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A Case of Swyer-James Syndrome, with Reference to Pathophysiologic and Etiologic Aspect

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ABSTRACT

A 21-year-old male of Swyer-James syndrome is presented with clinical, angiographic, bronchographic and scintigraphic studies. He had a past history of recurrent bronchial asthma. A chest roentgenogram revealed unilateral hyperlucency of the left lung without any changes in volume. The small size of the pulmonary artery and its branches of the left lower lobe was visible by pulmonary angiography. Bronchography showed the bronchiectatic changes which consisted of moderate bronchial dilatation in an irregular beaded pattern with clubbed or tapered ends accompanied by absence of alveolar filling. A perfusion lung scan using ^{99m}Tc-macroaggregated albumin (^{99m}Tc-MAA) illustrated a perfusion defect in the left lower lobe. It was thought that recurrent pulmonary infections and asthmatic attacks were responsible for a pathological condition of this disease.

The term "Swyer-James syndrome" is used as a roentgenographic designation for the idiopathic unilateral hyperlucent lung[9,10]. There are two distinct schools of thought concerning whether vascular changes are primary[4,5,11] or that the hyperlucent lung is the result of primary parenchymal or bronchial disease[2,7,8,10].

This communication is concerned with unilateral hyperlucency involving the lower lobe of the left lung in a 21-year-old man who had been carefully examined with bronchography, pulmonary angiography and perfusion lung scan, and undergone left lower lobectomy.

CASE REPORT

A 21-year-old man was hospitalized to our institute on October 15, 1981 with the chief complaint of cough productive of a large amount of bloody sputum. He was well until 10 years old, when he suffered from respiratory embarrassment. After this episode of illness, the patient was followed with clinical and roentgenographic examinations and treated as bronchial asthma until entry. But he suffered repeated attacks of respiratory insufficiency. He had no history of smoking. Family history was not contributory.

Physical examination revealed a well developed, but chronically ill appearing man. The temperature was 37°C. Blood pressure was

128/80. A few medium sized rale were heard with deep inspiration in the left base of the chest. Pulmonary function studies revealed nothing of significance. The complete blood count, urinalysis and serological findings on admission were within normal limits.

Chest film and computed tomography showed a diffuse hyperlucency of the left lung accompanied by small hilar shadow and sparse intrapulmonary vascular markings. The affected lung was less reduced in volume (Fig. 1, 2).

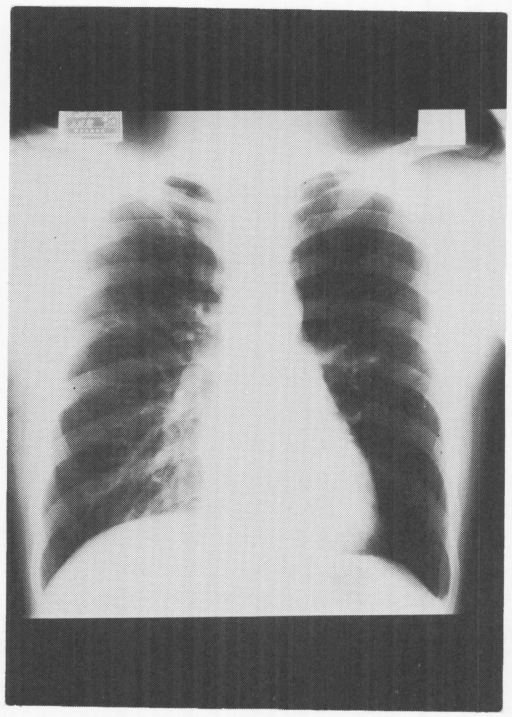


Fig. 1. Plain chest roentgenography showed unilateral hyperlucency of the left lung with small pulmonary artery and its branches.

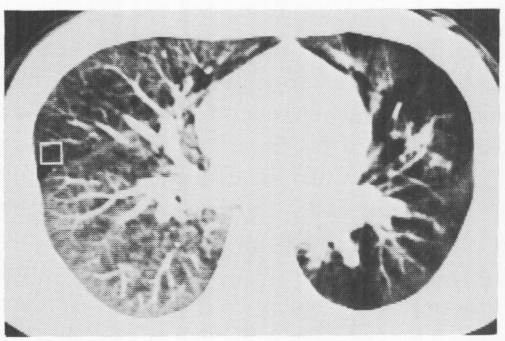


Fig. 2. Computed tomography of both lungs

A sequence of pulmonary angiograms revealed the inferior branch of left pulmonary artery to be markedly diminutive in size with very few, thin ramifications, in contrast, its upper branch was well demonstrated (Fig. 3). Also pulmonary angiography indicated that the volume of lower lobe of left lung kept normal. It was shown that bronchial artery circulation on the left lower lobe was markedly increased (Fig. 4). On aortography Takayasu aortitis was ruled out.

Bronchographically, the upper branch showed no evident bronchiectatic changes. But the bronchogram of left lower branch illustrated the usual findings in this case, with dilated, cylindrical bronchi, clubbing of some ends of smaller bronchi and lack of alveolar filling (Fig. 5).

A perfusion lung scan employing ^{99m}Tc-macroaggregated human serum albumin showed a marked reduction of blood flow to the lower part of the left lung (Fig. 6).

Since repeated severe hemoptysis failed to respond to various methods of treatment, the pulmonary lobectomy of the left lower lobe was performed with clinical improvement.

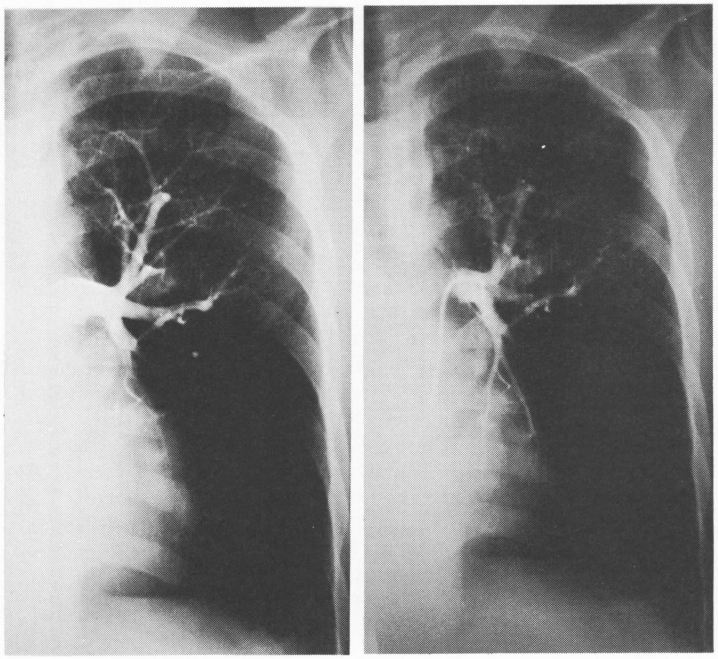
DISCUSSION

Swyer and James first described a 6-year-old child with unilateral pulmonary emphysema associated with a small pulmonary artery in 1953[10]. The most prominent clinical feature of Swyer-James syndrome is the presence of a unilateral hyperlucency on the chest reontgenogram which may involve a single lobe or the entire lung. Clinically the diagnosis of it is mainly roentgenographic and the following findings are usually evident[3,5]:

1) Abnormal bronchogram on the affected side or area which includes a) patent major bronchi b) small bronchi, mildly-dilated and beaded c) clubbing of some ends of smaller bronchi d) lack of alveolar and peripheral level filling.

2) Pulmonary angiography shows the narrowing of the branches of the pulmonary artery, the major pulmonary arteries being of normal size, and bronchial artery circulation increased on the affected site.

Since definitive pathologic studies of the unilateral hyperlucent lung are relatively few in number, the reason why the involved lung is less reduced in volume is still not completely known. According to the experimental study of Culiner



(a) (b)

Fig. 3. Selective pulmonary angiography showed the normal left upper branch (a), contrasting with small branch of left lower lobe (b).

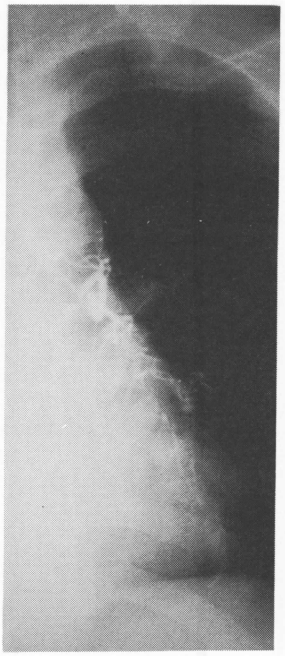


Fig. 4. Arterial supply of the left lung was contributed by the bronchial arteries which were considerably enlarged and hypertrophied.

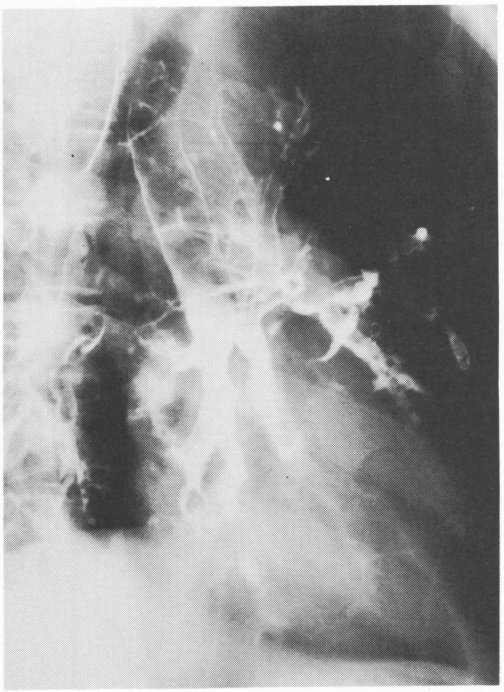


Fig. 5. Bronchography revealed the causative bronchial lesion, comprising moderate bronchial dilatation and an irregular pattern with clubbed or tapered ends.

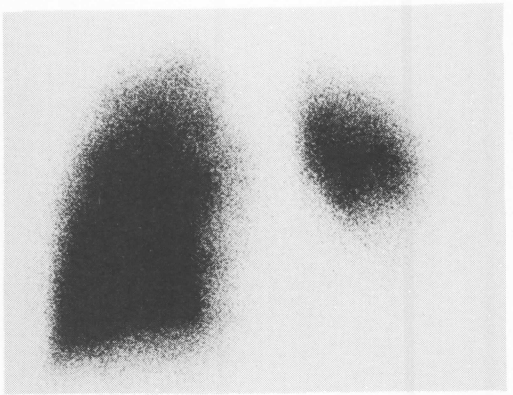


Fig. 6. ^{99m}Tc-MAA perfusion anterior scan showed a perfusion defect in the left lower lobe.

and Reich[1], some kind of check valve mechanism had occurred at the pores of Kohn and channels of Lambert, when the pulmonary pressure was raised over 40cm of water. Since no other causes of unilateral hyperlucency, such as obstructive or nonobstructive overinflation and pulmonary embolism were found in our case, it would seem conceivable that the basic lesion of

unilateral hyperlucent lung could be organic inflammatory changes on bronchi and bronchioles resulting from acute "bronchiolitis" in his childhood[6,8], followed by pulmonary air trapping and overinflation when air way pressure was raised during recurrent asthmatic attacks.

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