CHRONIC EFFUSIVE PERICARDITIS*

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The clinical features of pericardial effusion vary according to the volume of fluid effused, the rate at which it accumulates, and the nature of the underlying disease. An acute pericardial effusion of rheumatic or pyogenic origin may provoke dramatic symptoms which were recognized and described long ago. Cardiac tamponade was described by Richard Lower in 1669, as follows.

“the walls of the heart are compressed by fluid settling everywhere so that they cannot dilate sufficiently to receive blood; then the pulse becomes exceedingly small until it becomes utterly suppressed by the great inundation of fluid, whence succeed syncope, and death itself.”

Older physicians stressed the tendency to syncope, and the extreme orthopnoea which sometimes provoked characteristic postures aptly portrayed in the “pillow sign” of Blechmann (Fig. 1) and the “Mohamedan’s Prayer” sign of Hirtz (1911) (Fig. 2). According to Blechmann (1922), squatting, or *accroupissement*, was described as characteristic of pericardial effusion long before Fallot described his tetralogy, and indeed it was mentioned by Zehetmayer of Vienna in his textbook of 1845.

When the effusion accumulates more gradually, as in tuberculous pericarditis, the symptoms are far less dramatic, and in chronic effusive pericarditis with which this lecture is concerned, patients often continue at work for years with a massive pericardial effusion. Indeed one of my patients had played a round of golf shortly before over 6 pints of fluid were aspirated from his pericardium at successive paracenteses, and I shall start by describing this case.

CASE REPORTS

Case 1. P.W., a publican aged 41, first seen in October 1949, with Dr. Barrington Prowse, had had rheumatic fever at the age of 8, followed by signs of mitral disease. For many years, he had consumed a bottle of whisky a day, and was known to be hypertensive. In 1948, he developed paroxysmal nocturnal dyspnoea and was treated for heart failure, but recovered well and returned to work.

In September 1949, nocturnal dyspnoea recurred and was accompanied by fever, tachycardia, and signs of consolidation at the base of the left lung. Bacterial endocarditis was suspected, but there were no embolic signs and blood cultures were negative. Nevertheless, he was given a month’s course of penicillin, and routine therapy for heart failure.

Seen at this time (October 1949), the apex beat was in the anterior axilla, there was widespread palpable præcordial pulsation, and a systolic murmur and thrill at all areas, suggesting an enlarged heart with rheumatic mitral disease. Blood pressure was 170/112 mm. Hg. The venous pressure was slightly raised, the liver was palpable, and there was slight oedema. The electrocardiogram showed a typical “left heart strain” pattern. Radiography showed a huge globular heart shadow suggestive of pericardial effusion (Fig. 3), but in view of the clinical signs, the rheumatic valve disease, and hypertension, an enlarged heart was diagnosed.

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FIG. 1.—The “pillow sign” of acute pericardial effusion (from Blechmann, 1922).

FIG. 2.—The “Mohamedan's Prayer” sign of acute pericardial effusion (from Blechmann, 1922).

Fig. 3

Fig. 4

Fig. 5

FIG. 3.—Radiograph from Case 1 when first seen in 1949, showing pericardial effusion.

FIG. 4.—Radiograph of Case 1 after paracentesis and air replacement, showing large heart, prominent pulmonary trunk, and parietal pericardium.

FIG. 5.—Radiograph of Case 1 in 1953, after pleuro-pericardial fenestration, showing enlarged heart.

He recovered well, returned to work, and carried on without difficulty for the next two years, leading an active life including golf, and taking alcohol very freely. His average blood pressure was 210/120 mm. Hg, and radioscopy always showed a huge heart shadow with diminished pulsation, often recorded as "like a pericardial effusion".

In 1952, he developed ascites, attributed to cirrhosis of the liver, and was persuaded with difficulty to come into hospital. By this time, a pericardial effusion was suspected and was confirmed by angiocardio-graphy. Paracentesis of the pericardium produced 25 oz. of clear pale yellow fluid, s.g. 1021, containing 6.1 g. of protein per 100 ml. The fluid was sterile and cultures for tubercle bacilli were negative. Several further paracenteses yielded a total of 5½ pints (3 litres) of fluid. Air replacement after paracentesis showed an enlarged heart with prominent pulmonary trunk; the parietal pericardium did not appear to be grossly thickened (Fig. 4). He improved and left hospital but the effusion soon reaccumulated, and another 3 litres of hæmorrhagic fluid were aspirated at successive taps. He became anæmic, and developed a left pleural effusion, so that surgical treatment was advised. Before operation, another 3½ pints (2 litres) of hæmorrhagic fluid were aspirated. In September 1952, Sir Thomas Holmes Sellors resected a window, 4×3 in. (10×8 cm.), in the parietal pericardium, opening into the left pleural cavity. The pericardium was ¼ in. (0.6 cm.) thick, fairly soft, and pliable, and it contained several pints of hæmorrhagic effusion. Sections of the excised pericardium showed fibrous thickening and foci of cellular infiltration, but no evidence of tuberculosis or any other specific ætiology. A left pleural effusion was aspirated after operation and he left hospital quite free from signs of heart failure, but with a considerably enlarged heart (Fig. 5).

He continued at work for the next four years, taking alcohol very freely and medical treatment very irregularly. There was no recurrence of the pericardial effusion, but the heart remained enlarged, the signs of mitral disease were unchanged, and the hypertension persisted.

In 1958, he developed atrial fibrillation, since which he has frequently been in hospital with heart failure, usually precipitated by alcoholic excess, respiratory infection, cessation of treatment, or physical over-activity, but always responding to routine therapy.

Comment. This patient's pericardial effusion probably developed in 1948 and was certainly present in 1949, after which it persisted unchanged for three years during which he remained at work almost continuously. Altogether, 19 pints of fluid were aspirated from the pericardium before and during operation.

The distinction between a large heart and a pericardial effusion in an acutely ill patient is a time-honoured problem but not one that often arises in an ambulant patient carrying on his normal duties. When I encountered this case, I was quite unfamiliar with the condition of chronic massive pericardial effusion without tamponade in an afebrile patient, and so it took two years to reach the correct diagnosis.

The ætiology of the pericardial effusion has not been determined. All tests for tuberculosis, including histological examination of the pericardium, were negative, lupus erythematosus cells were never found, and there was no evidence of myxœdema or other known cause of pericardial effusion. Recently, Dr. Doniach has carried out tests for auto-antibodies, and the immunofluorescent nuclear staining test gave a strongly positive result for antinuclear factors. This suggests some form of auto-immune disease and I shall discuss this later.

The main feature of the case was the disproportion between the huge heart shadow and the relatively mild disability. The lesson learned was to think of chronic pericardial effusion whenever the heart seems too large for the symptoms, and I soon encountered another such case.

Case 2. W.E., a male clerk, aged 48, was found by chance to have an enlarged heart at mass miniature radiography and was, therefore, sent to out-patients' department in September, 1951. He had no complaints, and his heart had never been questioned in the past.

Radiographs showed a grossly enlarged heart shadow, cardio-thoracic ratio (C.T.R.) 73 per cent, with narrow vascular pedicle, and no hilar congestion; on screening, pulsation was diminished (Fig. 6). The apex beat was easily felt 5½ in. (14 cm.) from the mid-line, there was a triple rhythm, and moderate apical systolic murmur. Blood pressure was 120/80 mm. Hg. The electrocardiogram showed a "left heart strain" pattern with bifid P waves in leads 1 and 2. The diagnosis recorded was cardiomegaly of unknown origin. By the time I saw him again the following year, I had learned my lesson, and admitted him to hospital where

FIG. 6.—Radiograph of Case 2, when first seen in 1951, showing pericardial effusion with narrow vascular pedicle and absence of hilar congestion.

FIG. 8.—Radiograph of Case 2 in 1955, showing subsidence of effusion three years after paracentesis and diuretic therapy.

an angiocardiogram disclosed a large pericardial effusion (Fig. 7). Paracentesis yielded 20 oz. of clear serous fluid which was sterile, and guinea-pig inoculation was negative for tubercle bacilli.

He had two brief paroxysms of fibrillation in hospital and a mild pleural reaction after paracentesis. In the absence of cardiac symptoms no further paracentesis was done; he left hospital and returned to work. The pericardial effusion persisted unchanged for a year, after which it gradually subsided (Fig. 8). When

FIG. 7.—Angiocardiogram of Case 2, showing right atrial border within the pericardial effusion.

last seen in 1957, he was well and at work, but died suddenly at a railway station a few months later. A coroner's post-mortem examination reported left ventricular hypertrophy and fibrous degeneration of the myocardium without coronary occlusion. The cause of death was given as heart failure due to hypertension, though his blood pressure had always been normal.

Comment. This chronic pericardial effusion of unknown duration was discovered by chance. It persisted for two years in spite of paracentesis and caused little disability. His cardiac enlargement must be regarded as due to idiopathic hypertrophy or cardiomyopathy. As in Case 1, the clinical signs of an enlarged heart delayed diagnosis of the pericardial effusion, but again the clue was a heart shadow too large for the symptoms.

Case 3. E.B., a lady aged 60 with a bronchitic history, had been unable to sleep flat in comfort for two years, when a large heart was found at chest radiography, and she was referred as an outpatient in 1951 by Dr. A. G. Emslie who suspected a pericardial effusion.

On examination, she appeared acromegalic. She was afebrile, with a normal pulse rate, and blood pressure 190/130 mm. Hg. Apex beat in anterior axilla was diffuse. Heart sounds were distant; no murmurs. Radiography showed a huge pyriform heart shadow (C.T.R. 79%) which pulsated little on screening, but the heart contour changed noticeably with change of posture (Fig. 9). The electrocardiogram was normal. There were no signs of failure or of tamponade. A diagnosis of chronic pericardial effusion was made and she was admitted to hospital where angiocardiology confirmed the diagnosis (Fig. 10).

Paracentesis yielded 26 oz. of clear fluid, s.g. 1014, containing 1.6 g. protein/100 ml., and scanty endothelial cells and leucocytes. It was sterile and cultures for tubercle bacilli were negative. A second aspiration yielded blood-stained fluid.

She improved, but the effusion persisted for about a year, after which it subsided gradually, leaving a moderately enlarged heart (C.T.R. 62%) (Fig. 11). She is still alive, aged 72, but is unable to travel following a cerebral vascular episode in 1961.

Comment. This acromegalic patient with moderate hypertension and enlarged heart probably had a chronic pericardial effusion of two years' duration when first seen, and, in spite of paracentesis, it persisted for another year. No ætiological cause was found and tuberculosis was reasonably excluded.

FIG. 9.—Radiographs of Case 3 when first seen, (A) vertical and (B) horizontal posture, showing change from pyriform to globular contour. Note absence of hilar congestion and short narrow pedicle.

FIG. 10.—Angiocardiogram of Case 3, showing pericardial effusion extending beyond right atrial border.

FIG. 11.—Radiograph of Case 3, 12 years later, showing subsidence of effusion.

Case 4. In 1957, when visiting Kingston, Ontario, I saw a man of 59, with a large chronic pericardial effusion, who was carrying on his work as a postmaster, and Professor Ford Connell has kindly allowed me to record this unpublished case of his.

The patient had been somewhat short of breath for about 10 years but took little notice of it. In 34 years he had only been off duty from illness for one week until 1955 when he had a feverish cold, followed by nocturnal dyspnoea, but he continued to work until a large heart was discovered at miniature radiography after which he rested at home for five months. He was admitted to hospital for investigation in April 1956.

On examination, he was afebrile, the jugular venous pressure was raised, the liver enlarged, and there was moderate peripheral oedema. Radiography showed a massive pericardial effusion, confirmed by angiocardiography, at which the heart itself looked normal in size. The electrocardiogram showed low voltage QRS and T waves. Blood count was normal. Erythrocyte sedimentation rate was 22 mm./1 hour. Blood cholesterol 202 mg./100 ml. Histoplasmin skin test positive.

Paracentesis of pericardium yielded 500 ml. and 1425 ml. of brownish fluid at successive taps, and the heart shadow was reduced almost to normal size. The pericardial fluid was sterile, culture for tubercle bacilli negative, and contained amorphous protein material and a few leucocytes, but no malignant cells.

When he left hospital, the effusion re-collected and during the next 20 months he was re-examined in hospital on six occasions, the effusion persisting (C.T.R. 70%). Further aspirations produced sterile fluid. Test for lupus erythematosus cells negative. He remained at work with few complaints until November 1962, when he had an attack of retrosternal pain lasting 18 hours. The electrocardiogram remained normal, and radiography still suggested a pericardial effusion though less massive than when first seen.

Comment. This patient probably had a longstanding pericardial effusion when first seen, and in spite of numerous aspirations, it has persisted for 6 years during which he has remained ambulant and at work up to the age of 64. He had nocturnal orthopnoea and slight tamponade when first seen, otherwise the effusion has been well tolerated. No aetiological cause has been discovered.

Case 5. A.G., a man aged 69, at work as a toolmaker, was seen as an out patient in June 1957 on account of hypertension and slight oedema of the legs due to varicose veins, but he denied having any cardiac symptoms.

Examination revealed signs of aortic incompetence, a blood pressure of 220/60 mm. Hg, but no evidence of heart failure. Radioscopy showed an enormous pyriform heart shadow, C.T.R. 70 per cent (Fig. 12A). The electrocardiogram showed a slight digitalis effect only. The absence of symptoms with such a large heart suggested a chronic pericardial effusion, and he was admitted to hospital.

FIG. 12.—Radiographs of Case 5, (A) when first seen, showing pyriform contour of chronic effusive pericarditis and (B) five years later, with larger pericardial effusion.

Angiocardiography demonstrated a considerable pericardial effusion. The Wassermann reaction was positive, but other investigations were negative. He was given a course of penicillin and mercurial diuretics, after which his heart shadow became smaller (C.T.R. 66%).

He returned to work, and continued reasonably well for the next five years, though the pericardial effusion persisted. In August 1962, he had an attack of dyspnoea and a huge pericardial effusion was apparent on radiography, C.T.R. 93 per cent (Fig. 12B).

Paracentesis of the pericardium yielded 30 oz. of clear serous fluid containing 5 g. protein/100 ml., and scanty leucocytes. The culture was sterile and tests for lupus erythematosus cells were negative. Test for anti-nuclear factor was strongly positive. Liver and spleen were slightly enlarged, trivial oedema, but no rise of venous pressure. The electrocardiogram showed low voltage QRS and T waves, with inversion of T in left chest leads. Following paracentesis and diuretics the effusion became smaller and he left hospital, but the fluid soon re-collected, and he has just been readmitted with an enormous pericardial effusion, of which two pints have been aspirated. The electrocardiogram now shows atrial fibrillation.

Comment. This elderly patient appeared to have syphilitic aortic incompetence and a massive pericardial effusion which was well tolerated. The positive test for anti-nuclear factor raised the possibility of a collagen disease causing a false positive Wassermann reaction. Surgical treatment is under consideration, but in view of his advanced age he has been put on steroid therapy, without as yet any dramatic benefit.

Case 6. K.K., an Indian woman, aged 57, was admitted to St. Stephen's Hospital in 1962 as a case of cardiomegaly of unknown origin with heart failure. The history was vague, but she had been treated for heart trouble in India in 1958, and in 1959 developed Raynaud's disease of the hands and feet.

Investigations by Dr. Harvey suggested a chronic pericardial effusion with tamponade, and paracentesis yielded 800 ml. of straw-coloured fluid which later became blood-stained. The fluid was sterile and guinea-pig inoculation was negative. Air replacement suggested a normal heart size (Fig. 13 (A) and (B)). All investigations as to the cause of the effusion were negative, but she was given anti-tuberculous therapy. She failed to improve and was transferred to the Middlesex Hospital in August 1962, with a huge pericardial effusion (Fig. 14A), small pulse, raised venous pressure, enlarged liver, and slight oedema. Heart sounds were distant with triple rhythm. Blood pressure 100/70 mm. Hg. Pulse was 80 regular; she was afebrile. There was marked scleroderma of hands and feet.

FIG. 13.—Radiographs of Case 6, (A) when first seen, showing pericardial effusion and (B) after paracentesis and air replacement, showing parietal pericardium, normal-sized heart, and residual effusion.

Paracentesis yielded 500 ml. of heavily blood-stained fluid; culture sterile. Tests for lupus erythematosus cells negative. Wassermann reaction negative. Electrophoretic pattern showed increased gamma globulin. Auto-antibody test strongly positive for anti-nuclear factor (Dr. Doniach).

Partial pericardectomy was performed by Sir Thomas Holmes Sellors in September 1962. At operation, the pericardium was thickened and contained 400 ml. of effusion, but no evidence of tuberculosis was found, and the heart appeared normal in size. Sections of the pericardium showed dense hyaline fibrosis with perivascular lymphocytic infiltration compatible with scleroderma pericarditis. After operation, she improved rapidly, the venous pressure fell, and the heart shadow became quite small (Fig. 14 (B)). When last seen as an out patient, she was well, and the effusion had not recurred.

FIG. 14.—Radiographs of Case 6, showing (A) recurrence of pericardial effusion after paracentesis, and (B) five months later, after pericardectomy.

Comment. Pericardial involvement is found in about two-thirds of all cases of scleroderma examined *post mortem* (Oram and Stokes, 1961), but large effusions are unusual. Meltzer (1956) has described two cases of massive pericardial effusion in generalized scleroderma, neither of which had signs of cardiac tamponade, and he emphasizes that scleroderma pericarditis may have a protracted course.

To illustrate the variety of underlying heart conditions that may be complicated by chronic pericardial effusion I shall briefly cite one further case.

Case 7. K.B., a woman, aged 50, was sent to the out-patient department in March 1957, with ascites and heart failure, diagnosed as constrictive pericarditis. At the age of 6 she had had "rheumatism" but remained well up to the age of 40, when she had several blackouts within a few weeks. At the age of 44, she developed dyspnoea and ascites which had been tapped repeatedly ever since, and lately as much as 30-40 pints had been removed every few weeks. She was admitted to hospital for investigation.

Examination showed atrial fibrillation, a very large heart, a blowing mitral pan-systolic murmur, a raised venous pressure, enlarged liver, oedema, and massive ascites. The left radial pulse was small and

FIG. 15.—Radiograph of Case 7, showing chronic pericardial effusion and calcified aorta, probably old dissecting aneurysm.

delayed. Blood pressure in the left arm was 90/70 mm. Hg, compared with 160/80 mm. Hg in the right arm. The electrocardiogram showed atrial fibrillation and left bundle-branch block. Radiography showed a large pyriform heart shadow (C.T.R. 79%) with heavy calcification of the dilated ascending aorta and arch (Fig. 15). Angiocardiography showed a considerable pericardial effusion surrounding a large heart. Wassermann reaction negative. Serum protein 4.7 g.; albumin 2.9 g., globulin 1.8 g. The test for lupus erythematosus cells was negative. Paracentesis was difficult owing to the ascites, but 10 ml. of blood-stained fluid were aspirated. She was treated by diuretics and abdominal paracentesis and left hospital free from ascites and much improved, having lost two stone in weight. She died at home in March 1958.

Comment. While the clinical course seemed characteristic of constrictive pericarditis, the grossly enlarged heart shadow suggested chronic pericardial effusion which was confirmed by angiocardiography and paracentesis. The heavily calcified aorta and diminished left radial pulse suggested the possibility of an old dissecting aneurysm perhaps complicated by hæmopericardium, and this probably occurred at the time of her syncopal attacks 10 years before death.

DISCUSSION

As long ago as 1945, Blumenfeld and Thomas reported a single case of "chronic massive pericardial effusion" following radiotherapy for cancer of the breast; otherwise our knowledge of this condition dates from 1950, when Barker and Johnston described three cases of "chronic pericarditis with effusion", remarking that the disease might run a chronic afebrile course for years and simulate an enlarged heart. Guidotti and Puddu (1954) described two further cases of pericardial effusion persisting for years in ambulant and almost asymptomatic patients, and Contro, De Giuli, and Ragazzini (1955) applied the term "chronic effusive pericarditis" to an effusion of four years' duration in an asymptomatic girl aged 7. Since these early papers, the condition has been further reported by Gonin and Perrin (1956), Goldner and Knoop (1956), Connolly *et al.* (1959), Genecin (1959), Scheuer (1960), and Soulié *et al.* (1960).

These papers indicate that massive pericardial effusion may persist for years as a chronic afebrile condition which is often well tolerated and sometimes virtually asymptomatic. Not infrequently the effusion is discovered accidentally at routine radiography when it is usually mistaken for an enlarged heart. There is rarely any history of acute pericarditis to indicate the date of onset, but the effusion is probably longstanding in most cases when first recognized.

Cardiac tamponade and the distressing symptoms of acute pericardial effusion result from rapid accumulation of fluid under high pressure, but when the effusion develops slowly, the pericardium is gradually stretched and the intrapericardial pressure remains low, so that even a massive effusion does not embarrass the heart unduly. Four of my patients continued at work for years with large effusions, and one of Soulié's, first seen at the age of 9 and followed up for 10 years, was able to take part in competitive sports. The gradual onset of dyspnoea or nocturnal orthopnoea may be the first indication of cardiac embarrassment, and sooner or later signs of right heart failure may appear, but how far this is due to tamponade and how far to underlying heart disease is difficult to decide. One of my patients (Case 5) presented some of the typical clinical features of constrictive pericarditis with atrial fibrillation and chronic ascites, but signs of rheumatic valve disease and of gross cardiac enlargement were evidence against such a diagnosis.

Diagnosis. When the heart itself is enlarged, the clinical diagnosis of pericardial effusion is often difficult. The outstanding feature in most of these cases is the disproportion between the apparent size of the heart and the relatively mild symptoms. Whenever the heart is too big for the symptoms we should suspect chronic pericardial effusion, the other condition to which this sometimes applies being Ebstein's disease.

Radiology. The grossly enlarged cardiac shadow is usually pyriform rather than globular in shape, and the lax pericardium allows the contour to change considerably with change of posture (Fig. 9). As in acute effusions, the normal arcs of the cardiac contour are ironed out, and pulsation is diminished or absent, but the vascular pedicle is usually short and narrow, as the superior vena cava is not dilated in the absence of tamponade. The absence of hilar congestion is an important feature in favour of a pericardial effusion rather than a grossly enlarged heart.

Selective angiocardiology provides a simple and safe diagnostic procedure and it is only necessary to inject 10–20 ml. of contrast medium by catheter passed into the right atrium to outline the right heart border within the pericardial effusion. Angiocardiology may also demonstrate pericardial neoplasm, primary or secondary, when it presents as a chronic pericardial effusion (Steinberg, 1960, 1961).

Once the diagnosis is established, a diagnostic paracentesis should be performed, and the fluid removed should be replaced by 100–200 ml. of air, which permits visualization of the parietal pericardium and of the heart within the effusion.

The electrocardiogram never shows the classical pattern of acute pericarditis. It may be normal (Case 3) or may show changes due to the underlying heart lesion, but the most characteristic finding is low voltage of QRS and flattened T waves (Fig. 16). In cases with sinus rhythm bifid P waves are sometimes seen and, as in constrictive pericarditis, atrial fibrillation or flutter may occur eventually as happened in four of my cases.

FIG. 16.—Electrocardiogram of Case 6, showing low voltage and inverted T waves in præcordial leads.

The pericardial fluid is usually clear and straw-coloured, s.g. around 1020, and containing 5–6 g. protein per 100 ml. Cultures are always sterile, and cultures for tubercle bacilli negative. After repeated aspiration, the fluid often becomes blood stained or frankly hæmorrhagic. In cholesterol pericarditis the fluid is usually described as resembling gold paint and contains abundant cholesterol crystals. If the fluid is hæmorrhagic at the initial tap, neoplasm should be suspected and a search for malignant cells should be made, though certain identification is often difficult. However, angiocardiology may be diagnostic.

Aetiology. Excluding tuberculosis from this discussion, chronic pericardial effusion has been reported in scleroderma (Meltzer, 1956), generalized lupus erythematosus (Bergen, 1960; Kong, Kellum, and Haserick, 1962), myxœdema (Kern *et al.*, 1949), chronic anæmia (Soloff and Bello, 1950), after radiotherapy (Blumenfeld and Thomas, 1945), in pericardial neoplasm (Gonin and Perrin, 1956; Steinberg, 1960), and after traumatic hæmopericardium (Tabatznik and Isaacs, 1961).

So-called “cholesterol pericarditis” may be associated with myxœdema, but often it is not and the blood-cholesterol level may be normal (Crow, 1961). Histological examination of the pericardium shows nothing specific apart from cholesterol deposits, and the clinical course differs in no way from that of chronic idiopathic pericardial effusion, so that there is no reason to regard cholesterol pericarditis as a disease *sui generis*.

There is no evidence that chronic effusive pericarditis is associated with any particular form of underlying heart disease. The heart may be normal in size or enlarged from known or unknown causes. Two of my patients (Cases 1 and 3) had hypertension, and Wood (1956) cited 6 cases of chronic pericardial effusion associated with hypertension: 2 (Cases 1 and 7) probably had old rheumatic mitral disease, one with hypertension and the other with extensive aortic calcification suggestive of healed dissecting aneurysm. Alcoholism has been mentioned as an ætiological factor (Goldner and Kroop, 1956), and one of my patients was an alcoholic. In Case 2, gross cardiac hypertrophy of unknown ætiology was found at necropsy. In Case 5, there was aortic incompetence and a positive Wassermann reaction, but the serum also gave a positive test for anti-nuclear factor, suggesting possible collagen disease, and Case 6 had scleroderma. In the majority of reported cases, full investigation failed to show any recognized ætiological cause for the pericardial

FIG. 17.—Radiographs showing (A) pericardial and left pleural effusions complicating neoplasm (thymoma) in woman aged 40, and (B) three months later, after radiotherapy, and (C) 22 years later, aged 62, after several courses of radiotherapy.

effusion, and histological examination of the pericardium at operation or at necropsy has usually shown a chronic non-specific fibrous pericarditis.

It is important to bear in mind that pericardial tumours, innocent or malignant, may present as a chronic pericardial effusion. Some such tumours are operable and others are radio-sensitive, so that their diagnosis is of practical importance. For example, the x-ray picture in Fig. 17(A) was taken over 25 years ago in the case of a woman aged 40 who presented all the clinical signs of pericardial effusion with tamponade. In aspirating blood-stained fluid from the pericardium, I gained the impression of penetrating tumour, and she was treated by radiotherapy. The effusion subsided, and the tumour, probably a thymoma, became evident and responded to further treatment (Fig. 17(B)). She has had several further courses of radiotherapy and is still alive today, aged 66, with a normal heart shadow and a calcific residue of her tumour (Fig. 17(C)). I have also encountered a chronic pericardial effusion in Hodgkin's disease, which responded to radiotherapy. Mahaim (1945) cites the case of a child aged 4 who died from a massive chronic pericardial effusion, and in whom a leiomyofibroma was found at necropsy, which might have been operable; Gonin and Perrin (1956) described two cases of chronic pericardial effusion due to malignant cœlotheliomata.

The occasional occurrence of chronic effusive pericarditis in various collagen diseases raises the possibility of a disturbed immunological mechanism as the underlying cause. Recurrent pericarditis after cardiac infarction, after surgical pericardiotomy, and after traumatic hæmopericardium has been attributed to an auto-immune reaction. In three of my present cases immunological tests supported the idea that an immunopathological reaction was in operation.

The evidence available certainly suggests that chronic effusive pericarditis is usually due to some general cause rather than to any primary disease of the heart or pericardium, and an auto-immune mechanism is therefore a plausible explanation.

Prognosis and Treatment. The clinical course depends a good deal on the underlying heart condition. If the heart is relatively intact, a massive pericardial effusion may be well tolerated for years—over 10 years in several reported cases, and often the condition is longstanding when first recognized. Sooner or later, signs of heart failure may appear, but the symptoms are never comparable with the tamponade of acute pericardial effusion, though one case presented the clinical picture of constrictive pericarditis with recurrent ascites. In Cases 2 and 3, in spite of reaccumulation after paracentesis, the effusion subsided gradually on prolonged diuretic therapy. Both patients who died had serious underlying heart disease.

I have never seen constrictive pericarditis develop in these cases of chronic idiopathic pericardial effusion, and in most reported cases treated surgically, an effusion was still present at operation, but

Scheuer (1960) mentions several cases which did eventually develop constriction while under observation. In regard to treatment, paracentesis may give temporary relief, but the effusion almost invariably reaccumulates, and repeated paracentesis may provoke an undesirable reaction in which the effusion becomes hæmorrhagic or a pleural effusion develops. The only effective treatment is surgical, either pleuro-pericardial fenestration or more radical pericardectomy. In the presence of an effusion, resection of the parietal pericardium is technically far easier and less hazardous than in the case of constrictive pericarditis, and is usually well tolerated even when the heart is enlarged.

Pleuro-pericardial fenestration was successful in abolishing the pericardial effusion in Case 1, and though he later developed heart failure due to underlying heart disease, the effusion has not recurred. However, a more extensive pericardectomy is now advocated for chronic pericardial effusion both in adults (Blakemore *et al.*, 1960) and in children (Roshe and Shumacker, 1959), and also for recurrent idiopathic pericarditis (Zinsser, Johnson, and Blakemore, 1959). In my patient aged 57 with scleroderma, pericardectomy was well tolerated, and since operation the heart has been normal in size and she has been quite free from heart failure. Experience suggests that, once the diagnosis of chronic idiopathic pericardial effusion has been established, surgical treatment should not be delayed as the effusion is unlikely to subside either spontaneously or as a result of paracentesis.

Steroid therapy is often effective in treating pleuro-pericarditis of the post-cardiotomy syndrome, in which an auto-immune reaction has been postulated, and, therefore, it seems reasonable to try it in cases of chronic effusive pericarditis, at least when immunological tests are positive. Dr. W. A. Oille informed me of a case of his which responded favourably to steroids, and, at his suggestion, Case 5 is being treated with prednisone, but so far no dramatic benefit has occurred.

SUMMARY AND CONCLUSIONS

A massive chronic pericardial effusion may exist for years without cardiac tamponade or other classical signs of pericardial effusion, and may be remarkably well tolerated. Often, it is discovered by chance at routine chest radiography and mistaken for an enlarged heart. Eventually signs of heart failure or of mild tamponade may appear, or rarely chronic tamponade with hepatomegaly and ascites simulating constrictive pericarditis.

The main diagnostic clue is the discrepancy between the huge heart shadow in the radiograph and the relatively mild symptoms. Once suspected, the diagnosis is readily confirmed by angiocardigraphy.

Chronic effusive pericarditis may complicate some recognized form of heart disease such as hypertension, rheumatic valve disease, pericardial neoplasms, or traumatic hæmopericardium, and it may be associated with some recognized general disease such as chronic anæmia, myxœdema, scleroderma, generalized lupus erythematosus, or other collagen disease, but in a majority of cases no specific cause for the effusion can be identified. In three of the cases reported, the test for anti-nuclear antibodies was positive, suggesting that an auto-immune mechanism was responsible.

In regard to treatment, the effusion nearly always reaccumulates rapidly after paracentesis, and though it may sometimes subside slowly with prolonged diuretic therapy, the only effective treatment is surgical, either pleuro-pericardial fenestration or a more radical pericardectomy.

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