PULMONARY ARTERIO-VENOUS FISTULA

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SYNOPSIS

Pulmonary arterio-venous fistula is an uncommon condition, and up to 1955, only 150 cases have been documented in the literature. This paper describes three cases seen in Singapore. The first patient, a 9 year old Chinese boy, was detected to be cyanosed by his school doctor. Pulmonary angiography revealed multiple bilateral pulmonary arterio-venous fistulae in both lungs. He underwent thoractomy but succumbed to respiratory failure soon after the operation. The second patient, a 20 year old Chinese man, presented with recurrent fainting attacks. Pulmonary angiography showed multiple bilateral pulmonary arterio-venous fistulae. He has been treated conservatively and has remained well over a 4 year period. The third patient, a 15 year old Chinese man, was noted to have a rounded shadow in the lower zone of his left lung on routine radiography of the chest. Pulmonary angiography confirmed a solitary pulmonary arterio-venous fistula. The patient has refused operation and has remained well over a 7 year period.

The literature regarding pulmonary arterio-venous fistula is reviewed.

Ever since the first clinical diagnosis of a pulmonary arterio-venous fistula (PAVF) was made by Smith and Horton (1939) more than 30 years ago, this anomaly of the pulmonary vessels has been recognised more and more frequently. With the advent of angiography, confirmation of the diagnosis of PAVF is now possible in life. However, PAVF is an uncommon condition, and up to 1955, only 150 cases have been documented in the literature (Muir, 1955). In this paper, we report three further cases of PAVF as seen in Singapore.

CASE RECORDS

Case 1

A 9 year old Chinese boy was first seen in July 1965, after having been detected to be cyanosed by his school doctor. The patient has 5 other siblings and all of them, together with their parents, have been examined and were found to be clinically normal.

Clinical examination of the patient showed that he had mild cyanosis of both his fingers and toes, but no clubbing. No telangiectasia were seen. There was a continuous murmur heard over the sixth right intercostal space at the anterior axillary line. A soft systolic murmur was also heard medial to the right scapula and over the base of the right lung posteriorly. The electrocardiogram was normal. The skiagram of the chest showed a normal sized heart and multiple rounded opacities in the mid and lower zones of the right lung and in the mid-zone of the left lung (Fig. 1). The haemoglobin level was 19 gms.%.

A right-heart cardiac catheterization was done and the pressures recorded in mms. were as follows:- pulmonary capillary wedge—13 (mean), pulmonary artery—36/21, 28 (mean), right ventricle—36/0-5, right atrium—5 (mean). The systemic cardiac output was 4·3 L/min. and the right to left shunt was calculated as 2·1 L/min. The brachial artery oxygen saturation was 85%.

A pulmonary angiogram demonstrated the presence of multiple arterio-venous malformations in both the lung fields, mostly in the mid and lower zones (Fig. 2).

On 11.7.66, a thoracotomy of the right lung with excision of the fistulae was attempted. 3 large fistulae, 2 in the mid-zone of the fissure between the right upper and the right lower lobe and one situated medially over the mediastinal visceral pleura were found. The arterial and venous supply of each fistula was dissected out and ligated; the bronchus to each of these areas was also dissected and divided between ligatures, and each lung segment was stripped out to the periphery.

24 hours after the operation, the patient became very dyspnoeic as a result of respiratory failure. Despite resuscitative measures, he died soon afterwards. Histological examination of the dissected

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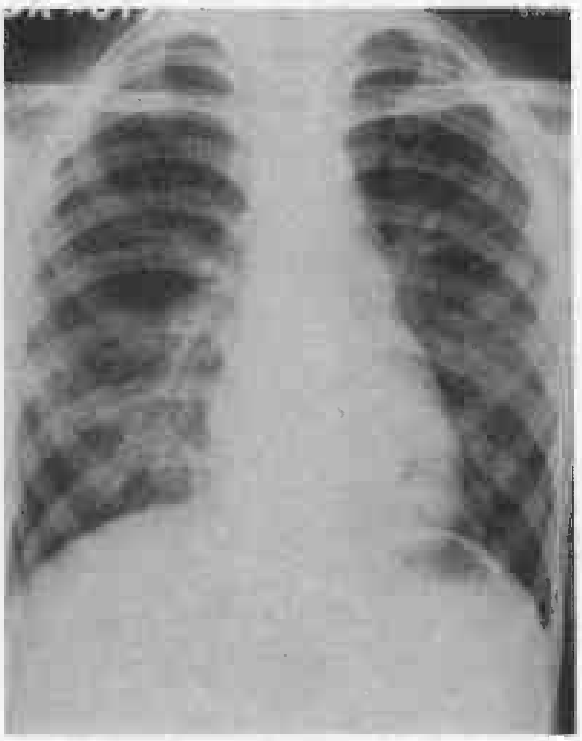

Fig. 1. Skiagram of the chest in Case 1, showing multiple rounded opacities in the mid and lower zones of the right lung and in the mid zone of the left lung (see text).

lung specimens showed changes consistent with pulmonary arterio-venous fistulae.

Case 2

A twenty year old Chinese man first presented in January 1968 because he experienced four attacks of fainting spells during that month. He also gave a history of being blue since birth. There was no history of haemoptysis or haematemesis. All his five other siblings and both his parents were examined and found to be normal.

Clinical examination revealed that he had a plethoric facies together with cyanosis of his fingers and toes. A few telangiectactic lesions were seen on the arms and over the anterior part of his chest. The blood pressure was 100/60. A systolic bruit was heard over the left lung posteriorly.

The chest radiogram showed that there were in the middle and lower zones of both lung fields a few nodular opacities of various sizes, with feeding vessels, thus suggesting the diagnosis of multiple pulmonary arterio-venous fistulae. The electrocardiogram was normal. The haemoglobin level was 22·8 gm.%, the packed cell volume was 70%, and both the total white and platelet counts were within the normal limits. A pulmonary angiogram done revealed a large number of arterio-venous malformations in both lungs below the level of the main pulmonary artery. Three large malformations were present, one at the right cardiac border, one at the left cardiac border and one near the bifurcation of the right main pulmonary artery (Fig. 3).

The patient has been followed up closely and has been keeping well. When seen recently, however, his haemoglobin level was 26·2 gms.%.

Case 3

A 15 year old Chinese boy was first seen in 1965. He had no symptoms, but a routine chest radiogram revealed a rounded opacity in the lower zone of the left lung. Clinical examination was completely normal except for a continuous murmur, heard best in the left axilla at the sixth intercostal space. This murmur was loudest in inspiration. The blood pressure was 90/60. The electrocardiogram and routine haematological investigations were all within the normal limits. In the skiagram of the chest, there was a rounded opacity in the lower zone of the left lung. Tomogram showed that this opacity was fed by a leash of vessels originating from the left hilum (Fig. 4). The patient was the only child and both his parents were unavailable for clinical examination.

A right-heart cardiac catheterization was done. The pressures recorded were as follows:- pulmonary capillary wedge—8 mms. (mean), pulmonary artery— 20/10 mms., 13 (mean), right ventricle— 20/0-4 mms. and right atrial —5 mms. (mean). The systemic cardiac index was 3·0 L/min./M^2. The brachial artery oxygen saturation was 98%. Selective pulmonary angiogram (Fig. 5) showed a large artery supplying an arterio-venous malformation, which corresponded to the rounded opacity seen in the chest radiogram.

The patient refused operation and has been followed up regularly. When seen recently, he was still keeping well with no symptoms at all.

DISCUSSION

Pulmonary arterio-venous fistula (PAVF) is a condition where there is one or more direct communications between the pulmonary arterial and venous systems. These haemangiomatous malformations of the pulmonary vascular bed are also known variously as cavernous haemangioma of the lung, congenital pulmonary telangiectasia and pulmonary arterio-venous aneurysm. The term PAVF probably is the most appropriate because it highlights the afferent arterial and efferent venous connection to the malformation seen characteristically in this condition.

The incidence of PAVF is low although there have been increasing reports in the literature. In one large centre (New York Hospital—Cornell Medical Centre), only 9 cases were collected in a

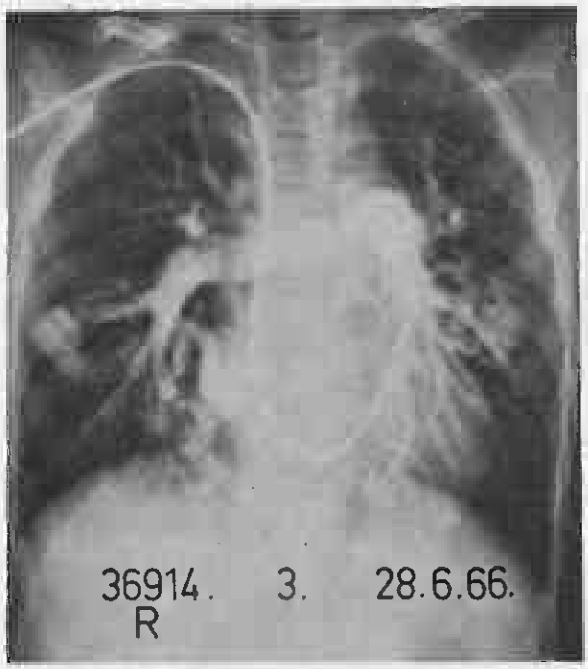

Fig. 2. Pulmonary angiogram of Case 1, showing multiple arterio-venous fistulae in both the lung fields, mostly in the mid and lower zones (see text).

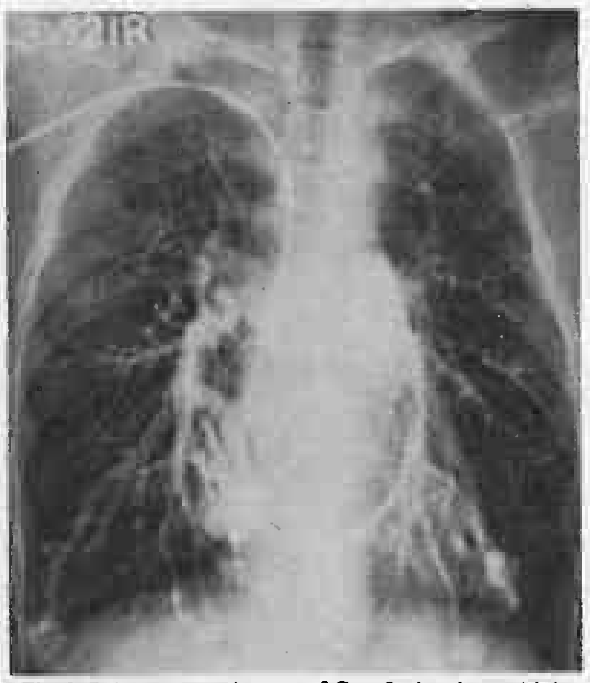

Fig. 3. Pulmonary angiogram of Case 2, showing multiple arterio-venous fistulae in both the lung fields (see text).

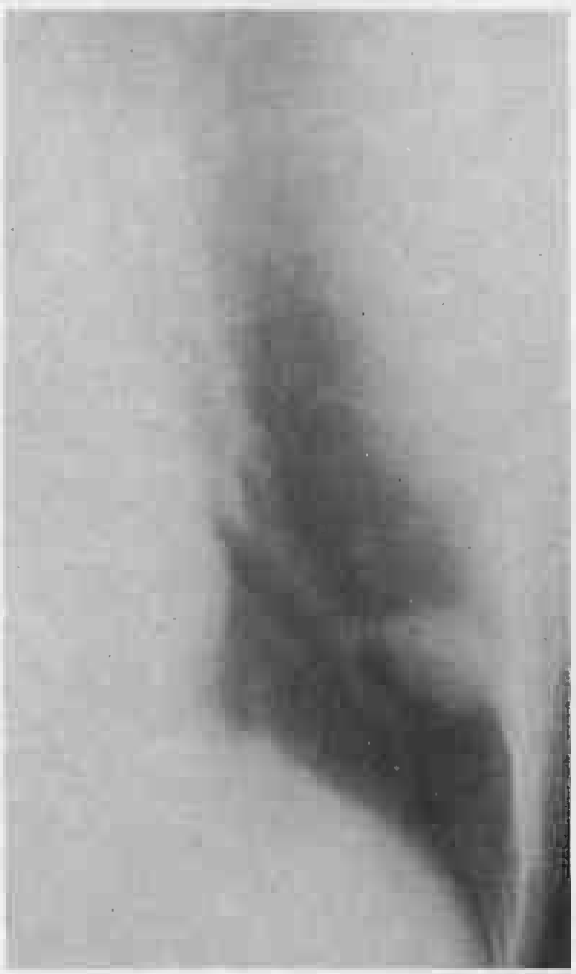

Fig. 4. Tomogram of the left lung in Case 3, showing a rounded opacity in the lower zone, fed by a leash of vessels originating from the left hilum (see text).

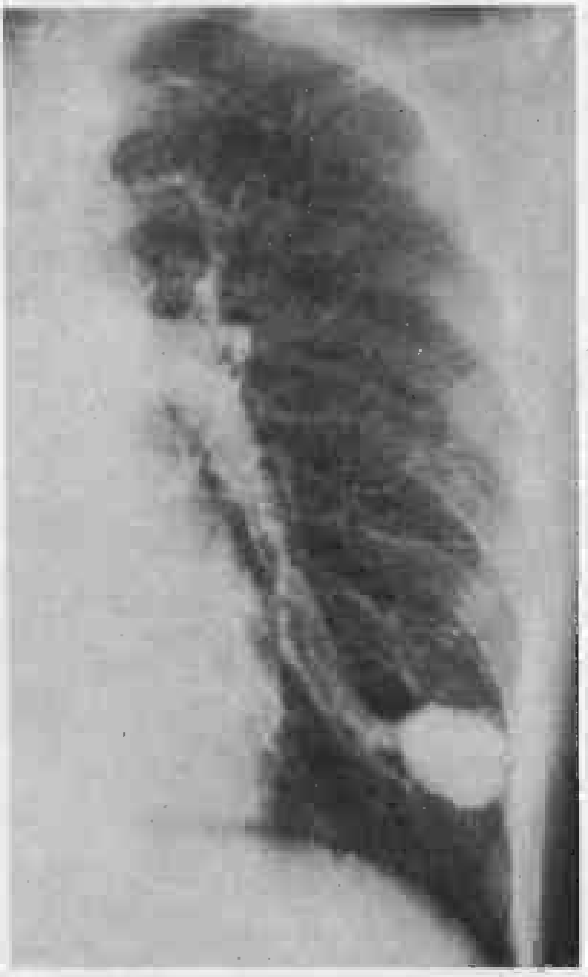

Fig. 5. Pulmonary angiogram in Case 3, showing pulmonary arterio-venous fistula in the lower zone of the left lung (see text).

total of 2,000 patients studied angiographically over an 8 year period (Steinberg and McClenahan, 1955).

There is often a familial tendency in PAVF and cases have been described in siblings (Moyer and Ackerman, 1948) and in parent and child (Tobin and Wilder, 1953). However, in our two cases (Cases 1 and 2) none of the immediate family members examined clinically were found to be affected. A strong association with hereditary haemorrhagic telangiectasia (Rendu-Osler Weber Disease) is also frequently noted. Thus, in the literature, 15% of patients with PAVF have a familial history of this disease (Sloan and Cooley, 1953). Furthermore, 40% of reported cases of PAVF will show telangiectatic lesions elsewhere in the body, as is seen in Case 2.

The clinical presentation of PAVF depends largely on the size and the number of lesions in the lungs. Cyanosis (seen in Cases 1 and 2) is present if there is a significant shunting of venous blood in the lungs. As a result of the anoxaemia, secondary polycythaemia may result as is seen in Cases 1 and 2, where the haemoglobin levels were 19 gms.% and 26 gms.% respectively. Haemoptysis due to rupture of the PAVF into the adjoining bronchi and dyspnoea are also seen. Although symptoms may occur in childhood (seen in Case 1) and infancy, they classically manifest during early adult life (seen in Case 2).

Neurological disturbances such as fainting attacks (seen in Case 2), dizziness, diplopia and motor weakness are commonly encountered and have been attributed to cerebral anoxaemia. Hemiparesis may also occur as a result of cerebral thrombosis, cerebral abscess or paradoxical embolus (Kaplan, 1968).

Clinical examination of the heart often reveals normal findings, except for a systolic or continuous bruit heard over the site of the fistulae. The cardiac size and electrocardiogram are frequently normal, due to an absence of increased pulmonary vascular resistance seen in this condition.

The radiological appearances of the chest depend upon the number and the size of the PAVF present. Localised anomaly are seen more commonly in the lower zone of the right lung. Large fistulae are easily identified in the chest radiogram as multiple opacities with vascular shadows radiating to the hilar (Shumacker and Walhausen, 1963). Small lesions which are missed in the chest X-ray, may however be identified by tomography of the lungs (Lodin, 1952).

Right heart catheterization nearly always reveals normal findings. Pulmonary angiography is the single most important investigation, as it not only confirms the presence of PAVF, but also demonstrates the site, number, extent and distribution of the lesions (Seaman and Goldman, 1952).

The treatment of choice for PAVF is excision of the lesion, with conservation of as much normal lung tissue as possible during the surgical procedure. Successful operation corrects the anoxaemia, cyanosis and polycythaemia present, and also prevents the occurrence of cerebral abscesses. There are many reports in the literature regarding successful surgery in this condition (Goldman, 1947; Hepburn and Dauphiner, 1942; Burchell and Clagett, 1947). However, cases have been reported where pre-existing PAVF increased in size after excision of associated lesions. From the literature, it would appear that the best chance of a cure lies in the solitary lesion. In Case 1, multiple large PAVF were present. The patient underwent surgery where three large PAVF in the right lung were resected. Unfortunately, he developed respiratory failure 24 hours after the operation and succumbed to this complication.

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