CASE REPORTS

CONGENITAL ABSENCE OF LEFT PULMONARY ARTERY WITH COARCTATION OF RIGHT PULMONARY ARTERY

BY

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Congenital unilateral absence of a pulmonary artery is a rare condition. Pool, Vogel, and Blount (1962), in a review of the English published material, collected 98 cases and described 4 of their own. Of this group 23 were proven examples of absence of the left pulmonary artery. Oakley, Glick, and McCredie (1963) described a further case, and Sherrick, Kincaid, and DuShane (1962) encountered 8 examples. This condition is uncommon as an isolated anomaly, and is most frequently seen in conjunction with the tetralogy of Fallot, but may also be associated with atrial and ventricular septal defects, and less commonly with patent ductus arteriosus. Apparently unilateral absence of the left pulmonary artery has not been reported in association with a coarctation of the right pulmonary artery.

Case History

An African man, aged 41 years, was admitted to hospital for investigation of mild exertional dyspnoea of two years' duration. He had never experienced hæmoptysis or chest pain, and there was no history of rheumatic fever. Clinical examination revealed a well-built man in no distress with no evidence of cardiac failure. The blood pressure was 110/70 mm. Hg. All the pulses were present and equal. The heart size was difficult to determine clinically; an ejection systolic murmur was audible at the pulmonary area and radiated poorly to the neck and mitral area, but was well heard in the right chest and was maximal in the right axilla. This murmur became louder after the inhalation of amyl nitrite. The second sound was normally split with slight accentuation of the pulmonary component. In addition a localized blowing early diastolic murmur which waxed on inspiration was heard at the pulmonary area, and was thought to be the murmur of pulmonary incompetence.

The left chest was smaller and moved less well than the right, and the air entry was diminished on the left side. Further physical examination was negative. A full blood count was normal and the Wasserman reaction was negative.

An electrocardiogram (Fig. 1) showed a mean frontal plane vector of plus 100° with a tall R wave in lead V1 suggestive of right ventricular hypertrophy. A chest radiograph (Fig. 2) showed conspicuous dilatation of the main pulmonary artery and an absent hilar comma on the left. The left hemithorax was much smaller than the right, and the vascular markings in the left lung were absent. The streaky shadowing present in the left upper lobe was considered to be crowded bronchial markings. Bronchography (Fig. 3) revealed an essentially normal bronchial tree except for some crowding of the bronchi in the left upper lobe.

Cardiac catheterization revealed a right atrial pressure of 5/0 mm. Hg, right ventricular pressure of 65/0 mm. Hg, and main pulmonary artery pressure of 65/12 mm. Hg. The pressure in the right pulmonary artery was 30/12 mm. Hg with a gradient of 35 mm. Hg on withdrawing the catheter across the right pulmonary artery. The pressure tracing in the main pulmonary artery was almost identical with the tracing in the right ventricle and was suggestive of pulmonary incompetence (Fig. 4). The catheter could not be made to enter the left pulmonary artery. The oxygen saturations on the right side of the heart were normal and averaged 75 per cent. The systemic arterial saturation was 98 per cent. The cardiac index was 3.1 l./m.²/min.

FIG. 1.—Electrocardiogram showing right axis deviation and right ventricular dominance.

Cine-angiography from the right ventricle and main pulmonary and right pulmonary arteries showed a very dilated main pulmonary artery and a coarctation of the right pulmonary artery close to its site of origin from the main pulmonary artery. The left pulmonary artery and its branches failed to fill, but in the aortic phase some dye was visualized in small vessels of the left lung. These vessels were presumably filled from bronchial collaterals. There was a consistent reflux of dye from the main pulmonary artery into the right ventricle due to pulmonary incompetence.

Single plane angiograms (Fig. 5) confirmed the above findings.

FIG. 2.—Postero-anterior 6-foot chest radiograph (see text).

FIG. 3.—Normal bronchogram showing some crowding of the bronchi of the left upper lobe.

FIG. 4.—Withdrawal pressure tracing from right pulmonary artery (RPA) to right ventricle (RV). (MPA=main pulmonary artery).

FIG. 5.—Pulmonary arteriogram (antero-posterior projection). The left pulmonary artery is absent, and there is coarctation of the right pulmonary artery. The main pulmonary artery and the right main pulmonary artery distal to the coarctation are enlarged.

Discussion

Congenital coarctation of the pulmonary artery is not uncommon, and may be unilateral, bilateral, single, or multiple. It is reported as an isolated lesion in 40 per cent of cases, the remaining 60 per cent being associated with other congenital cardiac anomalies, most frequently valvular pulmonary stenosis (Gay *et al.*, 1963). The condition may give rise to severe pulmonary arterial hypertension. Coarctation of a pulmonary artery has not been described in association with congenital absence of the other pulmonary artery. In the absence of confirmation at necropsy, it is remotely possible that the failure to demonstrate blood supply to the left lung angiographically may be due to a superimposed thrombus on the pre-existing coarctation of the origin of the left pulmonary artery. However, the absence of any previous episode of chest pain or hæmoptysis and the disparity in size between the two lung fields make this possibility unlikely.

Pool *et al.* found pulmonary hypertension in 19 per cent of cases of isolated unilateral absence

of a pulmonary artery and in 88 per cent of those patients with a shunt in addition. They postulated that increased pulmonary blood flow from the time of birth was an important factor in the development of pulmonary hypertension. This case demonstrates that coarctation of the opposite pulmonary artery in the absence of a shunt may be another cause of pulmonary hypertension in cases of unilateral absence of the pulmonary artery.

Summary

A case is described of congenital unilateral absence of the left pulmonary artery with pulmonary hypertension due to an associated congenital coarctation of the right pulmonary artery in a man aged 41 years.

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