Pulmonary Intralobar Sequestration Accompanied by Aneurysm of an Anomalous Arterial Supply

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A 47-year-old woman presented with hemoptysis and her chest X-ray films showed an opacity suggesting a mass in the left lower lung field. Based on radiographic investigations, the mass was diagnosed as an aneurysm develop in an anomalous vessel and was considered to be a Pryce type I pulmonary intralobar sequestration. Resection of the left lower lobes was performed and the aneurysm was found to be filled with thrombus. It is rare for an aneurysm to form in an aberrant vessel. This complication may have been the result of regional sclerosis affecting the anomalous artery as well as systemic atherosclerosis.

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Key words: Pryce type I, hemoptysis, thrombus

Introduction

Pulmonary sequestration is the term used when an aberrant artery of the systemic circulation system directly supplies an area of the lung parenchyma; this is believed to be a rare condition. This malformation was first reported by Huber (1) in 1777, and was later classified into 3 types by Pryce (2) in 1946, based on the pattern of pathology. Since the name pulmonary sequestration was given to this vascular anomaly, it has attracted clinical attention and an increasing number of case reports have been published in the literature.

Here, we report our recent experience with a case of pulmonary sequestration accompanied by a thrombotic aneurysm of anomalous vessels.

Case Report

The patient, a 47-year-old woman, had the chief complaint of hemoptysis. Both parents died of cerebral hemorrhage. For the past several years, she had suffered from hypertension but it was left untreated. She developed myocardial infarction in June 1988, and subsequently visited a general practitioner for treatment. Cerebral infarction occurred in January 1989. She had light hemoptysis in January and July of 1989, and then she was transferred to this hospital. Before the onset of myocardial infarction she had been obese as well as a

heavy smoker and drinker, with a daily intake of about 60 cigarettes and about 1,000 ml of rice wine (sake) for over 20 years. On admission, she appeared well-nourished, with an average physique. Left hemiplegia and dysarthria were present. There were no abnormal findings on physical examination of the chest and abdomen. Hematological tests on admission, including hemogram, biochemical tests, and serology tests showed no abnormalities.

On chest X-ray (Fig. 1), the frontal radiograph demonstrated an irregularly shaped mass (4.6×3.1 cm) with a relatively clear margin that overlapped the cardiac silhouette in the left lower lobe. Frontal tomograms confirmed the presence of the mass in the left lower lung (S¹⁰), and showed internal nonhomogenous attenuation of the lesion. An electrocardiogram demonstrated left ventricular hypertrophy with moderate ST-T changes. No relevant ECG record showing myocardial infarction was available. Chest CT scanning (Fig. 2), a computed tomographic (CT) scan of the chest disclosed an irregular mass abutting the descending aorta and heart. A nonhomogenous internal structure and partial calcification of this mass were suggested by internal mottling due to areas of decreased attenuation. Some parts of the mass were well enhanced by the injection of contrast material. Bronchography (Fig. 3), partial displacement and narrowing of the left basal bronchial tree due to pressure by the mass, but no filling defect or cyst formation was

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Pryce Type I Pulmonary Sequestration

Fig. 1. Chest X-ray film showing an irregularly shaped mass that overlapped the cardiac silhouette on the left.

Fig. 2. Chest CT scan showing an irregularly shaped mass abutting the descending aorta and heart.

found. Pulmonary angiography using DSA (Fig. 4), the pulmonary tree revealed a poor distribution of vessels in the left basal lung segment. Angiography of the thoracic aorta (Fig. 5), in the arterial phase showed an abnormal extension of the aorta into the left lower lung field at the 8th thoracic vertebral level, and further peripheral branching was also revealed. The venous return entered the lower pulmonary vein in the venous phase of the thoracic aortogram.

Based on these findings this case was diagnosed as intralobar sequestration of the left lung base, and the mass shown on the X-ray films was considered to be present in the anomalous artery supplying the sequestered lung. Therefore, thoracotomy was performed. During thoracotomy, an incision was made over the 6th rib, and

Fig. 3. Left bronchography showing partial displacement and narrowing due to pressure by the mass, but there were no filling defects or cyst formation.

Fig. 4. Pulmonary angiography (using DSA) showing a poor distribution of vessels in the left lower lung field.

the abnormal arterial supply leading into the left lower lobe and arising from the descending aorta was found at the T8 level. The vessel was 1.7 cm in diameter at its origin, and showed cystic dilatation where it entered the lung, forming an aneurysm 3.5 cm in diameter at the widest point. This anomalous vessel was excised at the root; the aortic site was patched and then the lower lobe of the left lung was resected.

Fig. 5. Thoracic aortogram showing an abnormal extension of the aorta into the left lower lung field, and the venous return which entered to the lower pulmonary vein.

Arteriography of the resected lobe including the anomalous vessel and the branch pulmonary artery was performed (Fig. 6). Arteriography of the anomalous artery (the lower vessel) revealed the presence of an aneurysm with irregular filling defects and then visualized the basal segment of the lung. Only A⁶ was enhanced by the injection of contrast medium into the branch of the pulmonary artery supplying this lobe (the upper vessel).

On observation of the cut surface of the resected lobe after fixation, no cystic or emphysematous change

Fig. 6. Arteriography of the resected lobe. Anomalous arteriography (the lower vessel) showing the presence of an aneurysm and the basal segment. Pulmonary arteriography (the upper vessel) showing only A⁶ lobe to be enhanced.

was seen in the basal lung segment. The inside of the aneurysm was filled with thrombus. Examination of the origin of the anomalous artery revealed distinct intimal

Table 1. The Case Reports of Pryce Type I Intralobar Pulmonary Sequestration in Japan

Case No.	Age and sex	Abnormal artery						Published year	Reference No.
		Location	Origin	No.	Size(mm)	Venous return	Aneurysm		
1	27 M	L BS	Thor. aorta					1956	5
2	34 M	L BS	Thor. aorta	1	8	LPV	(-)	1956	6
3	12 M	R S10	Thor. aorta	1	8	LPV	(-)	1971	7
4	28 F	R LL	Thor. aorta	1		LPV	(-)	1973	3
5	24 M	L LL	Thor. aorta	1		LPV	(-)	1973	3
6	18 F	L BS	Thor. aorta	1	10	LPV	(+)*	1975	8
7	6 F	R S9 + 10	Abd. aorta	1	10	UPV	(-)	1976	9
8	37 M	R S9 + 10	Thor. aorta	1	10	LPV	(-)	1978	10
9	5 M	L S10	Thor. aorta	1		LPV	(-)	1978	11
10	5 M	L BS	Thor. aorta	1	11	LPV	(-)	1979	12
11	9 F	R LL	Celiac a.	5	1-5	LPV	(-)	1982	13
12	26 M	L BS	Thor. aorta	1		LPV	(-)*	1983	14
13	1 F	L BS	Thor. aorta	1	8	LPV	(-)	1985	15
14	38 F	L LL	Abd. aorta	1	7	LPV	(-)	1987	16
15	53 M	L S9 + 10	Thor. aorta	1	15	LPV	(-)	1988	17
16	16 M	L BS	Thor. aorta	1	6	LPV	(-)*	1988	18
17	50 M	L BS	Thor. aorta	1	16	LPV	(-)	1988	19
18	61 M	L BS	Thor. aorta	1	7	LPV	(-)*	1989	20
19	13 M	L S10	Thor. aorta	1	8	LPV	(-)*	1990	21
20	20 F	L S8 + 9 + 10	Thor. aorta	1	8	LPV	(-)	1990	22
21	47 F	L BS	Thor. aorta	1	17	LPV	(+)*	Present case	

BS: basal segment, LL: lower lobe, LPV: lower pulmonary vein, UPV: upper pulmonary vein.

* Dilatation.

thickening due to severe atheromatous degeneration and destruction of the internal elastic lamina.

This patient did not have any pulmonary arterial branches in the left basal pulmonary segment, rather there was an aberrant artery which arose directly from the descending thoracic aorta. The bronchial system was normal. Therefore, Pryce type I intralobar sequestration of the left lung base was diagnosed in this patient. The thrombotic aneurysm that developed in the aberrant artery appeared as a mass on the chest X-ray films.

Discussion

In 1974, Pryce (2) classified this disease into 3 pathological types. The present case was a type I pulmonary sequestration, due to the abnormal arterial supply to normal lung tissue.

Pryce type I pulmonary sequestration is rarely seen, in contrast to types II or III (3, 4). As far as could be determined, only 21 cases have been reported (3, 5–22) in Japan, including the present case (Table 1). Six patients had anomalous arteries which were dilated after branching off from the systemic arterial supply (8, 14, 18, 20, 21), and only 2 of these (8) had an aneurysmal dilatation of the anomalous vessel. Furthermore, the present case was the only case where a cystic dilatation of the anomalous artery was filled with thrombus among the previous domestic case reports.

The world literature includes 400 case reports of pulmonary intralobar sequestration as reviewed by Savic et al (23), but none of these were similar to the present case. Only the 2 cases reported by Schuman et al (24) and McLaughlin et al (25), resemble the present case, although they were not Pryce type I lesions.

It has been thought that an anomalous artery exposed to the high blood pressure of the aorta is liable to undergo local changes, leading to regional sclerosis, and sclerosis localized in an abnormal arterial supply has been reported in many cases. However, one report (8) suggested that no lesions of the lung tissue occur as a result of the buffering effect of aneurysmal dilatation of the anomalous artery reducing systemic hypertension. There are only a limited number of cases of structural abnormalities in the adjacent lung tissue, even among the patients without dilated feeding vessels or those with an aberrant vessel dilated in a non-aneurysmal form. This suggests that the contribution of an aneurysm in the protection against potential lung injury can be ruled out.

The present patient was not elderly (47 years old), and no abnormalities of lipid metabolism or obesity were found on admission. However, she used to be obese, was a heavy drinker and smoker, and suffered from hypertension, myocardial infarction, and cerebral infarction. Also, the anomalous vessel showed severe intimal thickening due to atheromatous degeneration, destruction of the internal elastic lamina, and loss of the

intima and media of the aneurysmal wall with thrombus filling the lumen. These findings indicated that the aneurysm in the present patient had been formed by regional sclerosis in the anomalous feeding artery plus generalized arteriosclerotic changes.

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