PULMONARY VASCULAR SHUNTS

REPORT OF TWO CASES

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Two patients with pulmonary vascular abnormalities discovered as a result of a routine chest radiograph, are reported. The first patient had generalised hereditary telangiectasia whilst the second patient was found to have a communication between the bronchial and pulmonary arterial circulation.

CASE REPORT

CASE 1

Miss G.P., age 48 years, first had a chest radiograph taken in 1950 following an attack of post-herpetic neuralgia. This revealed two opacities in the right lung. She received no treatment. A radiograph taken two years later showed that the lesion had persisted (Figs. 1 & 2), and she was referred for operation.

The past history revealed a tendency to nosebleeds both as a child and as a young adult.

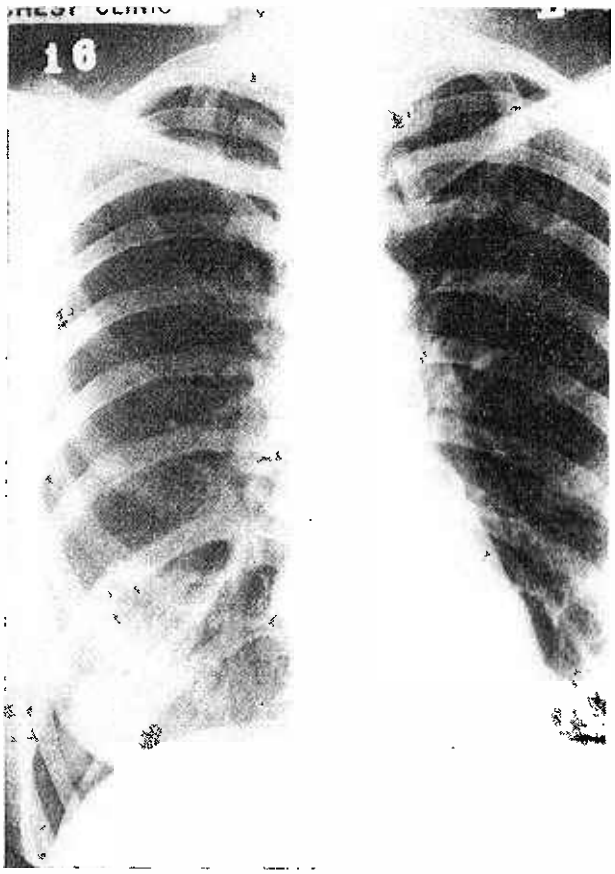

Fig. 1. One lesion has been outlined.

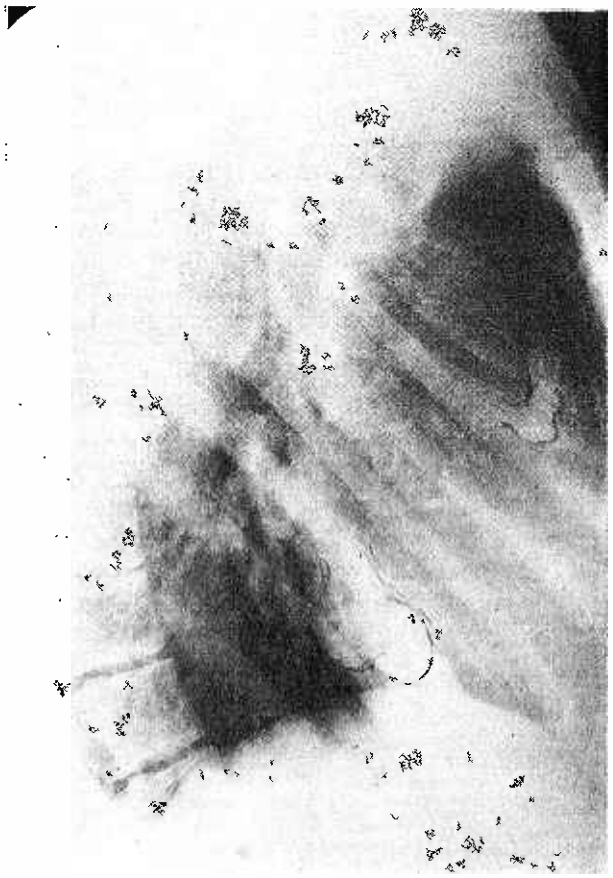

Fig. 2. Two lesions have been outlined.

Her father had dilated vessels over his face and a similar tendency to epistaxis. He died at the age of 65 years from prostatic obstruction. A post-mortem examination records that a "blood cyst" was found in one lung although no additional information was noted.

On the 9th of November 1953, the patient was admitted to the Thoracic Unit, the Royal North Shore Hospital of Sydney. Telangiectatic spots on the cheeks, palate and tongue were noted. No cyanosis, plethora or finger clubbing was present and the blood pressure was normal.

A machinery murmur was heard over the anterior lower third, the axillary, and the posterior areas of the right lung. The breath sounds were normal although the air entry was diminished anteriorly.

All other systems were normal.

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The blood count was as follows:

Haemoglobin	11.1 g/100 ml.
Total white cell count	6,400/c.mm.
Sedimentation rate	5 mm. per hour.

The differential count was within normal limits. The Mantoux Test was negative. Bronchoscopy revealed no abnormality.

A diagnosis of pulmonary arteriovenous fistulae associated with multiple familial telangiectases was made.

On the 18th November 1953, a right posterolateral thoracotomy was performed and two vascular tumours were found—one situated in the anterior segment of the right upper lobe and the other in the anterior basal segment of the right lower lobe.

The main pulmonary artery was isolated and temporarily occluded. The superior and inferior pulmonary veins were similarly controlled. The vein draining the upper lobe fistula, which was dilated and tortuous, was ligated and divided. The distal end of the vein was then followed peripherally to its junction with the dilated fistula and the sac was excised with a small wedge of lung tissue. Its arterial supply was then clamped and ligated.

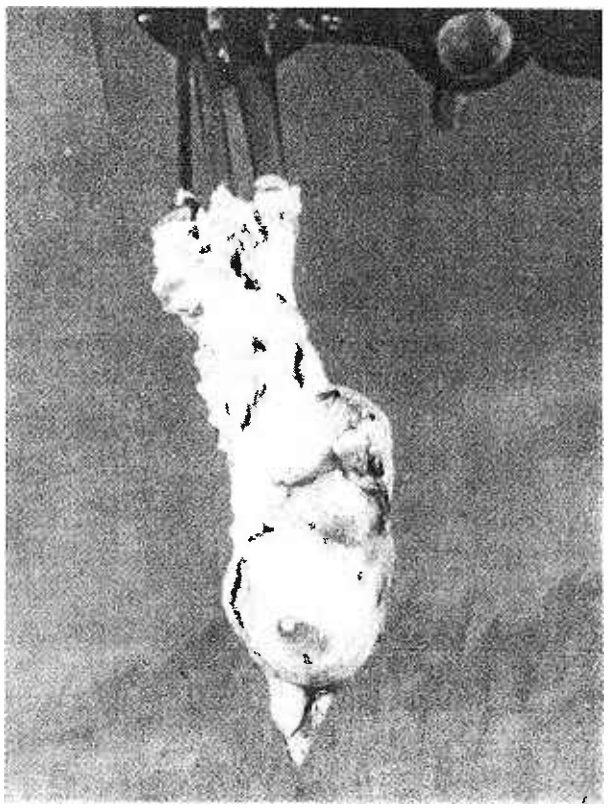

Fig. 3. Case 1. The vascular tumour was injected with formol-saline. When distended it measured 9 x 3 centimetres.

In the lower lobe, the artery to the sac and the adjacent venous tributary were found to be enlarged vessels of the anterior basal segment. The artery was ligated and divided first, then the vein. After securing these two vessels they were dissected peripherally to the dilated sac which was removed in continuity (Fig. 3). When the removed specimen was distended with fluid it was nine centimetres long and averaged three centimetres in width.

After the operation, recovery was uneventful.

The patient was seen 6½ years later in March 1960. Her chest was radiologically clear.

CASE 2

Miss C.B., 17 years old, was found to have a coin shaped opacity in the right lung following a mass x-ray in October 1959 (Fig. 4). A few months earlier she had been feeling more tired than usual. She was admitted to the Manly District Hospital and a Mantoux Test was found to be positive. The sputum was negative on direct smear and on culture on several occasions.

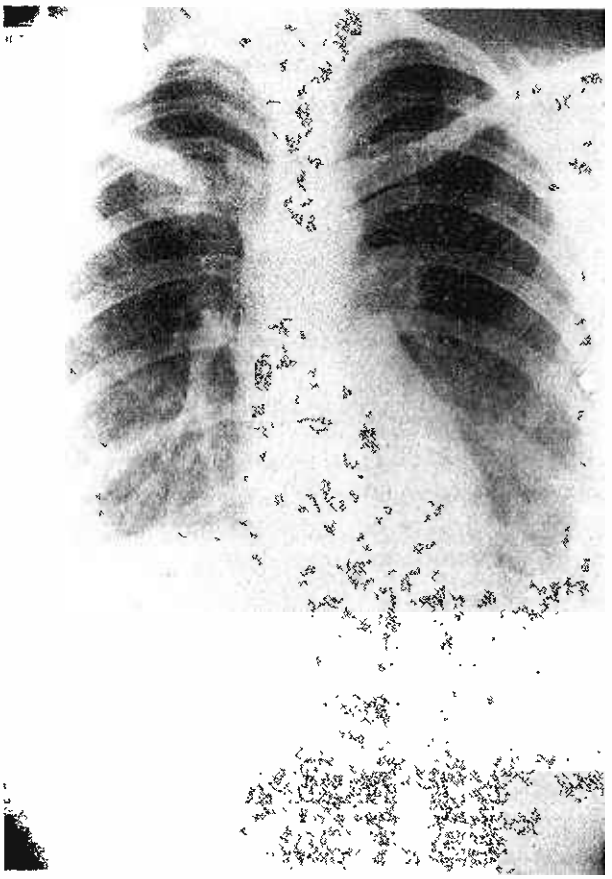

Fig. 4. Case 2. Opacity discovered in Mass X-ray.

Because of the positive Mantoux test and the X-ray appearance, a course of anti-tuberculous drugs was given over a period of three months.

The lesion remained unchanged so she was referred for operation.

There was no family history of epistaxis or generalised telangiectases. However, her mother had died suddenly at the age of 29 years and was said to have had congenital heart disease, although no details could be obtained.

The patient's general condition was good, there was no cyanosis or clubbing and no telangiectases were noted. No abnormality of the cardiovascular system or the lungs was found.

The blood count was as follows:

Haemoglobin	13.2 g/100 ml.
Total White Count	8,500/c.mm.
Sedimentation Rate	12 mm. per hour.

The differential leucocyte count was within normal limits.

On the 22nd February 1960, the chest was opened through a right postero-lateral thoracotomy. The upper lobe was freely mobile. A pulsating globular mass, about 2½ cms. in diameter, was found in the posterior segment of the upper lobe. It was situated deeply in the lung. Beneath the visceral pleura on the axillary surface of the upper lobe were some dilated vessels.

A large tortuous bronchial artery, about the size of the radial artery in an adult, became dilated and aneurysmal behind the right main bronchus. This dilatation of the bronchial artery measured about one and one half centimetres in diameter and two centimetres in length. On further dissection, the bronchial artery was found to continue into the posterior segment of the upper lobe. Its further course could not be followed because of the severe bleeding induced, so the posterior segmental artery was exposed. Pressures were then taken by inserting a 20 guage needle into the pulmonary artery, the aneurysmal dilatation of the bronchial artery, and the pulsating mass situated in the substance of the posterior segment of the right upper lobe.

The pressures recorded in these three areas were the same, i.e. twenty-five millimetres sytolic and ten millimetres of mercury diastolic pressure. Occlusion of the posterior segmental and bronchial arteries, both separately and simultaneously, did not alter the pressure in the pulsating tumour, indicating the presence of further communications within the lung. On the other hand when the intra-pulmonary vascular tumour was compressed between finger and thumb, a rise in pressure in the bronchial artery from twenty-five up to fifty millimetres of mercury was recorded. These observations demonstrated the existence of a shunt between the systemic (bronchial) and the pulmonary arterial systems.

The right middle and lower lobes were normal. No subpleural telangiectases were seen.

As a result of these findings, an upper lobectomy was undertaken in preference to a more localized resection and from this operation, her recovery was uneventful.

The resected right upper lobe was investigated by injecting lipiodal into the posterior segmental artery. An X-ray taken of the specimen showed that both the pulmonary and bronchial arteries communicated with each other by a large tortuous sac. (Fig. 5). Lipiodal was then run into a pulmonary vein and an X-ray showed that no communication could be demonstrated with the varix or sac (Fig. 6).

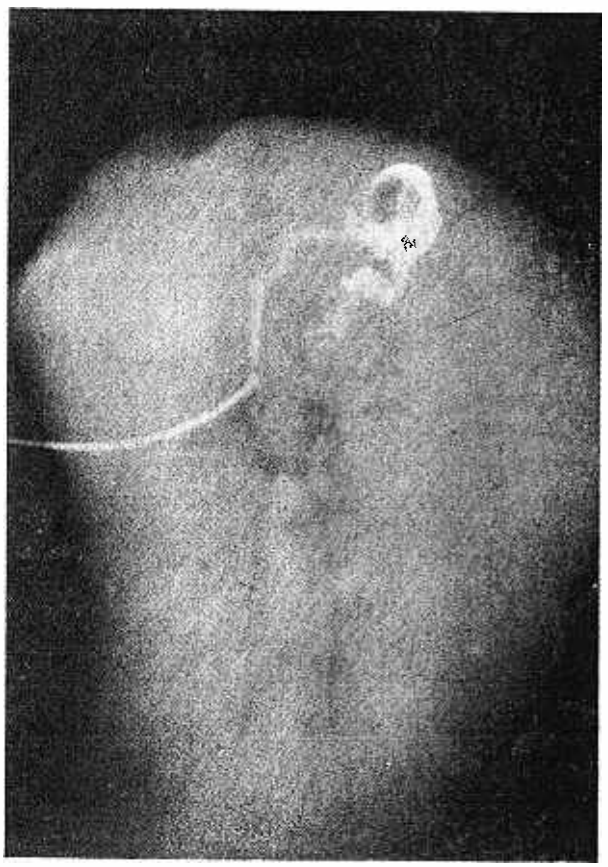

Fig. 5. A cannula has been placed in the posterior segmental artery and lipiodol injected to demonstrate the communication between the two arterial systems.

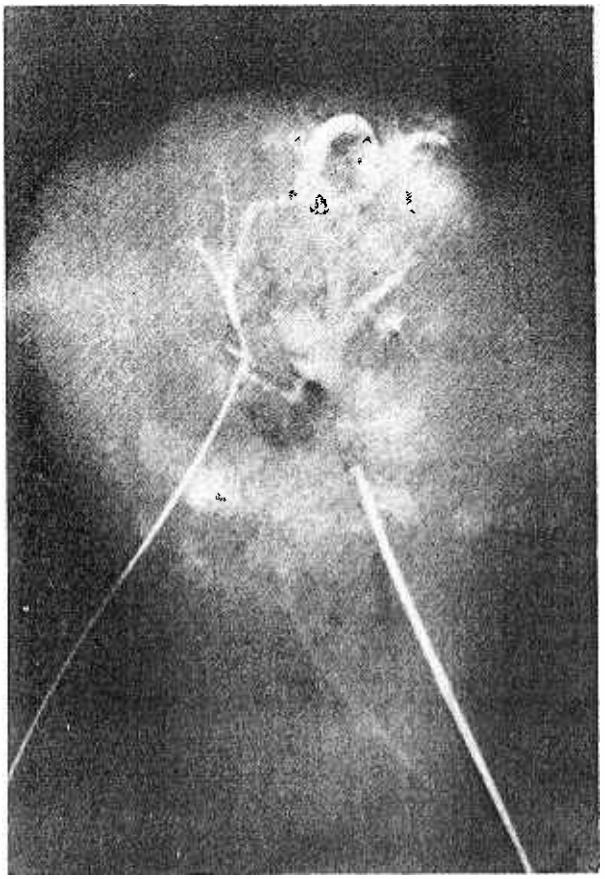

Fig. 6. The fine cannula is in the artery and the larger in the vein.

DISCUSSION

Certain anatomical observations have been recorded which are relevant to a discussion of these cases.

The existence of precapillary anastromatic channels was described in 1940 by Van Hoyek, and Tobin and Zorequiery (1950) demonstrated these arterio-venous anastomoses in the normal lung by the injection of glass spheres of graduated sizes into the pulmonary artery. Direct pulmonary arterio-venous communications up to 500 μ were found at the level of the lobular bronchovascular segments. Smaller shunts of 50 to 100 μ were present at the level of the smaller bronchi and respiratory bronchioles and those of 20 to 50 μ were found near the alveolar sacs and alveoli. Hales (1956) and Rydell et al. (1956) found in their cases small multiple pulmonary arterio-venous communications scattered throughout the substance of both lungs.

Hayward and Reid (1949) described a case in which the removed specimen consisted of a large sac supplied by an artery and drained by a vein being "a huge peripheral vessel in the distal capillary bed". Their patient had generalised telangiectasia which was familial. They pointed out that the pathology of the lung lesion was the same as that occurring in other parts of the body and was the pulmonary component of a generalised disease. Direct communications (or fistulae) therefore exist between the pulmonary arteries and veins at pre-capillary level as well as at the periphery.

Marchant et al. (1950) demonstrated, by means of bronchovascular casts, that there were anastomoses between the bronchial arteries and the pulmonary arteries at pre-capillary level. In the normal lung the bronchial artery becomes widened and tortuous near the site of anastomosis and gives off a branch which enters the pulmonary artery against the direction of its blood flow. They were able to demonstrate that these anastomoses enlarge considerably in pulmonary disease.

In 1936 Bowers reported a vascular tumour in the lung which he described as a haemangioma, and since then other abnormalities have been described. In one type of case blood passes from the pulmonary arterial to the pulmonary venous system without oxygenation and a right to left shunt of blood exists. In the other type of case a communication exists between the bronchial and pulmonary arterial systems, so that there is a left to right shunt of blood.

Bosher et al. (1959) analysed 350 cases and noted that a right to left shunt of blood was present in 338 patients. These lesions may be situated on the surface or deep in the lung. They may be single or multiple but confined to one lobe, or they may be scattered diffusely throughout both lungs.

The other type of case is very rare. There is a left to right shunt of blood through a communication between a systemic artery and the pulmonary circulation. Claiborne et al. (1956) reported a case in which they were able to demonstrate that the oxygen saturation of a sample of blood from the right pulmonary artery, where the fistula was situated, was higher than a sample from the left pulmonary artery. Bosher et al. (1959) collected 12 more cases. In 9 of these cases the anastomotic channel arose from the aorta, in two the intercoastal artery, and in one the pericardiophrenic artery. In such cases, systemic arterial oxygen saturation is normal. This type of fistula resembles a patent ductus arteriosus and it is conceivable that pulmonary hypertension could develop if the shunt were sufficiently large.

The mechanism of development of these abnormalities is unknown. When generalised telangiectases are present, the condition may be familial. It has been observed that some lesions

remain static for a long time following their discovery on routine chest film. Other lesions develop rapidly as indicated by progressive cyanosis and incapacity, and by the appearance of new lesions in serial radiographs. Cherbon, Adams and Carlson (1952) reported a case which developed three new lesions in the right lung following a left pneumonectomy for multiple arteriovenous fistulae five years after operation. The patient became so incapacitated that, seven years later, further surgery was performed on the remaining lung. Similar experience were recorded by Wodehouse (1948), Baer et al. (1950), Parker and Stallworth (1952), and Ronald (1954).

Progressive enlargement of the fistula with an increase in the shunt is a common feature of the condition. Maier et al. (1948) estimated that the shunt in their case was 58% of the cardiac output. In cases with large right to left shunts cyanosis may be severe and compensatory polycythaemia will occur. With the development of polycythaemia a tendency to thrombosis occurs in the peripheral vessels, notably the veins of the legs (Maier, 1948). Cerebral symptoms such as headaches, dizziness, fainting, thickness of speech, epileptiform seizures and hemiplegia may result from thrombosis of the cerebral vessels. A cerebral abscess has been reported by Wodehouse (1948) and a purulent meningitis by Hales (1956). It has been suggested that infection of the brain may occur either because of the growth of bacteria in an area of cerebral softening or from a cerebral embolus (Wechsler and Kaplan, 1940). The presence of a large right to left shunt may provide the passage of a paradoxical embolus.

Many patients are asymptomatic and have been discovered because of a routine radiograph. The presence of a sheaf of vessels leading to an opacity may suggest the diagnosis and this is confirmed if a murmur is heard over the site of the lesion. Angiocardiography has been used to establish the presence of a fistula and to more accurately localise it, although definite hazards to life are associated with its use.

Other cases may present with exertional dyspnoea, cyanosis, clubbing of the fingers and toes, and polycythaemia. The heart is usually normal or slightly enlarged, but no cardiac murmurs can be heard. In the absence of a detectable murmur some patients have been treated as having polycythaemia vera (Baer, 1950). In 1936 Hirsch, in an article entitled "Pulmonary Changes in Polycythaemia Vera", commented that the pulmonary shadows in a case of multiple arteriovenous fistulae confirmed at post mortem and reported by Rodes in 1938 had nodules "similar to those I have described".

Many of the cases reviewed by Bosher and his colleagues (1959) had hereditary telangiectasia or Osler-Weber-Rendu Disease. In those patients with a single arteriovenous lesion in the lung 36% had generalised telangiectasia, and in those with multiple lesions in the lungs 57% had generalised disease.

The lesion in the lung may bleed. Baer et al. (1950) collected 24 cases from the literature, 8 of whom had a history of haemoptysis. Wodehouse (1948) reported a case of multiple bilateral arteriovenous fistulae who had 2 episodes of severe haemoptysis following strenuous effort during military training. Fatal bleeding from the lungs has been reported in 3 cases and Rodes (1954) and Israel et al. (1953) reported 2 such cases. Rupture of an arteriovenous fistula into the pleural cavity was found by Bowers (1936) at autopsy in an infant 2 days old. The rarity of this complication is probably due to the low pressure existing in the pulmonary circulation.

With regard to operation for these lesions, as little lung tissue as possible should be excised. Bosher et al. (1959) found that 212 out of the 350 cases they collected were single lesions. In such single lesions an excision of the abnormal vessels may be possible without removing normal lung tissue. Where the lesions are multiple, extra efforts should be made to conserve lung tissue because of the tendency for new lesions to develop. In our first case almost no lung tissue was removed, whilst in our second a lobectomy was carried out.

Following the operation, patients should be watched to see if new lesions develop. Baer et al. (1950), Parker and Stallworth (1952), and Ronald (1954) found evidence of the appearance of new lesions in the lungs within 6 months after excision of the fistulae.

SUMMARY

1. Two patients with pulmonary vascular abnormalities are described. In one case two peripheral pulmonary arteriovenous communications existed and in the other an anastomotic channel between the bronchial and pulmonary arteries.

2. The morbid anatomy of these two different types of abnormality is discussed.

3. Some observations are made on the surgical removal of these lesions.

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