Fig. 1. Pulumonary arteriography.
Pulumonary arteriography showing almost normal bifurcation of the right pulmonary artery without the image of A2 branch and a v-shaped filling defect in segment 2.

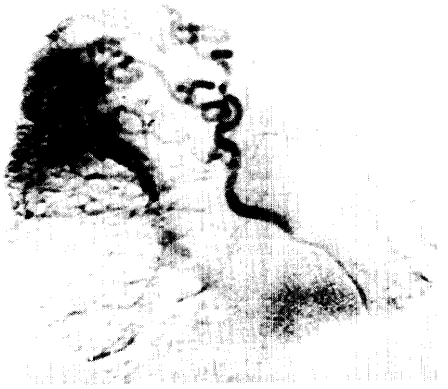

Fig. 2. Intercostal arteriography.
Intercostal arteriography showing a branch of a markedly distended tourtuous intercostal artery draining into the pulmonary trunk with a increased forming of wedge density.

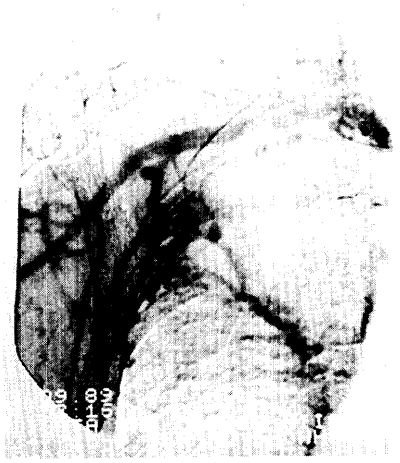

Fig. 3. Right subclavian arteriography.
Right subclavian arteriography showing an anomalous vessel branching from the right axilliary artery and draining into the tributary of the intercostal artery. This finding raises suspicion that this vessel blood is shortcircuited to segment 2.

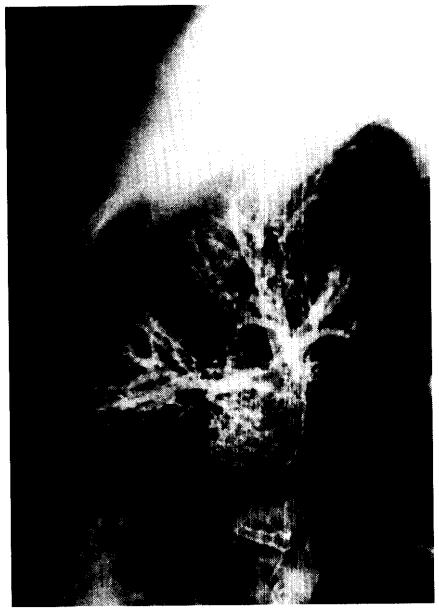

Fig. 4. Right upper lobe bronchography.
Right upper lobe bronchography showing that all bronchi in the right upper lobe are free from abnormalities.

Nonvisualization of the A2 arterial branch and v-shaped filling defect in segment 2 over the parenchymatous venous phase were depicted (Fig.1). Intercostalarteriogram revealed a branch of a markedly distended tourtuous intercostal artery draining into the pulmonary trunk with a formation of increased wedge density (Fig.2)

A right subclavian arteriogram showed an anomalous vessel branching from the right axilliary artery and draining into the tributary of the intercostal artery. This finding raised suspicion that this vessel blood was shortcircuited to S2(Fig.3). In a right upper

lobe bronchogram, all bronchi in the right upper lobe were free from abnormalities (Fig.4).

Arteriographic studies demonstrated a systemic arterial supply to the mass of normal lung tissue through vessels stemming from the intercostal and subclavian arteries in S2 of the right upper lobe of the lung with venous drainage into the pulmonary vein. These findings led to a diagnosis of quasi-intralobar pulmonary sequestration. In view of the patient's past history without hemorrhage or infection and current clinical findings, it consequently seemed appropriate to keep the patient under medical surveillance on an ambulatory basis.

DISCUSSION

In 1946 Pryce classificatied pulmonary sequestration into three types according to the presence or absence of abnormal lung tissue accompanied by an anomalous artery branching from the aorta (sequestered pulmonary segment), as follows: type 1, where there is no sequestered pulmonary segment and a portion of normal lung is supplied by an adjacent anomalous artery stemming from the aorta; type 2, both the sequestered pulmonary segment and adjacent normal lung are supplied by an anomalous artery; and type 3, the sequestered pulmonary segment is only supplied by an anomalous artery[1]. The present case corresponds to type 1 of Pryce's classification. The sequestered segment, through normal lung tissue, was supplied by an anomalous vessel of systemic arterial origin. However, the concept and the definition of pulmonary sequestration remain controversial. The majority of investigators adhere to the view that pulmonary sequestration is defined as a mass of lung tissue that is supplied by an anomalous vessel without communication of the tracheobronchial tree. Moreover, some investigators have insisted that type 1 of Pryce's classification should be excluded from sequestration of the lung. Heitzman advocated the concept of sequestration spectrum[2] based on his comprehensive elaborative studies of congenital developmental defects as: 1, anomaly of the pulmonary parenchyma; 2, anomaly of the bronchial system; 3, anomaly of the feeder artery; and 4, anomaly of draining vein. The present case showed anatomically normal bronchi by bronchography and the absence of a separate mass of lung tissue without bronchial communication. The pathological condition under investigation was thought to be a congenital anomaly characterized by a systemic arterial supply and anatomic as well as physiologic separation from adjacent lung tissue (corresponding to category 3 of Heitzman's classification above).

On the other hand, the plumonary arteriovenous fistula may be classified into two types, i.e. congenital and acquired. It has been postulated that a variety of pulmonary arteriovenous fistulae of the latter etiology occurs around an inflammatory lesion of the lung as a consequence of incomplete repair[2]. The pulmonary vascular disease in the present case seems very likely to be congenital in nature. A congenital pulmonary arteriovenous fistula rarely occurs in infancy or early childhood, but usually begins during the second and third decade of life, and is commonly diagnosed in the patient's thirties and forties. This implies that fistulous vessel walls become dilated with the advancement of age, and symptoms are relentlessly progressive[3]. The disease is also known to be associated with hereditary hemorrhagic telangioectasia (Rendu-Osler-Weber's disease) at a rate of 40% in a previous report[4]. In the present case, no signs of hemorrhagic telangioectasia on the skin or mucosa were noted. Thus, it is surmised that a fistulous communication between the right bronchial artery and the upper portion of the thoracic aorta was formed by some unknown causative agent. In normal development, the right bronchial artery connects the upper portion of the thoracic aorta and the upper intercostal arteries, the right bronchial artery extensive to the right subclavian artery, and the internal thoracic artery. All arteries are derived from the dorsal aorta during fetal life, but cease communications with the intrapulmonary vascular system at the periphery of the lung. These interconnecting channels functionally close in the course of normal development[8,9,10,11]. However, should such channels on the pulmonary artery close congenitally or postnatally, this could result in the compensatory development of anatomotical channels on the bronchial artery side through which to contribute to gas exchange[12]. In the present case, the bronchial tree appeared normal, implying that no anomalies took place during the development of the bronchi and their branching structures. Some shunting of arteries in the almost completely segment of normal lung tissue were probably formed by this process.

In cases of pulmonary arteriovenous fistula with an aberrant systemic artery but without associated pulmonary sequestration, hemoptysis may occasionally be the presenting symptom. In such an instance partial lobectomy becomes the choice of treatment for fear of copious hemorrhage. In the present case, in contrast, there was neither hemoptysis nor complications such as embolism and infections. Accordingly, a pulmonary arteriovenous fistula involving the systemic circulation is unlikely. Moreover, the dimi-

nution of peripheral resistance was slight, and there was virtually no increase in cardiac output. For this reason the patient was kept under medical surveillance without resorting to surgery, and his present condition remains favorable.

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