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MEDICAL MEMORANDUM

Congenital Aneurysm of the Right Pulmonary Artery Associated with Persistent Ductus Arteriosus, Ventricular Septal Defect and a Right-sided Hypoplastic Aorta

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Summary. A case of congenital right pulmonary artery aneurysm associated with persistent ductus arteriosus, right-sided hypoplastic aorta and ventricular septal defect in a 7-year-old child is reported. The most conspicuous symptom was cough associated with a single episode of haemoptysis.

Résumé. Un cas d'aneurysme de l'artère pulmonaire droite, associé avec ductus arteriosus persistant, aorte hypoplastique droite et inocclusion du septum, en enfant de sept ans, est rapporté. Le symptôme le plus marqué était une toux associé avec un épisode seul d'hémoptysie.

Congenital aneurysm of the pulmonary artery is one of the rare cardiovascular malformations. Johannsen & Connor (1943) reported only one case of this anomaly out of 28,180 necropsies carried out at the Bellevue Hospital; Deterling & Clagett (1947) at the Mayo Clinic also reported only one case out of 17,545 necropsies, and in their review of the world literature these authors found seven other documented cases. The purpose of this communication is to report a case of this rare malformation, the diagnosis of which was made during life in a 7-year-old Nigerian child. Although this case was the only one encountered among 300 necropsy and clinical cases of congenital heart defect studied at the University College Hospital (UCH) between 1964 and December 1970, Udekwu (1965) reported one other case diagnosed as necropsy from the same institution.

CASE REPORT

O.A. (UCH No. 117599) was a 7-year-old Yoruba female first seen in 1964 at the age of 2½ months. The significant history then included persistent cough and frequent vomiting. She

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had tachypnoea and tachycardia. There was cardiomegaly, a harsh pansystolic murmur maximal in the fourth left intercostal space. A chest radiograph showed cardiomegaly plethoric lungfields and a prominent vascular shadow in the right hilar region (Fig. 1). The clinical impression was congenital heart defect with a left-to-right shunt. On account of the tachycardia, cardiomegaly and hepatomegaly, a diagnosis of cardiac failure was made and she was therefore digitalized.

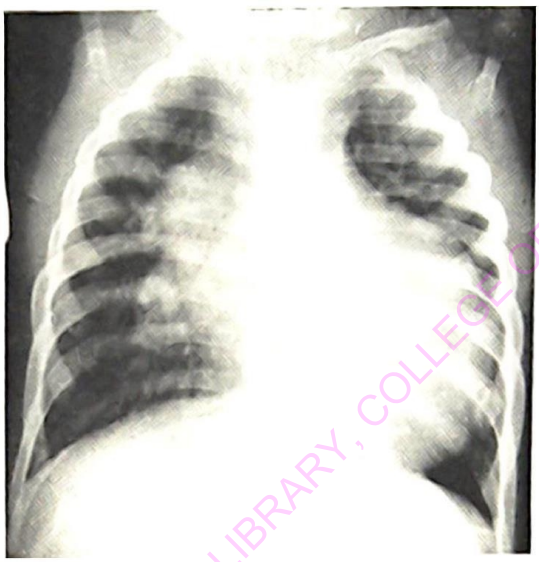

FIG. 1. Chest radiograph of the patient aged 2½ months. Note the conspicuous vascular shadow in the right hilum, lung plethora and cardiomegaly.

At subsequent out-patient follow-up, the essential clinical findings included further increase in the heart size, a loud first heart sound in the mitral area, accentuated second heart sound in the pulmonary area, mitral diastolic flow murmur, and a rough systolic murmur at the left sternal border. The findings in the chest radiograph were essentially the same as before except for a larger right hilar shadow than noted previously. The electrocardiographic (ECG) findings included sinus rhythm, heart rate 105/min, left axis deviation, biventricular hypertrophy and left atrial dilatation. The haemoglobin was 14·2 g (97%), PCV 45%, MCHC 32% and haemoglobin electrophoresis AA. The impression was still that of a left-to-right shunt, probably a ventricular septal defect (VSD). However, the heart shape, right hilar vascular shadow and ECG changes were atypical for an uncomplicated VSD. Central cyanosis was observed for the first time at the age of 2 years.

In January 1969, a right-sided angiocardiography was performed and the study revealed displacement of the superior vena cava and the left innominate vein by a round mass (Fig. 2a) which corresponded with the right hilar shadow in the plain chest radiograph. An aneurysm of the right pulmonary artery (Fig. 2b) was revealed in a later opacification of the vascular mass. The oarta was also found to be hypoplastic and right-sided.

A subsequent left-sided angiocardiographic study showed a persistent ductus arteriosus (Fig. 3), and further delineated the right pulmonary artery aneurysm.

Recently, the child has developed clubbing of the fingers and toes, and the cyanosis noted

previously has increased. On one occasion she had slight haemophysis. There is moderate reduction in her exercise tolerance.

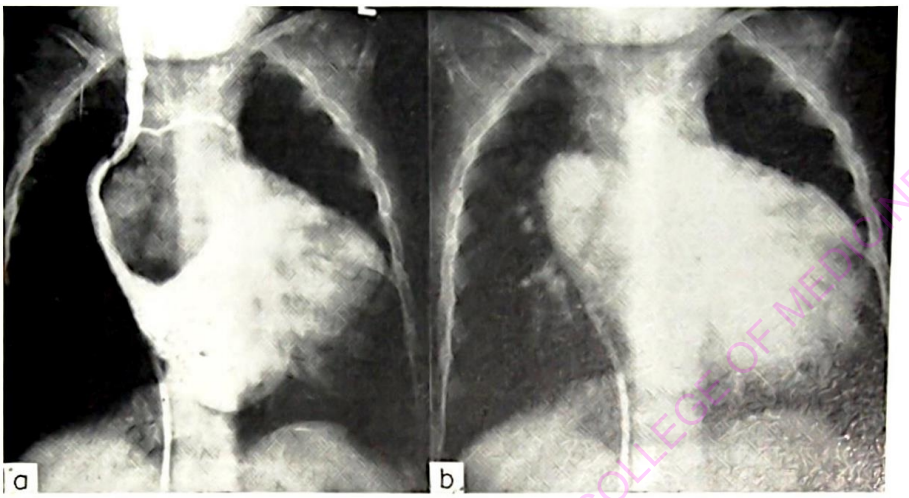

FIG. 2. Right-sided angiocardiogram. (a) The catheter is in the right atrium. Note the reflux of contrast material into the displaced superior vena cava and elevated left innominate vein by the vascular right hilar shadow. (b) At 2·0 sec from the injection of contrast material, the right pulmonary artery aneurysm is opacified. A small aortic knuckle in the midline is also opacified.

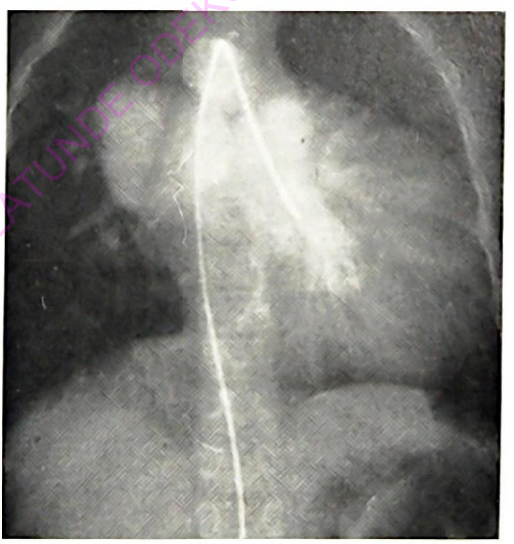

FIG. 3. Left-sided angiocardiogram showing the right pulmonary artery aneurysm, the small aortic knuckle and persistent ductus arteriosus. The tip of the catheter is in the left pulmonary artery, having passed through the persistent ductus.

COMMENT

Congenital aneurysm of the pulmonary artery is a rare malformation. In the past the diagnosis of this condition was usually made at necropsy. With modern cardiac diagnostic tools the diagnosis can now be easily made during life (Robb & Steinberg, 1939; Boyd & McGavack, 1939; Calenoff, 1964).

The malformation is more often than not associated with other congenital cardiovascular defects (Boyd & McGavack, 1939; Deterling & Clagett, 1947; D'Arbela *et al.*, 1970). In those cases reported by Boyd & McGavack and Deterling & Clagett, the incidence of associated cardiac malformations was 66 and 47% respectively. The most common associated defect is reported to be persistent ductus arteriosus. The incidence of this association in the series quoted above was 23 and 21% respectively. Persistent ductus arteriosus was an associated defect in both the present case and that recently reported by D'Arbela *et al.* (1970). The next common associated anomaly, VSD, reported by Deterling & Clagett, was present in our case. Associated hypoplasia of the aorta in the present case has also been reported by others (Joules, 1934; Wilkinson, 1940; Deterling & Clagett, 1947). An associated right-sided aorta has not, to our knowledge, been reported before. Another rare associated defect recently reported is pulmonary infundibular stenosis (D'Arbela *et al.*, 1970).

In spite of the commonly associated persistent ductus arteriosus with its characteristic continuous and easily detectable murmur, bedside diagnosis of congenital aneurysm of the pulmonary artery is by no means easy. In the present case as well as in the one reported by D'Arbela *et al.*, the typical murmur associated with persistent ductus arteriosus was not present. Experience gained from the present case shows that a plain chest radiograph together with evidence of cardiomegaly and the presence of a heart murmur is sufficient for a bedside diagnosis to be readily made. The important feature in the radiographic appearance is persistence of a vascular shadow in the hilar region. Fluoroscopy would reveal pulsation of the shadow, although in older patients pulsation may be obscured by calcification in the aneurysm.

Cough with haemoptysis has been reported to be the most common symptom (Boyd & McGavack, 1939). This was also the most prominent symptom in the present case. In addition, our patient has developed cyanosis and clubbing of the fingers which presumably result from a reversal of the left-to-right shunt.

There can be no doubt that the present case is of congenital origin. The clinical features were detected when the patient was 2½ months old. There was no evidence of other causes of pulmonary artery aneurysms, namely, Marfan's syndrome, long-standing pulmonary hypertension secondary to a large ventricular septal defect, or a complication of ligation of persistent ductus arteriosus.

ACKNOWLEDGMENT

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