IDIOPATHIC DILATATION OF THE PULMONARY ARTERY

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Idiopathic dilatation of the pulmonary artery is a rare clinical and postmortem finding. Greene *et al.*[1] found 8 cases, confirmed at autopsy, in the literature up to 1949.

CASE REPORT

An African male, aged 65, was admitted to Edendale Hospital, Pietermaritzburg, in August 1958. He complained of chest pain and dyspnoea, a poor appetite, and pain in the lower abdomen. These symptoms had been present for some months.

Examination showed gross wasting. The pulse was completely irregular. The liver was enlarged 4 fingers. The apex beat was palpable 4½ inches from the midline. A systolic thrill was felt and increased pulsation was visible over the pulmonary outflow tract. In the same area a systolic murmur was audible and the first sound split. The blood pressure was 90/80 mm.Hg. There was dullness on percussion over the left apex, with absent air entry.

X-ray of the chest showed gross patchy infiltration of both lung fields, suggestive of cardiac failure. There appeared to be collapse of the upper lobe of the left lung. There was a clearly circumscribed mass at the left hilum with the features of an aneurysm (Fig. 1).

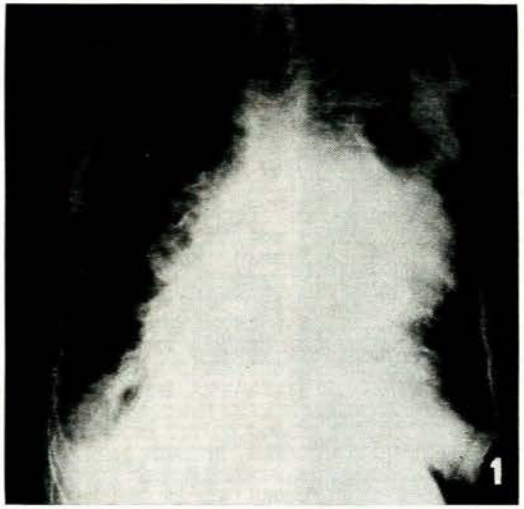

Fig. 1. X-ray of chest (see text).

The blood Wassermann reaction was negative, blood urea 40 mg. per 100 ml., haemoglobin 13·1 G per 100 ml., PCV 44%, MCHC 30%, sedimentation rate 17 mm. in the first hour, and white blood-cell count 14,000 cells per cu.mm.

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The patient died suddenly 2 days after admission.

Autopsy

Postmortem examination confirmed that death was due to pulmonary embolism. Evidence of congestive cardiac failure was present. There was considerable enlargement of the heart, with massive dilatation of the pulmonary artery and its major branches.

The *cardiac enlargement* was due to muscular hypertrophy. The normal 3:1 ratio between the thickness of the left and right ventricular walls was unchanged. The heart valves were all completely normal. The arch of the aorta was of normal size and configuration. The coronary ostia were normally placed and the vessels themselves normal in their branching and distribution. The brachiocephalic and left common carotid arteries had a common stem from the arch of the aorta (Fig. 2).

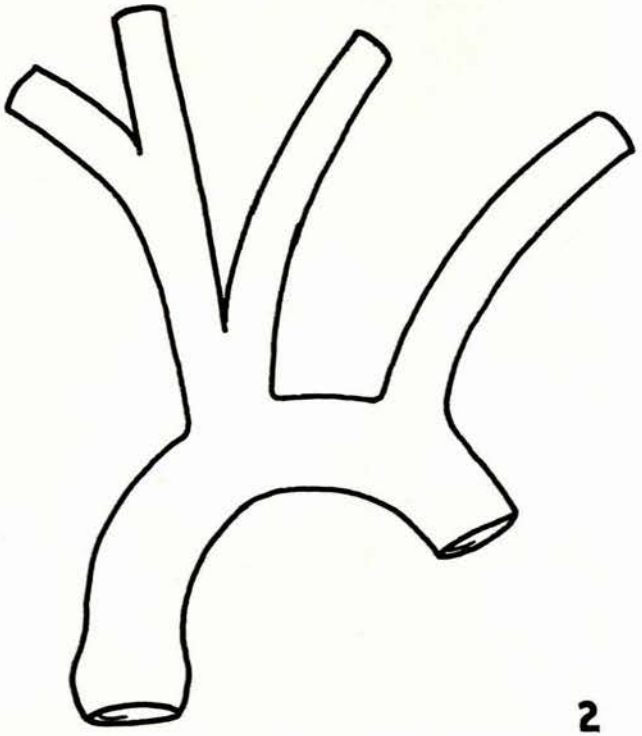

Fig. 2. The aortic arch, showing the common brachiocephalic left common carotid stem.

The *pulmonary artery* was 4·4 cm. in length (normal length

5 cm.) with a diameter of 5·2 cm. giving a cross-sectional area of 21 sq.cm. (normal diameter 3 cm. and area 7 sq.cm.). The left branch of the pulmonary artery, which is normally slightly

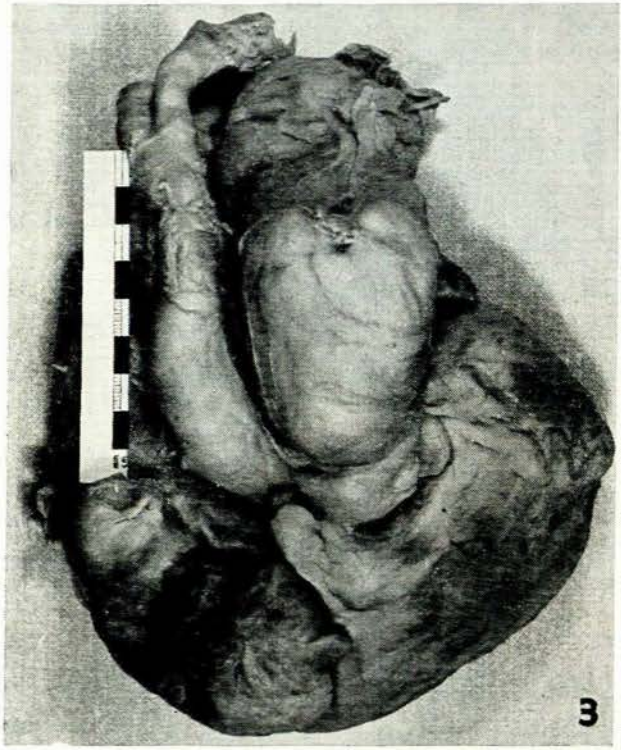

Fig. 3. Photograph of heart *postmortem* showing pulmonary artery (see text).

smaller and shorter than the right, was 5 cm. long (Fig. 3).

Histological examination of the lung, liver and kidney showed no significant abnormalities.

DISCUSSION

Idiopathic dilatation of the pulmonary artery is difficult to diagnose during life. It may present, as in this case, as a sharply demarcated mass in the hilum of the lung, indistinguishable from a neoplasm.[2] In such cases angiocardiography will assist in establishing the true nature of the mass. The condition needs to be differentiated from patent ductus arteriosus, pure pulmonary stenosis (in which a high right-ventricular pressure is demonstrable on cardiac catheterization), atrial and ventricular septal defects, idiopathic pulmonary hypertension, cor pulmonale owing to lung disease, Eisenmenger's syndrome, mitral stenosis, and pulmonary artery aneurysms. If X-ray of the chest shows an enlarged pulmonary artery and a systolic or diastolic murmur, or both, are heard in the second or third left interspace, idiopathic dilatation of the pulmonary artery needs to be considered.[3] Cardiac catheterization and angiocardiography are of considerable value in confirming the diagnosis.

The aetiology of the condition is uncertain. Goetz and Nellen[3] report 4 cases found in 70 consecutive cases of congenital heart disease. Taussig[4] records a congenital case where the dilatation was so extreme that cyanosis and severe dyspnoea resulted from the constriction of the trachea with the slightest variation in posture.

The condition should not be diagnosed when intra- or extracardiac shunts are present, in the presence of chronic pulmonary disease, or when the dilatation is more circumscribed or actually aneurysmal. With circumscribed dilatation about 23% of the cases will be the result of syphilis, mycotic degeneration, or atheroma, with disease of the arterial wall demonstrable at necropsy.[5, 6]

SUMMARY

1. A case of idiopathic dilatation of the pulmonary artery is reported. Autopsy was performed.

2. The clinical presentation, diagnosis and aetiology are briefly discussed.

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