Congenital pericardio-peritoneal communication with herniation of omentum into the pericardium

A rare cause of cardiomegaly

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A patient with a congenital defect of the pericardium with herniation of the greater omentum within the pericardial cavity is described; the condition was recognized during life. The value of echocardiography as a diagnostic tool is emphasized. Successful surgical treatment was carried out.

Congenital defects of the pericardium are rare (Ellis, Leeds, and Himmelstein, 1959) and only 10 cases where the defect was in the diaphragmatic portion of the pericardium have previously been described. This report records a patient in whom a considerable quantity of omentum had herniated through such a defect and presented as an unusual cause of cardiomegaly. The patient was successfully treated by operation.

Case report

A 28-year-old male Saudi Arabian army officer had, for five years, been complaining of intermittent sharp pains in his left shoulder blade lasting for a few seconds, not related to exertion. During the past 6 months before clinical examination in Saudi Arabia in April 1971 the pain had become more continuous, was located generally in the left chest, was unrelated to respiration but was made worse by lying on his left side, by raising his arms above his head, eating a large meal, or carrying heavy weights. He had lost 6 kg in weight during these 6 months. There was no cough, dyspnoea, vomiting, or ankle oedema. At this time his heart shadow was radiologically enlarged and a pericardial effusion was suspected, but two attempts at diagnostic pericardial aspiration produced no fluid. A clinical diagnosis of tuberculous pericarditis was made and a 3-month course of antituberculous chemotherapy was started. As his symptoms continued he was referred to one of us (J.F.G.) at Hammersmith Hospital for further investigation in September 1971. His general health was otherwise good and he had had no serious illness in the past. His father and mother were alive and well, as were 4 sisters and 1 brother; however, 4 other brothers

had died between the ages of 1 and 5 with no ascertained cause.

Physical examination revealed a fit, well-built, short man, with regular sinus rhythm and normal peripheral pulses without detectable paradox. His jugular venous pressure was slightly raised with 'a' and 'v' waves of 4 to 5 cm respectively at 45°, and his blood pressure was 110/80 mmHg, falling to 105/80 mmHg on inspiration. The cardiac impulse was not palpable. There was physio-

FIG. 1 Six foot posteroanterior chest x-ray showing cardiac enlargement.

FIG. 2 Angiocardiogram showing gross separation of ventricular cavities from pericardium. SVC injection outlining superior vena cava, right atrium, right ventricle, main pulmonary artery, and right pulmonary artery. P, pericardium; SVC, superior vena cava; RV, right ventricle; MPA, main pulmonary artery; RA, right atrium; RPA, right pulmonary artery.

logical splitting of the 2nd sound and a 4th heart sound was audible at the apex. His trachea was central and chest was clear to percussion and auscultation. Nothing abnormal was detected on abdominal examination.

Investigations: Hb, 15.1 g/100 ml; RBC, 4.96 million/mm³; PCV, 45 per cent; platelets, 135,000 per mm³; WBC, 4300 per mm³; ESR, 10 mm at 1 hour; urea, 29 mg/100 ml; electrolytes, normal; uric acid total protein, serum aspartate aminotransferase, albumin, calcium, inorganic phosphate, alkaline phosphatase, all normal; fasting cholesterol, 260 mg/100 ml; fasting triglyceride, 100 mg/100 ml; glucose tolerance test, normal; protein electrophoresis, normal; Mantoux test at 1/1000, positive; the electrocardiogram showed sinus rhythm, right axis deviation, and non-specific repolarization changes in leads II and V3-V7.

Phonocardiography (low frequency) confirmed, in addition to the first heart sound and physiological splitting of the second heart sound, the presence of a fourth heart sound.

Echocardiography showed normal apposition of echoes from anterior chest wall and heart wall, and thus no evidence of pericardial effusion. The mitral valve echoes were normal.

Plain radiography of the chest revealed moderate generalized cardiac enlargement with some prominence of the main pulmonary artery shadow. Pulmonary vascularity

was within normal limits (Fig. 1). Screening did not reveal any abnormality. Cardiac catheterization showed raised right atrial pressure of 10 mmHg (mean), slight increase in right ventricular end-diastolic pressure (30/5-9 mmHg), left atrial (wedge) pressure of 10 mmHg (mean), and slight increase of left ventricular end-diastolic pressure to 18 mmHg. The results suggested slight cardiac constriction.

Angiocardiography revealed gross separation of right and left ventricular cavities from the pericardium suggesting pericardial effusion, probably encysted, or the presence of a mass in the pericardial cavity (Fig. 2).

A diagnosis of loculated pericardial effusion was made. The patient was admitted for surgical exploration to exclude a tumour, to obtain a pericardial biopsy to establish the diagnosis, and for decompression of the heart. Operation was carried out on 18 October 1971.

Operation A left anterior thoracotomy was performed through the 5th intercostal space. The pericardium was incised and a large mass of omentum surrounding the inferior aspect of the heart was discovered. There was a 2.5 cm communication between the peritoneum and the pericardium lying anteriorly, just to the left of the mid-line and about 2.5 cm posterior to the sternocostal attachment of the diaphragm. The omentum could not be returned to the abdomen even after opening the abdomen through a midline incision. Part of the omentum was therefore resected and the remainder then reduced readily.

Stretching from the diaphragm just below the posterior margin of the defect and entering the inferior aspect of the right atrium or coronary sinus was a wide vascular channel about 1.5 cm in diameter - this presumably represented a persistent left-sided inferior vena cava. It was not possible, however, to identify the right inferior vena cava for technical reasons. The canal was securely closed with several interrupted sutures and the chest was closed with drainage.

Pathological examination of the resected omentum showed coarsely lobulated yellow-white fibro-fatty tissue. The entire specimen appeared to be mainly fat and a small amount of fibrous tissue, consistent with omentum. Microscopical examination revealed fatty tissue containing blood vessels.

Discussion

Absence, or deficiency, of the pericardium is one of the rarest of the congenital malformations. Historically, it is of interest that the first case of absence of the pericardium was described by Realdus Columbus in 1559 though this condition was first accurately

described by Baillie in 1793. Since then reports of individual cases have been published occasionally.

The partitioning of the pericardial cavity from the rest of the coelom is initially indicated by the formation of transverse septum through a transversely placed mass of mesoderm situated just caudal to the heart and extending dorsally from the ventral body wall. The developmental separation of the pericardial and pleural cavities is brought about by the growth of additional folds which became associated with the transverse septum.

While the precise mechanism is debated it is generally agreed that congenital pericardial defects result from incomplete development of either the transverse septum or of the pleuropericardial folds.

The great majority of these pericardial defects are on the left side; this was also the case in the patient described in this report. This occurrence on the left side may be due to premature atrophy of the left duct of cuvier (Perna, 1910) resulting in an incomplete left pleuropericardial membrane (a left pericardial defect).

The continuity between the pleural and pericardial cavities exposes the heart to pulmonary infection, and in 27 per cent of cases with pericardial defect death was due to pulmonary infection complicated by pleuropericarditis (Abbott, 1909).

The patient described in this report complained of almost continuous heaviness and discomfort around the left costal margin, unrelated to respiration, but usually after a heavy meal or carrying heavy weights. The pain may have been due to stretching of the pericardium as a result of the presence of a large mass of omentum or could have arisen from herniated omentum through the defect.

A clinical diagnosis of congenital pericardial deficiency is very difficult during life but may be suspected from an unexplained displacement of the heart and also abnormal mobility of the heart. Unexplained cardiac enlargement, if present, would be further evidence in favour of the diagnosis though its absence would not be significant (Southworth and Stevenson, 1938).

In general small defects or foramina in the left pericardium would not be expected to produce a specific abnormality on plain x-rays of the chest unless either heart or lung had herniated through the defect; in the latter case chest x-rays might show a mediastinal mass or deformity, but no such findings have been described to our knowledge.

The majority (73%) of these defects appear to affect men (Ellis *et al.*, 1959); this fact is also supported by the patient described in this communication.

Left-sided lesions vary from small defects with smooth edges in close relation to the left pulmonary

artery to total absence of the pericardium on the left side. Such defects are not uncommonly associated with cysts in the adjacent lung.

There are two groups of left-sided defects. In the first group which comprised the majority, there is a complete defect, the left pleural space and the pericardial cavity constituting one space, and the cardiac mass is not constrained by the left pericardium. In the second group, the minority, the pericardial defect is incomplete, and takes the form of a foramen. Our patient belongs to the second category.

Pericardial defects *per se* are rarely the cause of cardiac symptoms. Two cases have been reported in which death occurred which could be attributed to herniation of the heart through the defect, resulting in tension. This, however, is a rare event. Occasionally when the heart is partially outside the pericardium, pressure from the firm rim of the aperture may press on one or other of the coronary vessels and cause some degree of obstruction. No such factor appeared to be operating in the case reported here. Though the heart is enlarged in 50 per cent of the cases, there is nothing to suggest that this enlargement is due to the defect. Dahl (1937) identified gas in the pericardium associated with a left therapeutic pneumothorax for tuberculosis, and made the diagnosis of congenital pericardial defect. Rusby and Sellors (1945) noted gas about the heart after a diagnostic left pneumothorax for lung mass (cyst), but its significance was not appreciated until after a small left pericardial defect had been found at operation. A pneumothorax may not always yield diagnostic information, since in some cases of deficiency in the left pericardium, adhesions might prevent accumulation of gas over the surface of the heart; it was not attempted in the case discussed in this report, nor would it have provided any useful information as the defect did not communicate with the pleura.

Cardiac fluoroscopy is not expected to provide much help in the positive diagnosis of pericardial deficiency except occasionally when it might show unusual cardiac silhouette or abnormal mobility of the heart. In the case under discussion fluoroscopy did not reveal any abnormality.

Angiocardiography helps to rule out other cardiac abnormalities and makes the configuration of various cardiac chambers obvious. In the case under discussion angiocardiography revealed gross separation of right and left ventricular cavities from the pericardium suggesting an encysted pericardial effusion or the presence of a mass in the pericardial cavity. On cardiac catheterization the abnormal haemodynamic findings of a raised right ventricular end-diastolic pressure signified right and left ventricular dysfunction or external compression of cavities of

the chambers by fluid or mass in the pericardial cavity.

Of particular interest was the echocardiogram which showed no evidence of pericardial effusion when angiography suggested fluid or a mass.

This association of clinical and angiocardiographic evidence suggesting pericardial fluid with negative echocardiographic evidence may well prove to be of value in diagnosis. Since tissue in the pericardial space presents interfaces to reflected ultrasound it produces echoes similar to those of both chest wall and heart; the characteristic separation of the echoes from the wall of the heart and the structures outside the pericardium which identifies a pericardial effusion therefore does not occur.

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