Acquired pulmonary stenosis

W. A. LITTLER, J. B. MEADE, and D. I. HAMILTON

The Regional Cardio-thoracic Surgical Centre, Broadgreen Hospital, Liverpool, 14

Four cases of pulmonary artery stenosis resulting from extrinsic pressure are presented. All of these cases presented with the triad of chest pain, dyspnoea, and a pulmonary systolic murmur. Respiratory variation of this murmur was noted in three of the patients, the murmur increasing during expiration and diminishing or disappearing during inspiration. It is suggested that this may be a useful sign in diagnosing this syndrome. The tumour in these four cases was an intra-pericardial sarcoma, a benign teratoma, Hodgkin's disease, and a malignant thymoma respectively.

Acquired pulmonary stenosis is uncommon, particularly so when it is due to extrinsic compression by a mediastinal tumour. Prior to the series of cases reported by Gough, Gold, and Gibson (1967) and Seymour, Emanuel, and Pattinson (1968) only about a dozen single case reports had appeared in the literature. The tumours responsible for this syndrome included benign and malignant teratomas (Rusby, 1944; Maier, 1948; Fry, Klein, and Barton, 1955), lymphatic cyst (Wood, 1956), Hodgkin's disease (Winter, 1958; Shaver, Bailey, and Marrangoni, 1965), pericardial mesothelioma (Waldhausen, Lombardo, and Morrow, 1959), and lymphoblastoma (Babcock, Judge, and Bookstein, 1962).

Only 2 of the 7 patients reported by Gough *et al.* (1967) and 7 of the 13 patients of Seymour *et al.* (1968) had, in fact, a primary mediastinal tumour. Extrinsic compression in the remaining cases was due to aortic aneurysm, bronchial carcinoma, constrictive pericarditis, and pulmonary tuberculosis. We therefore feel justified in adding a further four cases to the literature.

CASE REPORTS

CASE 1 A 35-year-old man was referred to this centre with a short history of severe central chest pain and effort dyspnoea. Twelve months earlier he had been admitted to another hospital with a left-sided pulmonary embolus, for which no specific cause was found. He made a speedy recovery and on discharge from hospital his chest radiograph had been reported as clear.

Physical examination revealed a healthy man with a normal pulse and jugular venous pressure; the blood pressure was 90/70 mm. Hg. A grade 3/3 mid-systolic murmur was intermittently heard in the pul-

monary area; the pulmonary second sound was normal. The lungs were clinically normal.

The electrocardiogram was normal but the chest radiograph showed a large shadow in the region of the pulmonary bay and aortic knuckle (Fig. 1). This shadow appeared to pulsate on screening. An aortic or pulmonary artery aneurysm was considered as the likeliest diagnosis.

Cardiac catheterization revealed a gradient of 28 mm. Hg between the right ventricle and the pulmonary artery with a right ventricular pressure of 46/5. The mediastinal shadow could not be entered with the catheter. A pulmonary angiogram (Fig. 2) showed that the pulmonary artery was grossly narrowed as it crossed the mediastinum; the peripheral branches were normal. As the right main pulmonary artery was grossly narrowed and bowed inferiorly, we deduced that this must be an intrapericardial mass, or a mediastinal mass with intrapericardial extension. A left thoracotomy revealed a lobulated mass, the size of a golf ball, around the left pulmonary artery. The pericardium was thickened and hyperaemic and an effusion was present; on opening the pericardium a large fleshy tumour mass was found to be surrounding the main pulmonary artery and filling the transverse sinus.

The greater part of this tumour was removed. Histological examination showed it to be a low-grade sarcoma containing myxomatous and vascular areas.

Following removal of the tumour the patient underwent a course of radiotherapy and actinomycin D therapy. He has done well so far and the signs of pulmonary artery obstruction have subsided.

CASE 2 A 23-year-old man was referred to this Centre because of central chest pain and effort dyspnoea. Twelve months previously he had received a kick in the chest whilst playing football and from that time had suffered from intermittent attacks of central chest pain.

FIG. 1. *Case 1. Chest radiograph showing mass in the region of the pulmonary bay and aortic knuckle.*

FIG. 2. *Case 1. Pulmonary angiogram shows gross narrowing of the main pulmonary artery.*

FIG. 3. Case 2. Phonocardiogram showing marked respiratory variation of the pulmonary systolic murmur.

He was a healthy looking young man with a normal pulse and jugular venous pressure. A systolic thrill was felt over the pulmonary outflow tract and this was associated with a grade 3/3 mid-systolic murmur. The most striking feature of this murmur was that it disappeared almost completely on deep inspiration, whereas it was loudest at the end of full expiration (Fig. 3). The pulmonary second sound was loud but normally split. The other systems were clinically normal.

The electrocardiogram was normal but the chest radiograph showed mediastinal widening around the roots of the great vessels (Fig. 4); lateral views suggested that this was an anterior mediastinal mass (Fig. 5).

Cardiac catheterization revealed a gradient of 10–15 mm. Hg over the right ventricular outflow tract with a right ventricular pressure of 35/0. The right pulmonary artery could not be entered with the catheter.

A pulmonary angiogram showed compression of the main pulmonary artery, and particularly the right pulmonary artery, by a rounded mass descending from the superior mediastinum (Fig. 6). A biopsy of this mass was obtained through a small right anterior thoracotomy; histological examination showed it to be a benign teratoma composed of numerous mature tissues, viz., skin, respiratory epithelium, gastrointestinal epithelium, cartilage, muscle, sebaceous glands, and mucous glands.

A median sternotomy was performed, revealing a large solid tumour, the size of a cricket ball, arising from the body and lower poles of the thymus gland. This tumour was dissected off the pericardium, the innominate vein, the superior vena cava, and the main pulmonary artery. There was an obvious depression over the pulmonary outflow tract where a knuckle of the tumour had been pressing.

FIG. 4. Case 2. Chest radiograph showing widening of the mediastinum around the root of the great vessels.

FIG. 5. Case 2. Lateral chest radiograph showing the mass in the anterior mediastinum.

Signs of serious cardiac tamponade appeared on the eighth post-operative day; 500 ml. of straw-coloured fluid was aspirated and this was followed by a rapid improvement in the patient's condition. On discharge from hospital all his cardiac signs had disappeared.

CASE 3 A 32-year-old man presented at another hospital complaining of a 'flu-like illness associated with severe central chest pain and dyspnoea. On admission his chest radiograph showed that a pericardial effusion was probably present and the electrocardiogram was compatible with pericarditis. A pericardial aspiration produced 500 ml. of yellow fluid which had a high white cell count with 50% polymorpholeucocytes.

On admission to this centre he looked in good general condition and there were no signs of cardiac tamponade. There was no lymphadenopathy. A marked right ventricular lift was present and auscultation revealed a grade 2/3 mid-systolic murmur in the pulmonary area. This murmur was loudest at the end of full expiration and became very soft on deep inspiration. The pulmonary second sound was normal. A small left-sided pleural effusion was detected clinically.

The cardiac contour on the chest radiograph (Fig. 7) suggested that a small pericardial effusion was still present. There was prominence of the left side of the superior mediastinum; tomograms of this region suggested that a widespread mediastinal tumour was present.

The pleural aspirate was a pale, turbid fluid and did not contain malignant cells. At bronchoscopy the left main bronchus was found to be compressed from without at the level of the carina.

The left scalene node was excised and histological section showed this to be Hodgkin's disease.

The patient was given a course of intravenous nitrogen mustard followed by radiotherapy, which

FIG. 6. Case 2. Antero-posterior angiogram showing compression of the main pulmonary artery.

FIG. 7. Case 3. Chest radiograph showing an opacity in the left hilar region and a left pleural effusion.

produced shrinkage of the mediastinal mass and disappearance of the cardiac murmur.

CASE 4 A 20-year-old woman who, six weeks before admission, had been struck on the left side of the chest during a party, complained that since then she had noticed the gradual onset of effort dyspnoea, left-sided chest pain, and palpitations; she had also had one fainting attack after exertion.

Physical examination revealed a healthy girl with a normal pulse and jugular venous pressure; the blood pressure was 100/70 mm. Hg. A marked right ventricular heave was present and on auscultation a grade 3/3 mid-systolic murmur was heard at the left sternal edge. The pulmonary second sound was loud but normally split. Both the murmur and the pulmonary second sound diminished markedly on inspiration.

Examination of other systems was essentially normal and there was no evidence of muscle weakness on prolonged effort.

A chest radiograph (Fig. 8) showed a large rounded shadow in the anterior mediastinum.

Cardiac catheterization revealed a gradient across the pulmonary outflow tract of 20–30 mm. Hg with a pulmonary artery pressure of 15/4 and a right ventricular pressure of 50/0–7 mm. Hg.

Pulmonary angiography demonstrated gross narrowing of the pulmonary outflow tract. The diagnosis was considered to lie between a tumour and a haematoma pressing on the outflow tract of the right ventricle.

A left anterior thoracotomy was performed revealing a large, hard tumour surrounding the outflow tracts of the heart. A frozen section of this mass showed it to be a highly malignant tumour. During this procedure cardiac arrest occurred and resuscitation was not possible.

At necropsy a firm tumour mass was found filling the superior mediastinum and surrounding and compressing the great vessels. It was directly invading the upper lobes of both lungs and was adherent to the pericardium and projected into the lumen of the right atrium. There was massive right ventricular hypertrophy; the posterior wall of the right ventricle measured 22 cm. whilst the left ventricle measured 18 mm. Lymph node metastases were present down to the diaphragm. Histological section of this tumour showed it to be a malignant thymoma.

DISCUSSION

Winter (1958) and Waldhausen *et al.* (1959) have postulated that the rarity of this syndrome is due to the fact that anterior mediastinal tumours tend

FIG. 8. Case 4. Chest radiograph showing mass in left hilar region extending downwards.

to expand laterally into both sides of the chest and therefore need not necessarily encroach on the great vessels.

In an analysis of their series of cases, Seymour *et al.* (1968) found that the triad of chest pain, dyspnoea, and a loud pulmonary systolic murmur was the best guide to a diagnosis of the syndrome of pulmonary artery compression. Shaver *et al.* (1965), in a review of the published cases, came to the same conclusion. All four of our patients presented with chest pain and dyspnoea and all were found to have a pulmonary systolic murmur. No satisfactory explanation has been given as to why chest pain should be so common in this condition.

Gough *et al.* (1967) have pointed out that there are no specific physical signs to indicate that the pulmonary stenosis is due to extrinsic pulmonary compression and not, for example, congenital subvalvular or supravalvular stenosis. However, we have been impressed by the respiratory variation of the pulmonary murmur in three of our patients. As demonstrated in Fig. 3, the murmur

virtually disappears at the height of inspiration presumably when the tumour is lifted off the pulmonary artery, whilst at the end of full expiration, when the tumour presses against the artery the murmur is impressively loud. We feel that this may be a useful sign in the diagnosis of extrinsic compression of the pulmonary artery.

The essential investigation to establish that the stenosis is in fact extrinsic is a pulmonary angiogram; cardiac catheterization alone simply reveals the degree of the obstruction. Angiocardiography was not performed in case 3 since a diagnosis of Hodgkin's disease had been made on scalene node biopsy; the disappearance of the pulmonary murmur after radiotherapy confirms that the stenosis was almost certainly due to pressure from mediastinal glands.

The impressive right ventricular hypertrophy found at necropsy in case 4 suggests that compression had been present for a long time without producing symptoms. In case 1 the major part of the sarcomatous mass was intrapericardial. Only two other cases of an intrapericardial tumour

producing pulmonary artery compression have been recorded (Waldhausen *et al.*, 1959; Seymour *et al.*, 1968); these tumours were a mesothelioma and a sarcoma respectively.

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