

Simultaneous Occurrence of Primary Pulmonary Racemose Hemangioma and Multiple Sclerosing Hemangioma - A case report -

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Abstract

Primary racemose hemangioma of the bronchial artery is a rare cause of repeated hemoptysis, and multiple sclerosing hemangioma of the lung is a rare benign pulmonary neoplasm. A 26-year-old male with repeated hemoptysis for one month was admitted to our hospital. An endobronchial polypoid cyst-like mass with spontaneous bleeding was detected on bronchoscopy and a very tiny pulmonary nodule in the same lobe was revealed on computed tomographic scan. The left upper lung was resected, and the simultaneous existence of racemose hemangioma of the bronchial artery shunting to pulmonary artery and multiple sclerosing hemangioma in the same side lung was histopathologically confirmed. In respect of pathogenetic association of both lesions the author reviewed previous literatures and report this very rare case.

Key words : Hemoptysis, Bronchial artery, Bronchoscopy, Sclerosing Hemangioma, Multiple,

INTRODUCTION

Hemoptysis is a common complaint in patients with respiratory diseases. To evaluate the cause of hemoptysis and localized bleeding points, various diagnostic procedures, including angiography, computed tomographic scan (CT scan), and bronchoscopy are performed. The bronchoscopic detection of endobronchial vascular anomalies in patients with hemoptysis is rarely reported in the literature.¹⁾ Sclerosing hemangioma of the lung is a relatively rare neoplasm presenting as a solitary nodule, but multiplicity of the lesions has reported to account for 4% of all cases.²⁻³⁾ A case of primary racemose hemangioma of the bronchial artery, with multiple sclerosing hemangioma incidentally

found on the concomittent performed CT scan, is reported here, as a confirmed diagnosis could be made histopathologically after the surgical resection. Primary racemose hemangioma of the bronchial artery should be taken into consideration in case of endobronchial bulding lesions with hemoptysis. If biopsy is performed, it will result in a massive endobronchial hemorrhage.

CASE REPORT

A 25-year-old man (non-smoker) had repeated hemoptysis for one month. He visited a few hospitals but the cause of hemoptysis had not been identified. On admission to this hospital, physical examination revealed decreased breathing sound with craking sound on left upper lung field. A chest roentgenogram (chest X-P) and CT scan showed infiltrative shadows and a very tiny nodule in left upper lung field, but there was no frank abnormalities in the blood vessels. (Fig.1)

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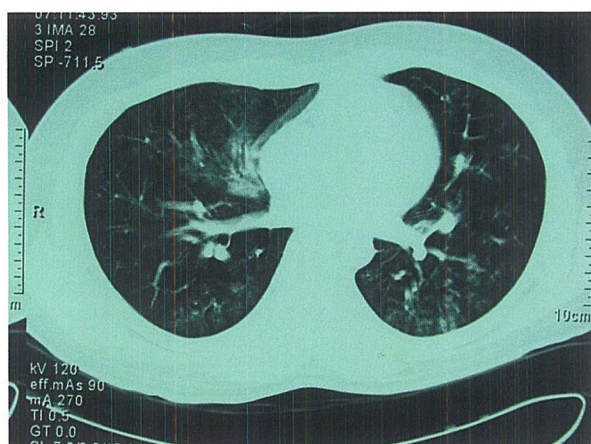


Fig. 1 Computed tomography scan (CT scan) shows infiltrative shadows and a very tiny nodule in left upper lung field, but there no frank abnormalities in the blood vessels.

Laboratory findings including blood cell count, serum biochemical analysis, anti-nuclear antibody (ANA) and anti-neutrophilic cytoplasmic antibody (ANCA) were normal. Bronchoscopic examination revealed the endobronchial polypoid mass nearly completely obstructing the orifice of left upper lobe. The mass appeared to be covered with hypervascular mucosa, with spontaneous bleeding. (Fig.2)



Fig. 2 Bronchoscopic examination revealed the endobronchial polypoid mass nearly completely obstructing the orifice of left upper lobe. The mass appeared to be covered with hypervascular mucosa, with spontaneous bleeding.

The bronchial mass was clinically diagnosed as bronchial

hemangioma, with multiple pulmonary nodules. He underwent left upper lobectomy in order to control the hemoptysis and confirm the histopathological diagnosis. The lobectomy specimen showed a well demarcated multilocular cystic mass measuring 1.0x0.8 cm in size, filled with blood and blood clots, nearly totally obstructing the bronchial lumen of the orifice of left upper lobe. (Fig.3A)

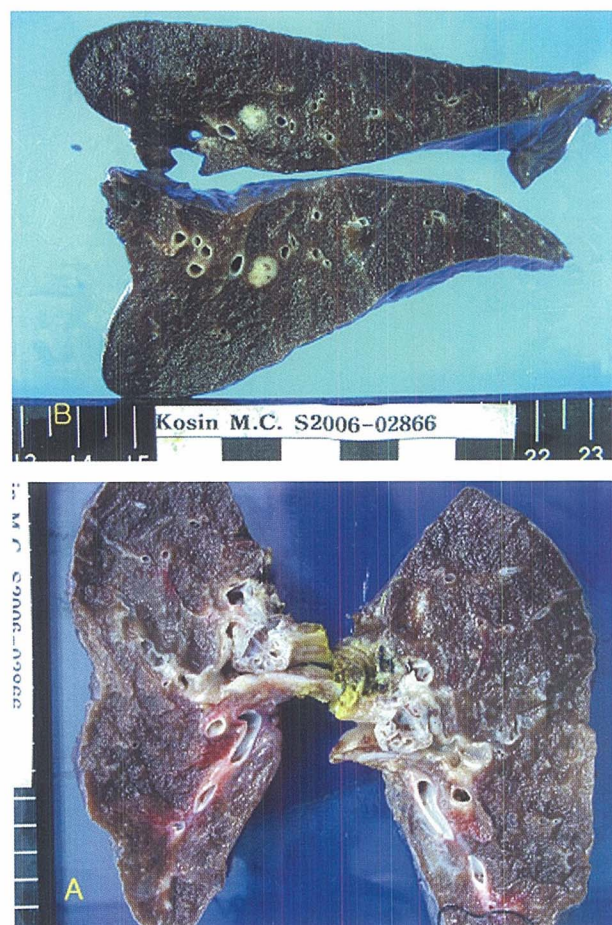


Fig. 3 A, The lobectomy specimen showed a well demarcated multilocular cystic mass measuring 1.0x0.8 cm in size, nearly totally obstructing the bronchial lumen of the orifice of left upper lobe. B, A small gray white solid nodule, 0.7 cm in greatest diameter, in lung parenchyma.

Parenchymal hemorrhage or necrosis was not evident. A small gray white solid nodule, 0.7 cm in greatest diameter, was seen in lung parenchyma. (Fig.3B)

Also two other smaller gray white solid nodules in left lower lobe were excised, 0.5 cm in greatest diameter, was

seen respectively. The solid nodules were well demarcated without encapsulation. Histologically the cystic mass was composed of convoluted thick-walled bronchial arteries directly interconnecting with dilated pulmonary artery (Fig.4).

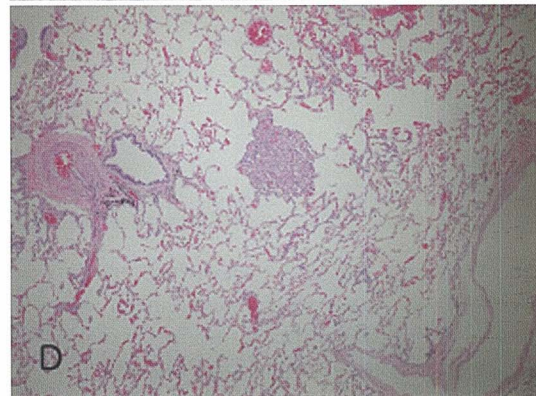
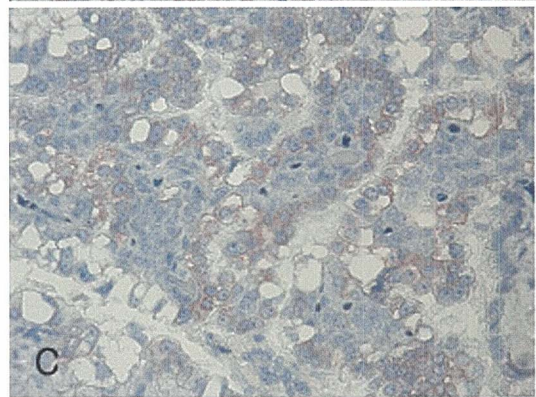
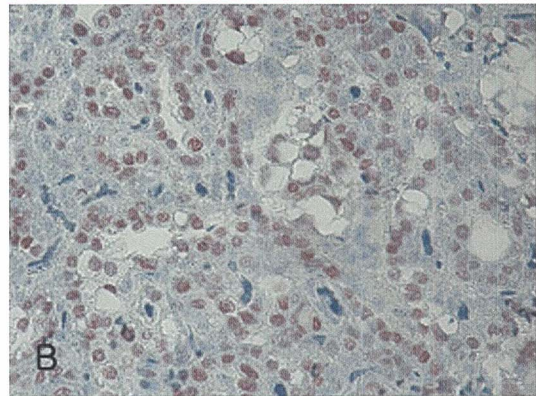
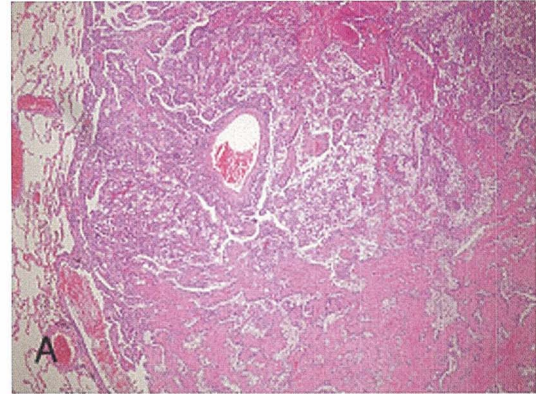
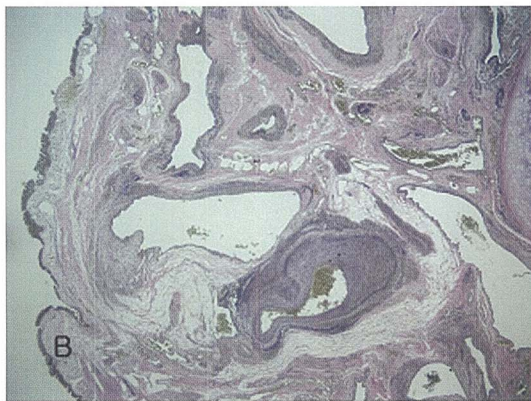
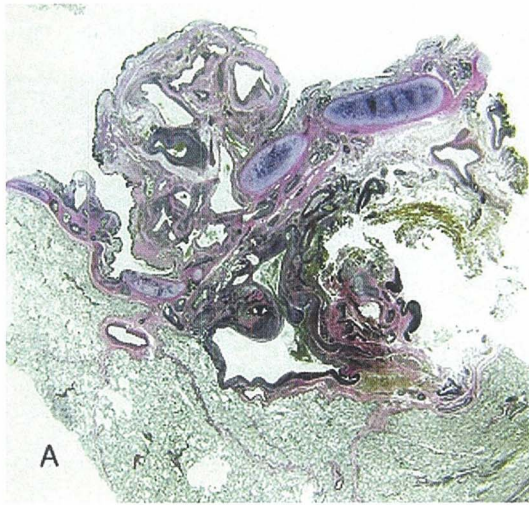


Fig. 4 The endobronchial cystic mass is composed of convoluted thick-walled bronchial arteries directly interconnecting with dilated pulmonary artery. (A, Elastic stain, whole mount scan; B, Elastic stain, x2)

The small solid nodules were composed of papillary architectures covered by cuboidal to columnar cells often with vacuolated cytoplasm, without lepidic growth of lining cuboidal cells.(Fig.5A)

Fig. 5 A, The small solid nodule composed of papillary architectures, shows two-type cells, lining cells often with vacuolated cytoplasm and round to polygonal cells in interstitium. (H-E stain, x2; inset, H-E stain, x200) B, Both surface cells and round cells of each solid nodules were diffuse immunopositive for TTF-1 in nucleus. (x400) C, Cytokeratin (MNFI16) reacted with the surface cells but did not reacted with the round cells. (x400) D, A very tiny nodules of micronodular pneumocyte hyperplasia, 0.7 cm from the racemose hemangioma of the bronchial artery. (H-E stain, x2)

Also noted were clusters or sheets of round to polygonal cells with uniform medium-sized polygonal nuclei and moderate amount of pale, eosinophilic or clear cytoplasm in the interstitial portion. Two of these nodules showed a thick-walled feeding vessel, respectively. Immunohistochemical study showed that both surface cells and round cells of each solid nodules were diffuse positive for TTF-1 in nucleus (Fig.5B) and EMA in cytoplasm, and negative for CD34, SMA and vimentin. Cytokeratin (MNFI16) reacted with the surface cells but did not reacted with the round cells in all nodules (Fig.5C). Both lining cuboidal cells and round cells showed no immunoreaction with p53 protein and the Ki-67 labeled proliferating index was less than 0.5 %, each solid nodules. The resected lung specimen also histologically showed a few very tiny nodules of micronodular pneumocyte hyperplasia, and one of which is 0.7 cm from the racemose hemangioma of the bronchial artery.(Fig.5D)

These findings confirmed the diagnosis of primary racemose hemangioma of bronchial artery for the bronchial cystic mass, and the diagnosis of multiple sclerosing hemangioma for the three pulmonary solid nodules. At one month after the operation, hemoptysis has not recurred.

DISCUSSION

Although hyperplastic changes in the bronchial arteries occasionally develop secondarily after inflammatory lung diseases such as bronchiectasis and bronchitis, hyperplastic changes in bronchial arteries without inflammation are to

be called primary angioma arteriovenosum racemosum.⁴⁻⁵⁾

Primary racemose hemangioma of the bronchial artery has been considered as inborn malformation, but its pathogenesis and natural behavior remain unknown. Primary racemose hemangioma is characterically composed of enlarged and convoluted bronchial artery shunting to the pulmonary artery or vein.⁶⁻⁷⁾ The majority of racemose hamangioma of the bronchial artery was detected as endobronchial polypoid lesions covered by normal mucosa on bronchoscopic examination.⁶⁾ In the present case, enlarged and distorted bronchial arteries formed the endobronchial polypoid mass and bronchial-pulmonary artery shunt was observed without inflammatory infiltrates in the resected lung specimen. These features are consistent with those reported for primary racemose hemangioma of the bronchial artery.

The bronchoscopic detection of endobronchial vascular anomalies in patients with hemoptysis is rarely reported in the literature. Park et al.¹⁾ classified three types of endobronchial vascular lesions as tubular bulding type, mass-like type and hemangioma type according to their bronchoscopic features. The present case showed racemose hemangioma of mass-like type, which felt soft and cyst-like. Some authors reported these endobronchial submucosal lesions poured a massive bleed upon biopsy.⁷⁾ Therefore if endobronchial vascular lesions as above on bronchoscopy is detected, there must be extreme caution to prevent fatal bleeding.

Pulmonary sclerosing hemangioma is thought to be rare benign neoplasm first described by Liebow and Hubell in 1956.⁸⁾ Histogically, sclerosing hemangioma is a mixture of four histologic patterns, such as papillary, solid, sclerotic and hemorrhagic components in typical cases. Each component of sclerosing hemangioma is composed of two populations of cells: cuboidal eosinophilic epithelial lining cells and solid-growing round to polygonal cells with pale to clear cytoplasm. Some authors have suggested that the origin of sclerosing hemangioma was mesothelial, mesenchymal and neuroendocrine derivation.⁹⁾ In recent

studies TTF-1 expression of both lining cells and round cells supported that these cells were derived from primitive respiratory epithelium or may represent poorly differentiated pneumocytes or Clara cells.³⁾ Most of the pulmonary sclerosing hemangioma are detected as a solitary nodule, but some papers have described multiple sclerosing hemangioma to account for 4-5% of all cases.²⁻³⁾ The possible pathogenesis of multiple sclerosing hemangioma has not been established. Although cases of atypical adenomatous hyperplasia (AAH) in continuity with multiple sclerosing hemangioma and alveolar adenoma with sclerosing hemangioma have been reported, whether the multiple lesions indicate multicentric origin of sclerosing adenoma or intrapulmonary metastasis, or incidental findings have not been verified.⁹⁾ In the literatures, it has not been fully studied whether the multiplicity of multiple sclerosing hemangiomas suggests a progressive clinical course or not. Although a solitary pulmonary sclerosing hemangioma with rapid progression was reported, several studies have showed that multiple lesions were slow-growing lesions.¹⁰⁾ In present case, three tiny sclerosing hemangioma of 0.5-0.7 cm size were distributed in upper and lower lobes of left lung. More tiny nodules were not detected in CT scan and in operation. One nodule of three nodules shows total papillary pattern without sclerosis or hemorrhage. Other two nodules show focal to total stromal sclerosis in papillary architectures. It has been unknown that the difference of stromal sclerosis in multiple lesions simultaneously detected in one patient is associated with pathogenetic significance and clinical course, including potential non-detectable nodules. Although all multiple lesions previously reported have been slow-growing cases, further analysis of a larger group of patients with a longer follow-up data is required.

Present case showed simultaneous detection of racemose hemangioma of bronchial artery and multiple sclerosing hemangiomas. In indexed medical literatures, the case showing the coexistence of racemose hemangioma of bronchial artery and multiple sclerosing hemangiomas has

not be described. Also it is unknown that the existence of one lesion is associated with occurrence of another lesion. Any syndrome are not associated with coexistence of both lesions or presence of one among two lesions. In present case, racemose hemangioma of the bronchial artery and the largest sclerosing hemangioma were in the same lobe, and other two sclerosing hemangiomas were in another lobe of the same lung. Also noted were a few nodules of micronodular pneumocyte hyperplasia very close to the racemose hemangioma of the bronchial artery. Although multifocal micronodular hyperplasia of lung is rarely found in patients with tuberous sclerosis complex,¹²⁾ whether a few foci of micronodular pneumocyte hyperplasia in present case are presumptive precursor lesion of multiple sclerosing hemangioma or are associated with tuberous sclerosis complex could not be verified. Although the simultaneous presence of a sclerosing hemangioma and racemose hemangioma of the bronchial artery in the same lobe and the presence of micronodular pneumocyte hyperplasia very close to racemose hemangioma of the bronchial artery indicate incidental findings as yet, further studies about its pathogenetic significance is required.

In conclusion, the author report an extremely rare case of simultaneous occurrence of primary pulmonary racemose hemangioma detected on bronchoscopy, multiple sclerosing hemangiomas and a few micronodular pneumocyte hyperplasia, surgically resected to control the repeat hemoptysis.

국문초록

기관지 동맥의 원발성 포도상 혈관종은 반복성 객혈의 드문 원인이며, 폐의 경화성 혈관종은 양성종양이며 드물게 다발성으로 발생한다. 26세 남자환자가 한 달간의 반복된 객혈을 주소로 본원에 내원했다. 기관지내시경검사서 우폐 상엽의 입구 기관지를 거의 폐쇄하는 낭종을 닮은 폴립양 종괴에서 다소 출혈이 있는 것을 관찰하였고, 컴퓨터 단층촬영 검사에서 같은 엽에 작은 결절이 관찰되었다. 좌폐우엽 절제술을 시행하였으며, 병리학적

으로 원발성 포도상 혈관종과 다발성 경화성 혈관종이 동일한 폐에 공존한다고 확진하였다. 두 질환의 병인학적 연관성에 관하여 문헌검토하였으며, 매우 드문 본 증례를 보고한다.

중심 단어 : 객혈, 기관지동맥, 기관지경, 경화성 혈관종, 다발성

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