CASE REPORTS

DISSECTING ANEURYSM OF THE AORTA ASSOCIATED WITH CONGENITAL PULMONARY VALVULAR STENOSIS

BY

BYRON EVANS AND T. M. DAUNCEY

From the United Cardiff Hospitals

The association of congenital cardiovascular defects with dissecting aneurysm of the aorta is well recognized, particularly in patients under the age of 40 years. The following case illustrates the rare association between dissecting aneurysm of the aorta and congenital pulmonary valvular stenosis.

Case Report

A man, aged 58 years, who was in hospital because of retention of urine, admitted only to attacks of bronchitis and mild breathlessness on exertion for some years. Examination, however, revealed collapse of the left lung (Fig. 1) and a striking systolic thrill and murmur, maximal in the pulmonary area. The

FIG. 1.—Radiograph of the chest, showing striking collapse and fibrosis of the left lung.

blood pressure was 165/85 mm. Hg. He continued to have attacks of bronchitis and occasional hæmoptysis but was able to return to work.

He was re-admitted five years later following the recent onset of a tearing pain between the shoulder blades and beneath the sternum, together with severe breathlessness. The clinical signs were unaltered from those observed five years before, except that the blood pressure was now 220/110 mm. Hg. The peripheral pulses were normal and no abnormal neurological signs, nor external congenital defects, were found. Chest X-ray confirmed collapse of the left lung and showed widening of the ascending aorta. Electrocardiograms showed right ventricular preponderance and no evidence of myocardial infarction. The serum Wassermann and Kahn reactions were negative. Four days after admission, he had further severe pain, followed by a subcutaneous effusion of blood at the root of the neck and front of the chest. After some weeks, signs of obstruction of the left jugular and axillary veins were seen, with paralysis of the left vocal cord and the left dome of the diaphragm. Heart failure with auricular fibrillation followed, and death occurred suddenly, eight weeks after admission.

Necropsy (Dr. B. H. Knight). The immediate cause of death was a massive hæmorrhage into the left pleural cavity. This came from a tear in the wall of a false aneurysmal sac, 8 cm. in diameter; it was enclosed within the upper lobe of the left lung, causing compression and fibrosis of the lung tissue. The cavity of the sac contained organized thrombus, and was part of a dissecting aneurysm from the arch of the aorta to the femoral arteries. The false lumen was completely endothelialized and communicated with the true lumen of the aorta at one site only by a smooth slit 2 cm. long just distal to the origin of the left subclavian artery. The superior mediastinum was full of organizing blood clot. Histological examination of the involved aorta demonstrated cystic medial necrosis.

There was also a pulmonary valvular stenosis of moderate degree (diameter of 1.25 cm.) with post-stenotic dilatation of the pulmonary arteries (Fig. 2). Both ventricles were hypertrophied and coronary atherosclerosis was present.

Discussion

The commonest congenital cardiovascular abnormality associated with dissecting aneurysm is coarctation of the aorta; thus, Abbott (1928) found that 38 of 200 cases of coarctation died of aortic rupture, and some degree of coarctation was discovered in 131 of 300 cases of aortic dissection reviewed by Shennan (1934) and in 45 of 141 reviewed by Schnitker and Bayer (1944). Other associated congenital defects include bicuspid aortic valves, aortic and sub-aortic stenosis, septal defects, and persistent ductus arteriosus. In many cases these are part of more widespread congenital defects thought to affect tissues of mesoblastic origin, such as arachnodactyly, in which dissecting aneurysm is a recognized cause of death. No reference was found to pulmonary stenosis associated with dissecting aneurysm, although Whitfield *et al.* (1951) mention pulmonary stenosis in arachnodactyly, and Abbott (1928) noted pulmonary atresia and stenosis in cases with coarctation of the aorta.

The association of congenital abnormalities and aortic dissection is of interest since a characteristic defect in the aortic media—cystic medial necrosis of Erdheim—has been described in most cases. This defect in dissecting aneurysms of younger age groups appears to be histologically identical with those occurring in pregnancy and in older hypertensive subjects. It is considered to be one of the most important factors in pathogenesis, although it is uncertain if it is a congenital defect of connective and elastic tissue of the media in the younger groups or a degeneration in the older. It is interesting to note that similar changes have been induced experimentally by feeding animals with tyramine (Duff, 1939) and certain seeds (Ponseti and Baird, 1952).

This case also demonstrates the remarkable occurrences of a silent, or asymptomatic, initial dissection and a prolonged course of at least five years. Although several cases have been reported, silent dissection is uncommon, but it may be that the pain is masked in some instances by syncope which is occasionally the presenting symptom. It is generally agreed that in about 10 per cent of all

cases, the dissecting aneurysm of the aorta becomes healed, and often the patient survives for up to eight years.

FIG. 2.—Photograph showing the dilated right pulmonary artery (R.P.A.) beneath the aortic arch (A.A.). Blood clot obscures the entrance (E) that leads from the lumen of the dissection (D) to the false sac in the left lung.

Summary

The clinical details and necropsy appearances are described of a case of silent dissection of the aorta with prolonged survival. The patient had also congenital pulmonary valvular stenosis.

References

- Abbott, M. (1928). *Amer. Heart J.*, 3, 392 and 574.
 Duff, G. L. (1939). *Proc. Soc. exp. Biol. N.Y.*, 41, 295.
 Ponseti, I. V., and Baird, W. A. (1952). *Amer. J. Path.*, 28, 1059.
 Schnitker, M. A., and Bayer, C. A. (1944). *Ann. intern. Med.*, 20, 486.
 Shennan, T. (1934). *Spec. Rep. Ser. Med. Res. Coun., Lond.*, No. 193.
 Whitfield, A. G. W., Arnott, W. M., and Stafford, J. L. (1951). *Lancet*, 1, 1387.