unaffected when subsequently thyrotoxicosis reappeared are all strong evidence against the etiologic significance of the latter.

By excluding therefore other potential causes, the

possible relationship of this syndrome to the potassium perchlorate, and particularly by a mechanism of toxic reaction, becomes more likely since allergic manifestations were not observed.

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EISENMENGER SYNDROME WITH RADIOLOGICAL CALCIFICATION OF MAIN PULMONARY ARTERIES

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CALCIFICATION of the main pulmonary artery or of its left and right branches gross enough to be visualized radiologically and diagnosed during life has not been described, as far as we could find from the literature. Atherosclerosis of the pulmonary arteries is of course common in long-standing pulmonary hypertension, and these atheromatous patches would naturally get calcified, although not large enough to be seen on X-ray examination. Thrombosis-in-situ of the large pulmonary arteries has been described by Brenner (1931), Dimond and Jones (1954), Magidson and Jacobson (1955), Ring and Bakke (1955) and by others. Magidson and Jacobson (1955), describing a series of these cases with autopsy findings did not mention any gross calcification of the pulmonary arteries. Closed chest injury has been mentioned as a cause of pulmonary artery thrombosis (Dimond and Jones, 1954; Don Michael, 1962, etc.)

Aortic valve cusp damage following non-penetrating injury of the chest and causing aortic incompetence has been described (Wood, 1956).

The case we describe below is of a young female who had pulmonary incompetence, with the clinical picture of Eisenmenger's syndrome and radiological calcification of the main pulmonary arteries, following a history of trauma to the chest.

Case Report

A 27-year-old unmarried female patient was admitted to the General Hospital, Colombo, in October 1962 with a history of dyspnoea on ordinary exertion, such as walking on the flat, since the age of ten years.

At the age of eight years the patient had fallen while playing and had struck the front of her chest on the edge of a stone step, with another child's weight falling on her back. For one month following this she had a pain in the precordial region. Two years after the chest injury she woke up one morning with a sense of dyspnoea and pain in the chest, and these symptoms increased as she was made to walk to school. She had a syncopal attack on the way to school and was cyanosed during the attack.

There had been no syncopal attacks since then, but anginal pain on walking had been experienced off and

FIG. 1.—Aneurysmal dilatation of pulmonary conus and right and left pulmonary arteries, showing horizontal streaks of calcification in right hilum and upper part of pulmonary conus.

FIG. 2.—Left anterior oblique (15 degrees) view of chest showing massive dilatation of left pulmonary artery, with calcification. Screening confirmed the abrupt constriction of the pulmonary artery distally.

on. Dyspnoea on ordinary effort was persistent from the age of ten years, and she was noticed to turn blue after exertion. She had also suffered from frequent attacks of 'bronchitis' about five to six times each year, since the time when she started getting the effort dyspnoea.

When she was warded in 1957, ECG examination had shown right ventricular preponderance, right axis deviation with a vertical heart, and pulmonary P-waves. Screening then had shown prominent pulmonary artery and main branches with exaggerated pulsation, and some right ventricular enlargement. She was then diagnosed as an A.S.D. with pulmonary hypertension and reversed shunt, after cardiac catheterization. In 1959 the patient had a bout of cough and hæmoptysis lasting for two days.

Two months before the present admission to hospital, her effort intolerance had increased, dyspnoea being brought on by less than ordinary exertion. During the same time she had frequent attacks of palpitation and pain over the precordium, even at rest. She also complained of dysphagia for solids.

The patient was of slender build and poorly nourished. The skin was xerodermic, especially in the legs. There were no deformities of the chest wall. The palate had a high-arched appearance. There was central cyanosis, and clubbing of the finger nails. There was no dependent oedema.

The jugular venous pressure was not raised; but a hepatogastric reflux was present. Pulse rate was 88/min. regular, and of moderate volume. B.P. 120/80 mm. Hg. The apex beat was in the fifth left intercostal space, $4\frac{1}{2}$ in. from the mid-line: it was a fairly vigorous 'right ventricular' tap. A left para-

sternal thrust was present, and pulmonary artery pulsations were felt in the second and third left intercostal spaces. A diastolic thrill was palpable, maximally in the third left intercostal space. There was percussion dullness in the second left intercostal space. The first heart sound was normal. There was a pulmonary ejection click and a short soft systolic murmur, best heard in the third left intercostal space. The second sound was normal in the aortic area, and very soft and single in the pulmonary area. There was a long, moderately loud, rough early diastolic murmur continuing into mid-diastole maximal in the third left intercostal space, but heard widely over the precordium.

The lungs were clinically normal. The liver was just palpable, the spleen was not palpable.

Investigations. Blood: Hb, 13.2 g./100 ml.; RBC 6 million/cu. mm.; PCV, 56%. The blood picture was normal. WBC, 11,200/cu. mm; polymorphs, 56%; lymphocytes, 36%; eosinophils, 8%. Serum calcium 10 mg./100 ml.; alkaline phosphatase, 7 K.A. units. Blood urea, 40 mg./100 ml.; blood cholesterol, 240 mg./100 ml. Blood for Kahn and V.D.R.L. tests negative.

ECG: There was marked right axis deviation, vertical heart, and clock-wise rotation. P-wave was 1 mm. in height in Lead 2 and in VI, tall R waves with inverted T waves in Lead 3, Lead 2, a VF, and in VI, denoting right ventricular strain. X-ray of chest showed gross enlargement of pulmonary arteries (right and left) and of pulmonary conus (Fig. 1) and calcification in left and right pulmonary arteries (Figs. 1 and 2).

Screening showed vigorous hilar pulsations and

FIG. 3.—Lateral view taken at screening with barium-swallow, to show ring-like appearance of calcification in cross-sectioned view of left pulmonary artery which is seen to displace the oesophagus backwards.

sudden narrowing in calibre of vessels (after the main right and left pulmonary arteries). There was displacement of oesophagus by enlargement of left pulmonary artery. Right ventricle was enlarged. Aorta was not easily identified, and appeared to be small. The radiologist gave a probable diagnosis of A.S.D., and aneurysmal dilatation of pulmonary arteries with calcification.

Discussion

The interesting features in this case were the clear history of crush injury to the chest in childhood, followed by chest pain, dyspnoea and the later development of the clinical picture of the Eisenmenger syndrome and radiological evidence of gross calcification of the main pulmonary arteries (Figs. 1, 2 and 3).

Wood (1958) defines the Eisenmenger syndrome as pulmonary hypertension with pulmonary vascular

resistance around systemic levels, and with a reversed shunt at any level centrally. The cases with the shunt at atrial level tend to develop reversal of shunt in adult life in 92% of cases, whereas those with a reversed shunt through a V.S.D. or a patent ductus had the syndrome manifesting in infancy in about 80% of cases (Wood, 1958). This late development of the syndrome in cases with the shunt at atrial level is considered to be due to pulmonary hypertension in atrial septal defects being a later acquired characteristic. The two most likely causes for sudden development of dyspnoea and cyanosis in later life with sudden rise of pulmonary vascular resistance are chronic bronchitis with emphysema, and multiple pulmonary embolism (Wood, 1956). Our patient had dyspnoea from ten years of age with cyanosis noticed on exertion, and was first seen and diagnosed as a case of the Eisenmenger syndrome with the shunt at atrial level, when she was 22 years old.

The factor that brought about acquired pulmonary hypertension in our case may well be repeated thrombo-embolism from thrombus formed in the main pulmonary arteries (right and left) following trauma. Mural thrombus following indirect trauma on a large pulmonary artery would get organized and later calcification could occur, as is shown in the ring-like appearance of the calcification in Fig. 3.

In cases of the Eisenmenger syndrome with A.S.D. a wide and fixed split of the second sound in the pulmonary area was found in 86% of cases in Wood's series and in no case was the second sound single in the pulmonary area (Wood, 1958). The single soft second sound in the pulmonary area in our case may either be due to the gross pulmonary incompetence, shown by the diastolic thrill and the long loud pulmonary diastolic murmur, or may be explained by the shunt occurring through a patent foramen ovale rather than an A.S.D. (This might also explain the absence of right bundle branch block in the E.C.G.) However, massive dilatation of pulmonary arteries is more a feature of A.S.D. than of pulmonary hypertension as such (Wood, 1958).

Summary

A case of Eisenmenger's syndrome with a history of crush injury to the chest wall in childhood followed by probable thrombosis in the main pulmonary arteries, giving rise to a thrombo-embolic type of pulmonary hypertension, and later calcification of the left and right main pulmonary arteries, is described and discussed.

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