions (4, 10). In a previous study, Lei et al. (4) reported a hypermetabolic lesion in only 1 of 5 patients. In our series, the SUV_{max} of the lesion on PET-CT ranged from 1.6 to 6.3, with a mean value of 3.5. It has been reported that the high uptake value may be related to the tumor size or the potentially low-grade malignant nature of SH (4, 10).

Preoperative diagnosis of the tumor is difficult (1, 3, 9). Pulmonary SH can be misdiagnosed preoperatively. It can be easily misdiagnosed as a malignant tumor during intraoperative frozen-section assessment and has a misdiagnosis rate of 25%–56% (3, 13). Precise diagnosis is established by surgical resection in most patients (1, 3, 9, 13, 14). Hematoxylin and eosin staining is the basis for diagnosis (9). Immunohistochemical analysis is performed to confirm the origin of the tumors (2, 9, 12). The tumor cells typically express epithelial membrane antigens, cytokeratin, vimentin, estrogen and progesterone receptor proteins, and thyroid transcription factor-1 and are negative for S-100 protein, carcinoembryonic antigen, and endothelial cell markers (16). SH shows 4 distinct histological patterns: papillary, solid, sclerotic, and hemorrhagic. The tumors can demonstrate a single pattern or more than one pattern (3, 9, 12). In our series, the tumor showed a single pattern in 5 patients but 2 patterns in 3 patients. The solid pattern was the most common. The histological differential diagnosis of SH includes benign lung tumors, malignant lung tumors, metastatic tumors, tuberculoma, and Castleman's disease (2, 3).

Pulmonary SH is generally considered a benign lesion. However, cases with multiple and bilateral lesions and lymph node or pulmonary metastasis or recurrence have been reported (2, 4, 6, 16-18). Most studies have reported that multiple lesions, lymph node metastasis, recurrence, and pulmonary metastasis do not affect the prognosis (4,

Table 4. Results of 3 published studies

	Reference		
	Kuo KT. (14)	Iyoda A. (18)	Lei Y. (4)
No. of patients	44	26	28
Female	37	25	25
Male	7	1	3
Female-to-male ratio	5.3:1	25:1	8.3:1
Mean age (range), years	46.5 (16–72)	46.2 (17–64)	46.1 (25–58)
Location n (%)			
Right upper lobe	10 (22.8)	4 (15.4)	5 (17.8)
Right middle lobe	8 (18.2)	4 (15.4)	4 (14.3)
Right lower lobe	11 (25)	8 (30.8)	6 (21.4)
Left upper lobe	7 (15.8)	2 (7.6)	3 (10.7)
Left lower lobe	8 (18.2)	8 (30.8)	9 (32.1)
Multiple	No	No	1 (3.7)
Pathological Pattern, n (%)			
Solid	19 (43.2)	25	Unknown
Papillary	29 (65.9)	23	Unknown
Sclerotic	32 (72.7)	26	Unknown
Hemorrhagic	28 (63.6)	23	Unknown
Survival data			
Follow-up interval, months	3–228	1–228	6–192
Alive	44	26	28
Died	No	No	No
Relapse	No	1	No

17). Surgical excision of the tumor is curative without the need for additional treatment (4, 9, 12). Surgical procedures include limited resections, such as wedge resection, tumor enucleation, and lobectomy (4, 8, 13, 14, 18). The recommended treatment for SH is wedge resection if a safe resection margin can be achieved (8, 13). Lobectomy is sometimes indicated for central tumors (14). The prognosis of patients with SH is excellent after surgical excision of the tumor. Recurrence is rare (4, 9, 13-15, 18). Wedge resection was the most common procedure among our patients. Our patients had good prognosis after surgical treatment, without evidence of recurrence. The results of 3 published studies are summarized in Table 4.

CONCLUSION

Sclerosing hemangioma is a rare tumor of the lungs. It can present with multiple or endobronchial lesions. The histological diagnosis of SH is established by surgical resection in most patients. The patients have excellent prognosis, with no evidence of recurrence following surgery.

Ethics Committee Approval: Study done by retrospective data analyses so ethics commitee approwal was not taken.

Informed Consent: Due to the retrospective design of the study, informed consent was not taken.

Peer-review: Externally peer-reviewed.

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