

Radiological-Pathological Findings of Central Sclerosing Hemangioma Initially Misdiagnosed as Papillary Adenoma by Bronchoscopic Biopsy: A Case Report¹

기관내시경 조직검사에서 유두상 선종으로 오진된 중심성 경화혈관종의
영상의학적-병리학적 소견: 증례 보고¹

Soo Hyun Kim, MD¹, Hyun Ju Seon, MD¹, Jang-Hyeon Song, MD¹, Seo Yeon Park, MD¹,
Yun-Hyeon Kim, MD¹, Yoo-Duk Choi, MD², Sang-Yun Song, MD³

Departments of ¹Radiology, ²Pathology, ³Thoracic and Cardiovascular Surgery, Chonnam National University Hospital, Chonnam National University Medical School, Gwangju, Korea

Sclerosing hemangioma is relatively rare, the second most common benign pulmonary neoplasm, which usually presents the peripheral location. Central location of this neoplasm is extremely rare with only a few reports. Herein, we would like to report an extremely rare case of central sclerosing hemangioma with descriptions of radiological characteristics. It was initially misdiagnosed as a papillary adenoma by bronchoscopic biopsy and mimicked central lung malignancies such as carcinoid tumors on non-invasive image evaluations. However the patient was finally confirmed with surgery.

Index terms

Sclerosing Hemangioma
Pneumocytoma
Endobronchial
Pulmonary Neoplasm

INTRODUCTION

Sclerosing hemangioma is relatively rare, the second most common benign pulmonary neoplasm (1), which is initially considered to be of vascular origin. However, recent immunohistochemical and genetic studies suggest that sclerosing hemangioma is an epithelial tumor, related to the pulmonary epithelium. Typically, this is a solitary, generally asymptomatic, and well-described lesion located in the periphery of the lung. Therefore, sclerosing hemangioma which contains endobronchial portion is extremely rare (2). The histological characteristics of sclerosing hemangioma have been well known for showing solid, papillary, sclerotic and hemorrhagic patterns (1). However, the specimens obtained by bronchoscopic biopsy may be limited to the papillary or other specific histologic patterns. It

can also be misdiagnosed. Therefore, we report an extremely rare case of central sclerosing hemangioma in 58-year-old woman with radiological-pathological findings, which was initially misdiagnosed as papillary adenoma by bronchoscopic biopsy and mimicked central lung malignancy on non-invasive image evaluations.

CASE REPORT

A solitary lung mass with central location in right upper lobe was discovered in 58-year-old female without specific symptoms. The initial chest PA radiograph showed about 3 cm sized nodular opacity in the right hilar area (Fig. 1A), and the initial chest CT revealed a well-defined centrally located mass of about 3 cm diameter, and heterogeneous contrast enhancement in the

Received May 28, 2013; Accepted July 26, 2013

Corresponding author: Hyun Ju Seon, MD
Department of Radiology, Chonnam National University Hospital, Chonnam National University Medical School, 160 Baekseo-ro, Dong-gu, Gwangju 501-746, Korea.
Tel. 82-62-220-5888 Fax. 82-62-226-4380
E-mail: sunaura@hanmail.net

This is an Open Access article distributed under the terms of the Creative Commons Attribution Non-Commercial License (<http://creativecommons.org/licenses/by-nc/3.0>) which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

right upper lobe (RUL) abutting the posterior segmental bronchus (Fig. 1B). There was no evidence of infiltration on the surrounding lung parenchyma or lymphadenopathy. Bronchoscopically, there was an endobronchial lesion obliterating posterior segmental bronchus of RUL with easy touch bleeding, and the bronchoscopic suspicion was a carcinoid tumor (Fig. 1C). The pathologic result of initial bronchoscopic biopsy was a papillary adenoma (Fig. 1D).

However, the serial of follow-up chest CT scans show gradual increases of the mass with small endobronchial portion. The last follow-up CT scan obtained 7 years after the initial exam indicated about 1.1 cm interval growth of central mass in the posterior segment of RUL with ice-burg sign. The large extraluminal portion with relatively small endobronchial portion is known as a typical radiologic finding of slow growing central carcinoid tumor (3), with newly developed distal obstructive pneumonitis and subsegmental atelectasis (Fig. 1E). The follow-up positron emission tomography-CT (PET-CT) scan also revealed high fluorodeoxyglucose (FDG) uptakes of the mass [maximal standardized uptake value (SUV_{max}): 5.8] (Fig. 1F). These image

findings suggested a slow growing low grade central lung malignancy with some hypervascularity such as a carcinoid tumor rather than benign neoplasm. For accurate diagnosis and treatment, the patient underwent posterior segmentectomy of the RUL, and the mass was completely removed. The mass showed papillary, hemorrhagic, and sclerotic pattern composed of surface cuboidal cells and sheets of round cells (Fig. 1G). There was no necrosis or mitotic figures. On the immunohistochemical stains, surface cuboidal cells were positive for cytokeratin (CK), epithelial membrane antigen (EMA), and thyroid transcription factor-1 (TTF-1), but round cells were positive for EMA and TTF-1, and negative for CK. Therefore, the final histopathologic diagnosis of the specimen was a sclerosing hemangioma.

DISCUSSION

Sclerosing hemangioma is relatively rare, the second most common benign pulmonary neoplasm, and named due to histologically prominent features of sclerotization and vascularization of the tumor (1). Many recent immunohistochemical stud-

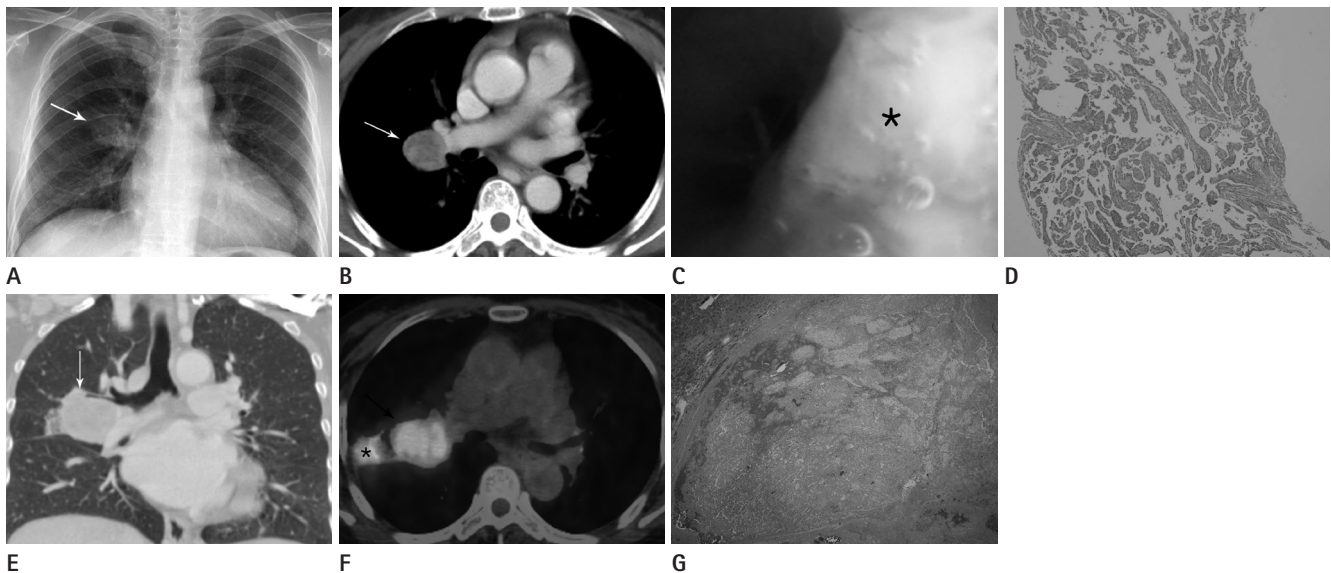


Fig. 1. Centrally located sclerosing hemangioma with endobronchial extension in a 58-year-old female.

- A.** Initial chest PA radiograph shows about 3 cm sized nodular opacity (arrow) in right hilar area.
- B.** The initial chest CT image shows about 3 cm sized well-defined central mass (arrow) with heterogeneous enhancement in right upper lobe (RUL).
- C.** Bronchoscopy shows a protruding endobronchial lesion in posterior segmental bronchus of RUL (*).
- D.** Only papillary pattern of tumor was obtained on initial biopsy (H&E, $\times 100$).
- E.** Follow-up CT image after 7 years from initial examination shows gradual increase of a central mass in RUL with endobronchial extension to posterior segmental bronchus (arrow).
- F.** Follow-up positron emission tomography-CT image shows high fluorodeoxyglucose uptake of the central mass (arrow) and distal obstructive pneumonia and atelectasis (*) of RUL.
- G.** A mixture of papillary, hemorrhagic, and sclerotic patterns is shown on final surgical specimen (H&E, $\times 10$).

ies supported that pulmonary sclerosing hemangioma arises from epithelial cells, probably the type II pneumocyte (4).

Typically, it occurs in middle-aged adults, with a predilection on females, who are usually asymptomatic until the time of diagnosis similar to this case (5). The tumor has well-defined margins, but not a definite capsule, and can grow expansively by compressing the adjacent lung parenchyma or bronchus. The CT findings show round or ovoid shape, a smooth margin, homogeneous attenuation, calcification, and strong early enhancement (6). In PET-CT scans, the majority of sclerosing hemangioma show increased FDG uptakes, the SUV_{max} of tumors ranged from 0.60 to 4.7 (median 2.30), and tumor ≥ 2 cm can frequently be falsely interpreted as malignancy (7). The tumor in our case shows intense FDG uptakes (SUV_{max} : 5.8). Single tumor localizations in the pulmonary parenchyma is a major concern. However, there are exceptional presentations such as endobronchial localization, which is extremely rare and mimics a slow growing low grade central lung malignancy such as carcinoid tumor. The reported doubling time of carcinoid tumor was 417 days and shorter than that of sclerosing hemangioma, which was reported to be 965 days (8, 9). The presence of sclerosing hemangioma inside a bronchus may not be a proliferation from the bronchus itself but a proliferation from peribronchial lung parenchyma invading adjacent bronchus (2). Histologically, the sclerosing hemangioma is a mixture of solid, papillary, sclerotic or hemorrhagic patterns. The proportions of these four components in the tumor usually vary, although one of them tends to predominate. Sugio et al. (1) reported that most sclerosing hemangioma cases exhibit a papillary pattern, while Devouassoux-Shisheboran et al. (10) reported that predominant endobronchial component by bronchial biopsy of this tumor was papillary and solid pattern.

The sclerosing hemangioma of our case exhibited endobronchial portion in the posterior segmental bronchus of RUL, and this tumor was initially misdiagnosed as a papillary adenoma. The initial specimen obtained by bronchoscopic biopsy contained only papillary patterns of the mass. The endobronchial mass biopsy obtained by bronchoscopy is widely used for the diagnosis of central lung lesions with endobronchial portion, regardless of benignity or malignancy as this method is less invasive and simpler than surgical biopsy. However, the bronchoscopic biopsy may not be a sufficient method for the pathologic

diagnosis of sclerosing hemangioma which has four different pathologic components. Nevertheless, a tiny specimen does not show the entire features of sclerosing hemangioma and may result in inaccurate diagnosis. Therefore, surgical methods, including wedge resection or segmentectomy, can exhibit entire features of sclerosing hemangioma and establish complete resection of the tumor. Furthermore, sclerosing hemangioma can grow and usually show high FDG-uptakes. Therefore, within such situations, results of limited biopsy specimens should be doubted.

Sclerosing hemangioma is a progressively growing benign tumor with high FDG uptakes and may have endobronchial portion which is extremely rare. Therefore, it can mimic slow growing central lung malignancy such as carcinoid tumor on imaging findings. Furthermore, this tumor is composed of four pathologic patterns: the papillary, sclerotic, solid and hemorrhagic components. Thus, the diagnosis by limited specimen through bronchoscopic biopsy may be insufficient. For accurate diagnosis, a complete surgical resection should be recommended in radiological suspicions of central lung malignancy or central sclerosing hemangioma with endobronchial extensions.

REFERENCES

1. Sugio K, Yokoyama H, Kaneko S, Ishida T, Sugimachi K. Sclerosing hemangioma of the lung: radiographic and pathological study. *Ann Thorac Surg* 1992;53:295-300
2. Boudaya MS, Falcoz PE, Alifano M, Camilleri-Broet S, Régnard JF. Endobronchial sclerosing hemangioma: a rare presentation of a parenchymal tumor. *Asian Cardiovasc Thorac Ann* 2008;16:57-58
3. Chong S, Lee KS, Chung MJ, Han J, Kwon OJ, Kim TS. Neuroendocrine tumors of the lung: clinical, pathologic, and imaging findings. *Radiographics* 2006;26:41-57; discussion 57-58
4. Devouassoux-Shisheboran M, Hayashi T, Linnoila RI, Koss MN, Travis WD. A clinicopathologic study of 100 cases of pulmonary sclerosing hemangioma with immunohistochemical studies: TTF-1 is expressed in both round and surface cells, suggesting an origin from primitive respiratory epithelium. *Am J Surg Pathol* 2000;24:906-916
5. Kim GY, Kim J, Choi YS, Kim HJ, Ahn G, Han J. Sixteen cases

- of sclerosing hemangioma of the lung including unusual presentations. *J Korean Med Sci* 2004;19:352-358
6. Chung MJ, Lee KS, Han J, Sung YM, Chong S, Kwon OJ. Pulmonary sclerosing hemangioma presenting as solitary pulmonary nodule: dynamic CT findings and histopathologic comparisons. *AJR Am J Roentgenol* 2006;187:430-437
7. 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
8. Oka S, Ono K, Kuwata T, Nagata Y, Baba T, Shigematsu Y, et al. [Surgical treatment for patients with pulmonary sclerosing hemangioma]. *J UOEH* 2011;33:41-45
9. Greengard O, Head JF, Goldberg SL, Kirschner PA. Pulmonary carcinoid tumors: enzymic discriminants, growth rate, and early age of inception. *Cancer Res* 1986;46:2600-2605
10. Devouassoux-Shisheboran M, de la Fouchardière A, Thiviolet-Béjui F, Sourisseau-Millan ML, Guerin JC, Travis WD. Endobronchial variant of sclerosing hemangioma of the lung: histological and cytological features on endobronchial material. *Mod Pathol* 2004;17:252-257

기관내시경 조직검사에서 유두상 선종으로 오진된 중심성 경화혈관종의 영상의학적-병리학적 소견: 증례 보고¹

김수현¹ · 선현주¹ · 송장현¹ · 박서연¹ · 김윤현¹ · 최유덕² · 송상운³

경화혈관종은 상대적으로 드문, 폐의 양성 종양 중 두 번째로 흔한 종양으로서 일반적으로 폐의 주변부에 위치한다. 중심성 경화혈관종은 극히 드물다고 알려져 있기 때문에 이의 영상의학적 소견에 대한 증례를 보고한다. 이 증례는 최초 기관지 내시경 생검 결과 유두상 선종으로 진단되었으나 영상의학적 검사에서 유암종 같은 중심성 악성 종양처럼 보였다. 수술적 절제를 시행하였고 경화혈관종으로 최종 진단되었다.

전남대학교 의과대학 전남대학교병원 ¹영상의학과, ²병리과, ³흉부외과