CAVERNOUS HAEMANGIOMA OF THE LUNG: REPORT OF A CASE

BY

W. P. CLELAND

London

Cavernous haemangiomas of the lung are uncommon. Recently Whitaker (1947) recorded two cases and was able to find only five other instances in the available French, English, and American literature. The pulmonary lesions are usually a manifestation of a widespread and often hereditary haemangiomatosis. The non-pulmonary lesions, apart from haemorrhage when near the surface sometimes resulting in severe anaemia or the occasional formation of a large skin tumour, seem to be of little significance in contrast to the pulmonary lesions, where, owing presumably to less resistance on the part of the surrounding tissues, considerable enlargement can occur with shunting of blood from the right to the left side of the heart. This fistulous communication accounts for many of the additional signs and symptoms encountered in such cases.

The following case presented many of the characteristic features of multiple haemangiomas, pulmonary masses, cyanosis, polycythaemia, clubbing, an exocardial to-and-fro murmur, and a collapsing pulse.

CASE REPORT

The patient, a married woman aged 51 years, had had three normal children. As a child she had always been breathless and blue and was under constant observation for "heart trouble." Her father also had been similarly affected. At school she was able to play some games, but her activities were limited by breathlessness. She married while a young woman and had three children without undue difficulty. Of recent years her symptoms had become worse and in addition she noticed undue fatigue, a winter cough associated with a little sputum, and severe epistaxes recurring at almost weekly intervals.

In 1937 she noticed the gradual appearance of pins and needles in her left leg followed by some loss of power; later she noticed clumsiness and weakness of her right hand and weakness of the muscles on the left side of the face. These phenomena gradually disappeared completely.

Early in 1947 she had an exacerbation of her symptoms with swelling of the ankles in addition. She was admitted to St. James's Hospital under the care of Dr. Barling, with congestive heart failure. A diagnosis of pulmonary haemangioma with cardiac failure was made; the latter responded to rest and routine measures, and later she was transferred to the Thoracic Unit at Horton Emergency Hospital.

On admission the intense cyanosis, combined with multiple small haemangiomas on the face, eyelids, lips, and tongue, gave her a striking appearance (Plate XXIVa). Haemangiomas were also present on the buccal mucosa, on the nasal septum, and at the base of the nails; the remainder of the skin surface showed no lesions. She was slightly dyspnoeic at rest, and her fingers showed gross clubbing of the drumstick type. The heart was not clinically enlarged, nor were there any cardiac murmurs. The pulse rate was between 80 and 90 per minute and the blood pressure 130/70 mm. Hg. There was dullness on percussion, with absent breath sounds at the base of the right lung posteriorly, and over this area there was a loud but fairly localized to-and-fro murmur accompanied by a thrill. The liver edge was palpable just below the costal margin, but there was no oedema of the ankles, nor albumin in the urine.

Radiographs of the chest (Plate XXIVb and c) showed the heart to be slightly enlarged in the transverse diameter. The right descending pulmonary artery and the pulmonary conus were larger than normal. There was a lobulated homogeneous shadow lying in the right cardio-phrenic angle, and in the left lower zone were several small, dense spherical shadows 1 to 2 cm. in diameter. In the lateral films the right-sided shadow was seen lying behind the heart in the lower lobe.

An electrocardiograph revealed a normal rhythm and P-R interval, with inversion of R and T waves in lead III. The circulation times were 5 seconds (arm to lung) and 10 seconds (arm to tongue).

Angiocardiography was kindly performed by Mr. R. C. Brock at Guy's Hospital (Plate XXVa). This showed the dye passing through a dilated right pulmonary artery into the lobulated tumour in the right lower lobe and back to the heart through a

dilated pulmonary vein. It also demonstrated the vascular nature of the left-sided tumours.

On July 25, 1947, a right thoracotomy was performed under anaesthesia by pentothal followed by cyclopropane, later changing to curare and trilene. A pulsating mass was found occupying the lower lobe and a thrill could be felt with ease. The pulmonary artery to the lower lobe was enormously dilated and its walls were very thin, but it was isolated in the fissure and ligated without difficulty. The tumour, however, continued to pulsate until an enormously dilated inferior pulmonary vein was ligated. The lobe was firmly adherent across the fissure to the middle lobe and even more so to the diaphragm where the wall of the tumour was calcified. The operation of lower lobectomy was completed without incident; the patient's condition at the end was good, with a pulse rate of 100, a blood pressure of 110/65 mm. Hg, and, for the first time, a pink complexion. Her general condition immediately after the operation caused no anxiety and she continued to improve slowly. Some twelve hours after its conclusion, however, the pulse began to rise and the blood pressure to fall, and in spite of various measures adopted her condition rapidly deteriorated and she died about twenty hours after the operation.

The right lower lobe (Plate XXVb and c) removed at operation showed a large loculated cavity 9 cm. by 5 cm. occupying the anterior aspect of the lower lobe. The pulmonary artery, 1.5 cm. in diameter, ran down behind the tumour before entering it lower down near the diaphragm; an enormously dilated inferior pulmonary vein 2 cm. in diameter was continuous with the upper pole of the cyst without an

obvious line of demarcation. The cyst wall was intact and lined by a smooth endothelium; its diaphragmatic aspect was calcified for an area of about 2 cm. square.

Autopsy revealed a congested right upper lobe although the bronchial tree was free from blood or secretions. The divided bronchus and vessels to the lower lobe were all satisfactorily secured. The left lung (Plate XXVI) showed several scattered haemangiomas in the lingular segment of the upper lobe and in the lower lobe. The heart showed dilatation of the right side but nothing else abnormal. The liver was fatty and showed surface haemangiomas, and there were submucous nodules in the stomach. The brain showed a large cyst in the left internal capsule but no other evidence of haemangiomas.

An adequate cause of death was not found at autopsy; the clinical condition suggested a severe degree of anoxia produced during the operation, although conditions favouring such an event were at no time experienced; alternatively the weakened heart may have been unable to adapt itself to the new vascular condition produced by closing the large arterio-venous fistula.

SUMMARY

A case of generalized haemangiomatosis with pulmonary manifestations is recorded. A large pulmonary haemangioma was removed by lobectomy; the patient lived for twenty hours after operation.

REFERENCE

Whitaker, W. (1947). *Thorax*, 2, 58.

(a)

PLATE XXIV.—(a) Haemangiomata on face, eyelids, and tongue. (b) and (c) Postero-anterior and right lateral radiographs of chest showing large lobulated opacity in the right lower lobe and the small spherical shadows in the lower zone of the left lung.

(b)

(c)

PLATE XXV.—(a) Angiocardiogram showing opaque dye in the tumour in the right lower lobe with dilated pulmonary artery and vein entering and leaving it. The vascular nature of the left-sided shadows is also indicated. (b) Operative specimen of right lower lobe, showing the grossly loculated cavity occupying the antero-lateral aspect of the lobe (lower right side of specimen). The hilum lies at the top right-hand corner and there is a probe in the pulmonary artery; the tip of the probe can be seen entering the lower left-hand corner of the cavity. The pulmonary vein is continuous with the top right-hand corner of the cavity. (c) Postero-lateral aspect of the right lower lobe to show the dilated pulmonary artery which runs down behind the cavity to enter its lower portion.

(a)

(b)

(c)

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PLATE XXVI.—Autopsy specimen of the left lung to show haemangiomata.