Recurrent pulmonary embolism due to hydatid disease of heart

Study of 3 cases, one with intermittent tricuspid valve obstruction (atrial pseudomyxoma)


From the Department of Internal Medicine, University Medical School, Madrid 3, Spain

Three cases of pulmonary hypertension caused by hydatid emboli from the right side of the heart are described; cardiac catheterisation was performed in 2. One case was confirmed at operation and 2 at necropsy. The pulmonary emboli were caused by hydatid vesicles in all 3 cases and in none was there pulmonary thrombosis; free scolices were found in the pulmonary alveoli in 2. In 1 patient with repeated syncopal attacks there was a pedunculated cyst in the right atrium which was thought to have intermittently obstructed the tricuspid valve. Gamma radiography, angiocardiography, and necropsy suggested a mechanical cause for the pulmonary hypertension with no vasoconstrictive element. The surgical patient was alive and well 18 months later.

Involvement of the heart in hydatid disease occurs in about 0.5 to 2.0 per cent of cases, and of the right side of the heart in about one-quarter of these (Dévé, 1946; Artucio et al., 1962; Di Bello et al., 1968, 1970). Hydatid embolism has rarely been reported. Three cases are described in which this diagnosis was made during life; 1 was operated on successfully and 2 were confirmed at necropsy. The clinical features are summarised in the Table.

Case reports

CASE 1
A 29-year-old man gave a one-month history of pleuritic pain, first on the left side of the chest and then on the right. In the chest x-ray (Fig. 1) there was a rounded prominence of the left cardiac border which was less opaque in its outer zone. In the right lung base there was a diffuse round shadow which had disappeared 15 days later. The electrocardiogram showed right axis deviation and T wave inversion in leads I and aVL, and a precordial record of a systolic lift to the left of the sternum. Cardiac catheterisation indicated pulmonary hypertension (44/30 mmHg). Right ventricular angiography showed a bulge of the upper part of the interventricular septum with a mobile projection into the outflow tract, and a round shadow in the lower branch of the left pulmonary artery. Coronary arteriography showed an arched displacement of the anterior descending coronary artery.

The deformity of the cardiac outline and the finding of 13 per cent eosinophils in the blood suggested the diagnosis of a hydatid cyst. The Casoni test was positive. The existence of pulmonary hypertension and the angiographic appearance suggested that the cyst was in the upper part of the septum and had perforated into the right ventricle.

Using cardiopulmonary bypass and a medial sternotomy the right ventricle was opened by a transverse incision. A cyst was found in the septum with vesicles in the right ventricle and pulmonary artery (Fig. 2). The contents of these were aspirated and the cyst membrane was excised. Eighteen months later the patient was alive and well, the chest x-ray was normal; the electrocardiogram showed evidence of septal necrosis.

CASE 2
A 63-year-old man gave a one-year history of increasing breathlessness, occasional pain in the right chest, a dry cough, and occasional haemoptysis. For 5 months he had had recurrent syncopal attacks without convulsions lasting for 2 or 3 minutes. These attacks were not related to coughing or to postural changes. At first they occurred only once or twice a month, but in the previous 20 days they happened once or twice a day.
The chest x-ray film (Fig. 3) showed multiple round cystic shadows in the lung fields, and there was a rounded prominence of the right cardiac border in the position of the right atrium. The electrocardiogram showed right ventricular preponderance and evidence of anteroseptal necrosis with an injury pattern.

Eight days later he developed left upper lobe pneumonia caused by a streptococcal infection from which he died.

The clinical diagnosis of hepatic, pulmonary, and probably cardiac hydatid cysts was confirmed at necropsy. A cyst 1.5 cm in diameter was found in the anterior wall of the right ventricle. Another large cyst occupied the right atrium with a pedunculated mass almost occluding the tricuspid valve (Fig. 4). Medium-sized pulmonary arteries were occluded by hydatid vesicles.

Microscopy of the pedunculated mass in the right atrium showed it to consist of hydatid membranes and organising blood clots. Section of the lungs revealed multiple cysts and some free scolices in the alveoli. Some pulmonary arteries were occluded by vesicles but there was no arterial thrombosis. Macroscopical and microscopical studies of the brain were normal.

There seems little doubt that the fainting episodes were caused by the pedunculated hydatid body in the right atrium intermittently occluding the tricuspid valve as may happen with an atrial myxoma.

**Case 3**

A 33-year-old woman was operated on for a hydatid cyst 6 years previously. Two years later she began to be breathless on exertion and to have asthmatic attacks. She also had two attacks of urticaria. She had acrocyanosis, and there were 7 per cent eosinophils in the leucocyte count.

The x-ray film of the chest (Fig. 5) showed an enlarged heart with prominence of the pulmonary artery, and two small cysts in the left lung. The electrocardiogram (Fig. 6) showed right axis deviation and incomplete right bundle-branch block. It suggested right atrial enlargement and right ventricular hypertrophy with systolic overload. Gamma radiography of the lungs showed severe perfusion defects in the right upper and left lower zones.

![Image](http://heart.bmj.com/content/554/3/321/F1.large.jpg)

**Fig. 1** Case 1. Chest x-ray film showing rounded prominence of left cardiac border, and a diffuse round shadow in the right base.
Recurrent pulmonary embolism due to hydatid disease of heart

At cardiac catheterisation the pulmonary arterial pressure was 69/26 mmHg. Pulmonary angiography (Fig. 7) showed dilatation of the pulmonary trunk and its main branches. The right upper lobe branch was occluded 1 cm from its origin and the inferior lobe branches were tortuous and irregular. There was an extensive avascular zone in the left lower lobe.

The patient died suddenly. At necropsy there was a hydatid cyst in the interventricular septum with several vesicles protruding into the right ventricle (Fig. 8). A cyst had also perforated into the left

---

Fig. 2  Case 1. Opened hydatid cyst occupying upper part of the interventricular septum and anterior wall of right ventricle. Several vesicles (daughter cysts) and the hydatid membrane are visible.

Fig. 3  Case 2. Check x-ray film showing a rounded prominence of the right cardiac border. Round cystic shadows in both lung fields.

Fig. 4  Case 2. The opened heart showing a large cyst in the right atrium with a pedunculated portion passing through the tricuspid valve into the right ventricle.
ventricle. Both lungs showed multiple arterial emboli caused by hydatid vesicles and free scolices, with recent areas of infarction but no arterial thrombosis. There were in addition three small cysts in the left lung, and aortic, common iliac, renal, and thyroid hydatid cysts were also found.

The history together with x-ray film, electrocardiogram, catheterisation, and angiographic data, aided by the gamma scan, suggested hydatid disease of the heart with multiple pulmonary embolism. Massive embolism caused sudden death before operation could be undertaken.

**Discussion**

Echinococcosis of the heart is usually associated with pulmonary or hepatic hydatidosis. One of these cases however (Case 1) presented as a solitary cardiac hydatid cyst. The heart muscle is an unfavourable site for the development of a cyst as the
Recurrent pulmonary embolism due to hydatid disease of heart

contraction of the heart compresses it intermittently and shortens its life; it is of importance prognostically, as it renders it liable to rupture either into the cavity of the heart or into the pericardium.

The embryo reaches the heart muscle by way of the coronary circulation and not directly from the cardiac cavity. Usually the embryos from ingested ova cross the intestinal wall to reach the portal or azygos vein. Those that are not arrested in the liver reach the right atrium and right ventricle and pass by way of the pulmonary artery and pulmonary vein to the left side of the heart, whence they reach the coronary circulation. Occasionally a pulmonary cyst may rupture into a pulmonary vein liberating scolices into the blood flowing to the left ventricle, aorta, and coronary arteries. Once in the myocardium and subject to continuous 'trauma' the cyst may grow, or calcify and die. A developing cyst is liable to rupture into the cavities of the heart, producing pulmonary embolism if into the right ventricle and systemic embolism if into the left; or it may rupture into the pericardium producing a pericardial effusion (Gibson, 1964), or later constriction (Heyat et al., 1971). Usually there is an asymptomatic period of one to five years, or even longer, before these complications occur (Ivanissevich and Rivas, 1962).

The diagnosis of cardiac hydatidosis is based on the clinical and electrocardiographic findings, and especially the radiological appearances, particularly when associated with cysts elsewhere, eosinophilia, and a positive Casoni test. Nevertheless the majority of cases which have been described were unsuspected necropsy or surgical findings because attention is usually concentrated on hepatic or pulmonary cysts. If there is a bulge of the cardiac outline of non-homogeneous density which is not accounted for by valvular disease, cardiac hydatidosis should be considered, and a diagnostic cardiac catheterisation with angiography performed.

Purriel and his colleagues (1970) reviewed 39 published cases of hydatid pulmonary embolism and added 4 cases of their own. Further cases have been published since including 2 by Perez-Gomez et al. (1973). Most authors classify hydatid pulmonary embolism in 3 groups:

(a) acute fatal cases,

(b) subacute pulmonary hypertension with death in less than a year,

(c) chronic pulmonary hypertensive cases.

The majority of cases appear to follow a course of prolonged pulmonary hypertension punctuated by acute embolic episodes. In our 3 cases the clinical symptoms of pulmonary hypertension were present for 1 month, 1 year, and 4 years, respectively.

The prognosis of hydatid pulmonary embolism is grave, and only one case, that of Artucio et al. (1962), had a survival time measured in years before operation. Our case 1 appears to be the second to be cured by operation.

The angiographic, and particularly the surgical and necropsy findings, show that the pulmonary emboli are caused by vesicles or daughter cysts, and by scolices, and in none of our cases was there blood clot or added thrombosis. The smooth-walled daughter cysts appear to act purely mechanically by blocking the blood flow and not by irritating the vascular endothelium. In case 3 the lung scan showed perfusion defects affecting the right upper and left lower lobes which the patient tolerated surprisingly well until he died some days later of a massive embolism. It seems that hydatid embolism produces very little pulmonary vas-constriction and that the haemodynamic effect depends on the number and size of the occluded vessels. Only 2 cases, an exceptional one of Dévé (1946) and one of Perez-Gomez et al., were associated with a thrombus.
Purriel et al. (1970) and other authors have commented on the protrusion of hydatid cysts into the cardiac cavities with the possibility of intermittently blocking a valve orifice, and syncope has been reported in some cases. Our case 2 is noteworthy in this respect in that he presented with repeated syncope attacks and was found to have a pedunculated hydatid cyst protruding into the right atrium which we suspect was intermittently obstructing the tricuspid valve in the same way as is seen with a myxoma in this cavity.

References


Requests for reprints to Professor V. Gilsanz, Cea Bermudez 61, Madrid-3, Spain.
Recurrent pulmonary embolism due to hydatid disease of heart. Study of 3 cases, one with intermittent tricuspid valve obstruction (atrial pseudomyxoma).

V Gilsanz, C Campo, R Cue, J Estella, R V Estrada, C Perez-oteiza, G Rabago, J L Rebollar and P Zarco

*Br Heart J* 1977 39: 553-558
doi: 10.1136/hrt.39.5.553