

## PULMONARY TELANGIECTASIS\*

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Pulmonary telangiectasis is not rare. Since Churton described his necropsy finding in 1897, over 100 cases have been reported in the literature according to a recent review (O'Neill, Fisher, McDowell, and Lauby, 1956). Doubtless many cases remain unrecorded, but nevertheless the condition is not so commonplace as to be familiar to all, therefore it has seemed worth while to present this series of six cases which were first seen between 1942 and 1949 and have been followed up since, the results of treatment being observed in five, and the progress without treatment in the sixth.

Two are personal cases, in three I was privileged to assist at operation, and for the details of the remaining case I am indebted to Sir Clement Price Thomas, and to him for the stimulus to gather these records together.

### CASE REPORTS

CASE 1.—This was an Army reservist aged 42 years. On September 9, 1942, six days after recall to the Forces, he collapsed while doing physical training. He gave a history of having had radium treatment for telangiectasis of the lips and nasal septum because of repeated epistaxis. However, bleeding had continued intermittently.

During the subsequent investigations a radiological opacity was noted in the left lung field. On October 20, 1942, he was transferred to the E.M.S. Chest Centre at King Edward VII Sanatorium, Midhurst. He was dyspnoeic on moderate exertion, and there was slight clubbing of the fingers and marked pallor of the skin and mucous membranes. The lips were scarred, the nasal septum was perforated and the edges of the perforation were unhealthy and prone to bleed.

To the left of the sternum there was a soft systolic murmur, the significance of which was not appreciated at first. The blood pressure was 110/70 mm. Hg, Hb 40%, red cell count 3,200,000 per c.mm., and white cell count 2,800 per c.mm. There was a radiological opacity in the left lung field (Fig. 1).

After blood transfusion a diagnostic pneumothorax was induced, and on October 30, 1942, Mr. Tudor Edwards performed a thoracoscopy. A tumour mass, which appeared to consist of dilated

blood vessels, protruded from the upper lobe surface, and a diagnosis of haemangioma was made. At thoracotomy on December 4, a tumour was present in the lingula area which had an expansile pulsation and a soft thrill could be felt. Two arteries were divided without any change in the tumour so an upper lobectomy was done. Post-operative convalescence was complicated by collapse of the lower lobe, but finally re-expansion was complete and the patient was discharged from hospital on January 13, 1943.

There has been no further evidence of pulmonary lesions in this patient, but the epistaxis has continued intermittently and he is at present (1958) receiving treatment for this condition.

CASE 2.—This was a man aged 22 years in whom an opacity had been noted in the left lung field on a mass miniature radiograph in July, 1944. He was symptomless and no action was taken until January, 1946, when he was admitted to Midhurst for investigation. The reason for this was that a slight degree of cyanosis was now present, although the patient himself had no complaints and played football regularly. A harsh systolic bruit was best heard in the fifth and sixth interspaces in the left nipple line. The

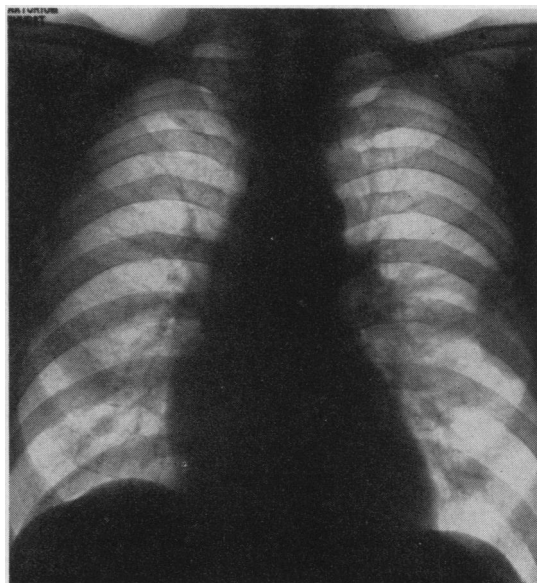


FIG. 1

\*Based on a short paper read before the Society of Thoracic Surgeons in November, 1956.

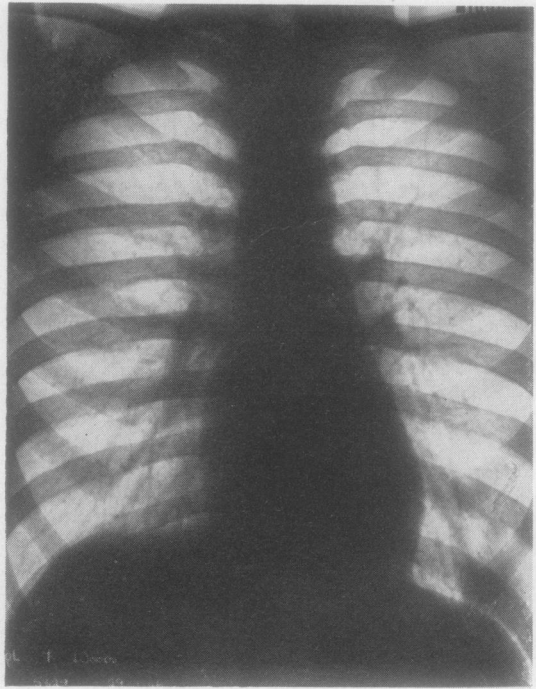


FIG. 2



FIG. 3

blood pressure was 124/70 mm. Hg, pulse 52, red cell count 6,144,000 per c.mm., and the Hb 118%. A radiograph showed a rounded opacity just above the left diaphragm with shadows extending to the hilum suggesting entering and leaving blood vessels (Fig. 2). A tomographic view confirmed this impression (Fig. 3).

Further investigations yielded nothing of importance, and, a diagnosis of arterio-venous fistula having been made, a thoracotomy was performed by Sir Clement Price Thomas on February 25, 1946. On

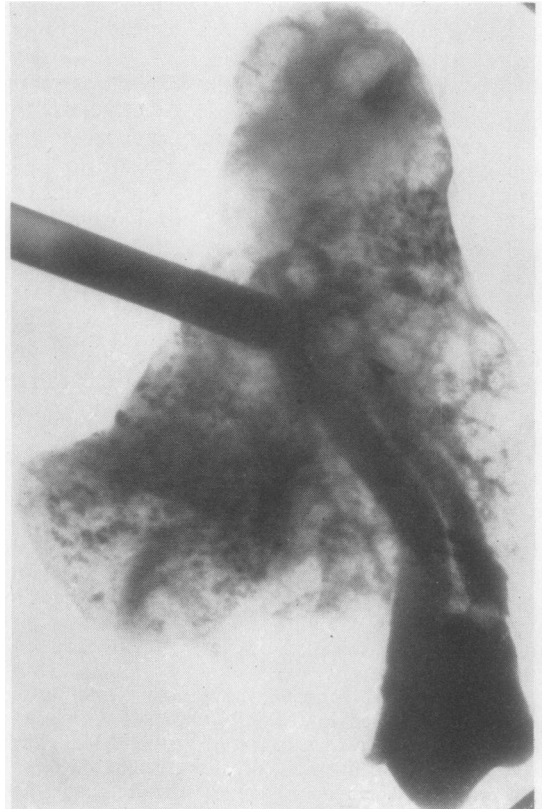


FIG. 4

the anterior surface of the lower lobe there was a vascular mass about  $1\frac{1}{4}$  in. across. The walls were thin and blood could be seen swirling through the tumour. An expansile pulsation was present and a marked systolic thrill palpable. A dissection lobectomy was done; the hilar structures were normal. Convalescence was uneventful. Examination of the removed lobe showed that the artery, immediately after giving off the branch to the apical segment, passed directly into a sac the size of a walnut, and this emptied into the inferior pulmonary vein, which subsequently drained the rest of the lobe (Fig. 4). This patient has remained well since.

CASE 3.—A woman aged 29 years had a haemoptysis in November, 1944. A bronchoscopy and



bronchogram done at that time revealed no abnormality. The haemoptysis continued intermittently and because of this she was admitted to the Brompton Hospital under Sir Clement Price Thomas. A radiograph of the chest showed enlargement of the left hilar shadow (Fig. 5). Bronchoscopy revealed slight bulging of the lateral bronchial wall below the

left upper lobe orifice and deep in the lingula branch there appeared to be a tumour mass.

A left thoracotomy was performed on July 11, 1946. A soft, non-pulsatile red mass about 1 in. in diameter presented into the fissure below the lingula, and could be emptied by pressure between the thumb and finger. Overlying the aortic arch and extending

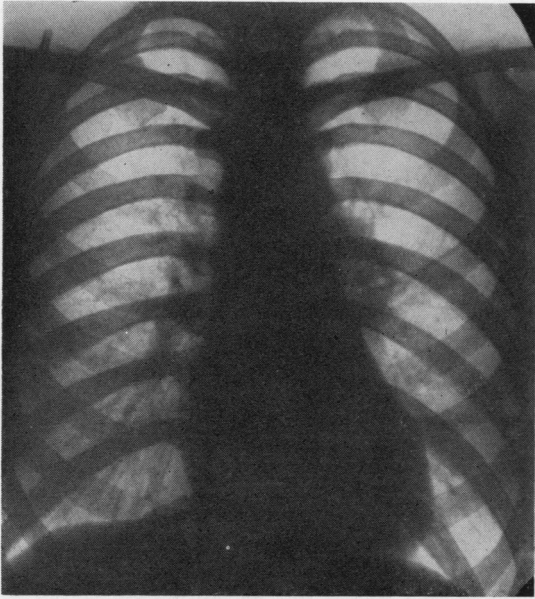


FIG. 5

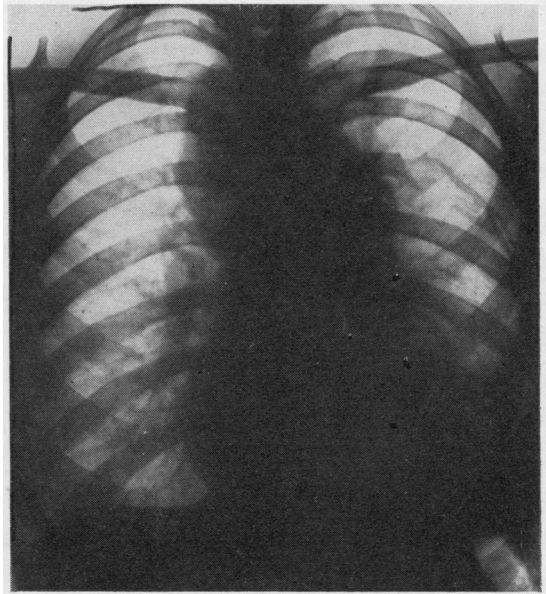


FIG. 7

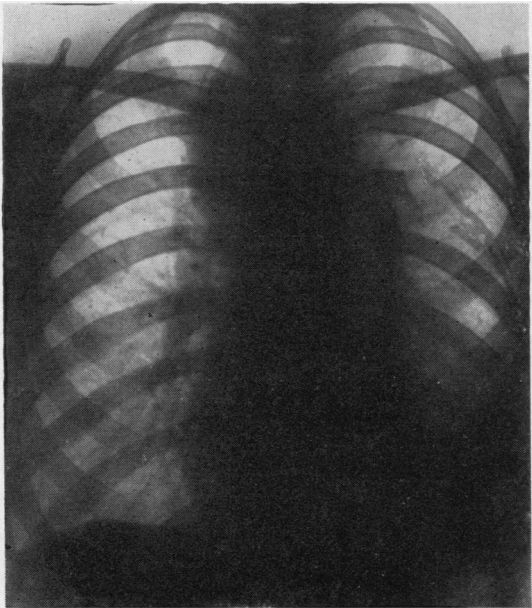


FIG. 6

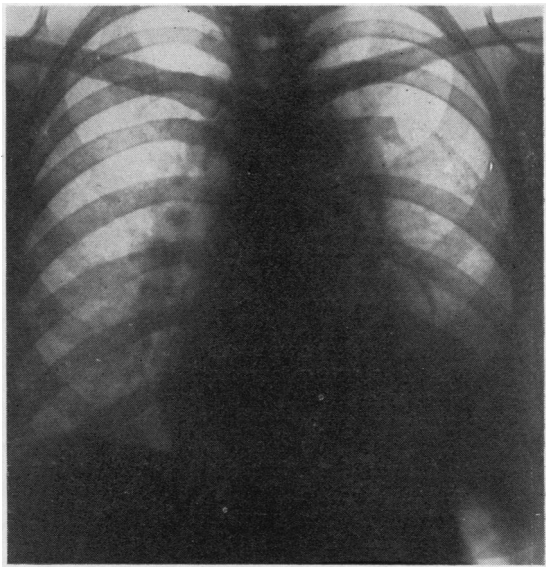


FIG. 8

down to the root of the lung covering the origin of the pulmonary artery and the ductus arteriosus was a trilobed similar mass which could not be emptied by pressure. Anteriorly the phrenic nerve crossed over the mass and the vagus nerve was applied to it posteriorly. After incising the pleura this mass was enucleated without difficulty, but a few small vessels entering the tumour required ligation. The mass in the lingula area was then dissected free until only a small pedicle remained and this was ligated and the mass removed. It had been intimately adherent to the upper and lower lobe bronchi and to the descending part of the pulmonary artery.

After operation the haemoptysis continued much as before, and on July 25 the patient was bronchoscoped again. The bulging of the bronchial wall had gone, but there was some blood clot in the upper lobe bronchus. On August 1, 1946, the chest was reopened and a left upper lobectomy was done. At the same time two large glands containing angiomatous tissue were removed; a few smaller ones passing down the left lower lobe bronchus were left.

After this operation there was no further haemoptysis and progress was satisfactory.

The resected lobe and glands were examined by Dr. J. W. Clegg, who in his report stated that the histological picture was difficult to interpret, but the condition might be malignant.

In 1947 the patient became pregnant and after delivery enlargement of the spleen was noted. In January, 1948, a splenectomy was done at the Westminster Hospital for what was at first thought to be a congenital cystic spleen, but was subsequently considered to be analogous to that found in the chest.

Excepting for a pneumonic illness in October, 1948, she has continued in good health, but in October, 1950, a radiograph of the chest showed a right paratracheal and left basal opacity (Fig. 6). A year later she was given a course of irradiation by Professor Smithers. For some months subsequently little change was discernible in the radiological appearances (Fig. 7). Then the opacities began to shrink and this process has continued (Fig. 8).

This patient has had occasional staining of the sputum over the past two years but otherwise has kept fairly well.

CASE 4.—This was a boy aged 18 years who was admitted to Midhurst on February 27, 1948, following a miniature mass radiography finding of an opacity in the right lung field (Fig. 9). He had no symptoms, and despite investigations the diagnosis was not established when a thoracotomy was done. The pleural space was obliterated by fine adhesions, and while separating these over the middle lobe there was a spurt of blood from the lung surface. Further separation showed that this came from a bluish mass which occupied most of the middle lobe. The lobe was removed, the hilar structures being small but otherwise normal. This was a mixed capillary and cavernous haemangioma, solitary as far as is known. Convalescence was uneventful and the patient remains well to-day.

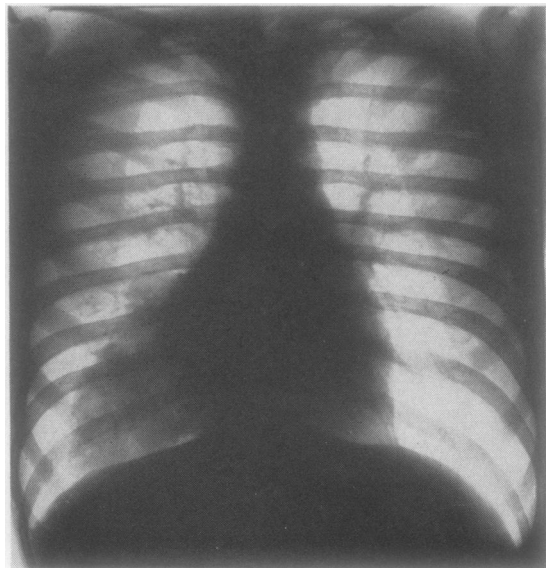


FIG. 9

CASE 5.—This man, aged 26, had had a radiograph in 1944 as a contact of his wife who had been found to have pulmonary tuberculosis. An opacity was noted in the left lung field. He had been kept under observation and the opacity had shown no change, but because from time to time he complained of pain in the chest he was admitted to Midhurst for investigation in July, 1948.

Clinical examination revealed no abnormality. The red cell count was 5,728,000 and Hb was 111%. A radiograph of the chest showed an opacity on the left side. This was considered to be a vascular tumour and tomograms tended to confirm this view, but an angiocardigram was inconclusive.

On September 6, 1948, a thoracotomy was done by Sir Clement Price Thomas. The left upper lobe contained a vascular mass which bulged into the fissure, and a thrill could be felt. A large vessel arising in the mass passed across the fissure and was subsequently traced into the inferior pulmonary vein after passing between the anterior basic bronchus and the remainder of the lower lobe bronchus. The arterial supply to the upper lobe consisted of one large branch to the upper part and one branch to the lingula. These vessels and the superior pulmonary vein were dealt with, but there still remained several small blood vessels near the tumour which had to be dealt with. Their origin was not clear, but they were thought to be bronchial. The lobe was removed. There was some post-operative intrapleural bleeding and the clot became infected, but after drainage the patient made a good recovery and has remained well.

CASE 6.—This girl, aged 23 years, was known to have a radiological opacity at the left base since 1944. Now, five years later, she complained of vague pain in the chest and was admitted to Midhurst for investi-



gation on September 30, 1949. The only abnormality noted on clinical examination was a port wine stain on the back of the neck. Again the radiographic appearances supported by tomography suggested a vascular tumour, but in this case no attempt was made to confirm this by angiocardiology.

A thoracotomy done on September 29, 1949, revealed an artery about 1 cm. in diameter arising from the aorta just above the diaphragm and passing into what appeared to be a small dissociated lung segment and thence on into the lower lobe. In the posterior mediastinum there was a large dilated venous plexus which passed upwards and over the root of the lung to the superior aspect of the pulmonary artery. Proximally the pulmonary artery appeared normal, as also was the superior pulmonary vein. The inferior pulmonary vein was deep to the venous plexus. A thrill was felt in the lower part of the plexus, which was abolished when the vessel arising from the aorta was compressed. Neither the extent of the anomaly nor the pattern could be made out with any certainty, but it seemed that if removal were embarked upon a pneumectomy might well prove necessary. This was considered unjustifiable in the absence of symptoms, and so the chest was closed without any surgical intervention. Since that time the patient has married and had two children and remained symptom-free, and without any apparent change in the lesion.

#### DISCUSSION

In recent years perhaps most interest has centred on the physiological response to an arterio-venous shunt in the pulmonary system. The first detailed investigations appear to be those recorded by Maier, Himmelstein, Riley, and Bunin (1948).

In this series, with the possible exception of Case 2, there were no features which could be attributed to a shunt. In this patient there was slight cyanosis, and perhaps the slightly raised red cell count of 6,144,000 (it was 5,500,000 some months after operation) was significant.

The pathogenesis of these vascular abnormalities, whether found in the lungs or elsewhere, has been the subject of much speculation and discussion, and the terminology applied varied and confusing. It now seems to be generally accepted that the majority of these lesions are developmental abnormalities of the blood vessels, often congenital but apparently sometimes acquired. The latter may possibly follow reparative processes (Willis, 1953) or be initiated by hormonal factors (Hayward and Reid, 1949).

Whatever may be the cause it is a matter of common observation that in pregnancy and in some diseases, particularly those involving the liver, cutaneous naevi may appear and in some cases disappear spontaneously.

A familial form has been recognized since the early writings of Rendu and later Osler (1901). It has for some time been established that the condition may be inherited as a simple autosomal dominant, and a recent investigation on an unprecedented scale by Bird, Hammarsten, Marshall, and Robinson (1957) makes fascinating reading and adds further confirmation. There remains a rare group of malignant angioblastic neoplasms (Willis, 1953).

The literature leaves one in no doubt that the distinction between these two groups, malignant and benign, is not easy, and this has led to confusion between the groups. It has resulted in such anomalous concepts as "benign metastasizing haemangioma" (Robinson and Castleman, 1936) and the "histologically non-malignant angioma with metastases" of Shennan (1914). Shennan postulated that the occurrence of metastases and not the histological features should be the deciding factor in assessing malignancy. But difficulty seems to have arisen in deciding what was and was not a metastasis. Robinson and Castleman record the case of a young girl with histologically benign haemangioma of the breast, with subsequent metastases which were definitely sarcomatous. They suggested that this case was a connecting link between the frankly malignant type and the benign metastasizing, in which the metastases were also histologically benign. Stout (1943), having been given the opportunity of re-examining the material, concluded that the primary tumour was malignant, a haemo-endothelioma. He concluded that there was no such entity as "benign metastasizing haemangioma," and, reviewing reports of 118 cases labelled with some name suggesting malignant tumour, felt compelled to reject 41, either because there was no or an inadequate histological report, or because in his opinion the illustrations and text described a tumour of a different kind.

Willis (1953) accepted a smaller number of the reported cases as authentic. He regarded the apparent metastases as due to canalization and establishment of blood flow in fresh parts of an already existing malformation, and emphasized the difficulties that may arise in distinguishing between true neoplasms and hamartomas of vascular origin, partly because of the histological features which sometimes show a seemingly infiltrative structure and partly because of the simulation of metastases by multicentric formation.

Unfortunately the slides and blocks of Case 3 were lost in the fire at the Brompton Hospital in

1950 and are not available for further study. However, I am indebted to Dr. Clegg for telling me that he came to the conclusion on histological grounds that the case might be malignant although malignancy is rare. He points out that, though the subsequent development of lesions elsewhere to some extent supported this view, it could equally be held that they were multiple developmental abnormalities. It is now almost 14 years since this patient's first haemoptysis. She remains relatively well at the present time. It cannot be doubted that this is an example of multicentric formation, but one which illustrates convincingly the difficulty that may arise in distinguishing between the malignant and the benign.

It seems probable that the effects of arteriovenous shunts are progressive and therefore there is a strong case for surgical intervention whenever possible. Treatment may also be necessary to control haemorrhage or to forestall it.

That all cases require treatment is debatable. Lister (1938), in a study of the natural history of strawberry naevi in infancy, showed that after an initial period of growth lesions sometimes regressed and might disappear completely. Similar observations were made earlier by Wolbach, and later these were elaborated in conjunction with Gross (Gross and Wolbach, 1943). Bailey and Ford (1942) reported a form of sclerosing angioma of the central nervous system

and recently Liebow and Hubbell (1956) report similar findings in pulmonary lesions. In Case 6 in our series there has been no apparent change in the lesion over a period of 14 years or more.

#### SUMMARY

Six patients suffering from pulmonary telangiectasis, all of whom have been observed over a period of years, are recorded.

The pathogenesis and the natural history of these lesions are discussed, the latter with particular reference to treatment.

One case is cited as an example of the difficulty that may arise in distinguishing benign malformations from malignant tumour formations.

My thanks are due to Sir Clement Price Thomas for permission to include his cases, and to Sir Geoffrey Todd for permission to include Case 1 and for access to radiographs and case notes.

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