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Hydatid disease of the heart

Report of five cases and review of the literature

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Calamai, G., Perna, A. M., and Venturini, A. (1974). *Thorax*, **29**, 451–458. **Hydatid disease of the heart: report of five cases and review of the literature.** The world literature on the surgical treatment of echinococcosis of the heart is reviewed. Few cases are surgically treated, although the disease has been known for a long time. Localization to the liver and lungs is the most frequent. Cardiopulmonary bypass techniques make possible surgical treatment of hydatid cyst of the heart. The present paper is concerned with five cases operated upon between 1959 and 1969, three males and two females, their ages ranging from 13 to 46 years. A preoperative diagnosis was made in each case. One case was operated upon under cardiopulmonary bypass. The need for cardiopulmonary bypass on a stand-by basis is emphasized. The localization of the hydatid cyst was in the left ventricular wall (three cases), right ventricular wall (one case), and multiple (one case). The frequency of cardiac echinococcosis ranges between 0·5% and 2% according to various authors. Diagnosis is achieved with the aid of laboratory tests, radiology, and angiography; but the presence of the disease must be suspected in all patients who come from endemic areas. Surgical therapy is mandatory. Due to the growth characteristics of the cyst itself, the danger of damaging the ventricular wall at operation is increased; thus it is essential to have cardiopulmonary bypass facilities immediately available.

Surgical treatment of a cardiac hydatid cyst was first attempted by Marten and de Crespigny (1921). The first case successfully operated upon was described by Long (1932). D'Abreu (1950) reported another case. Muller (1957) reported 26 surgically treated cases and 13 in which it was not clearly specified whether the treatment had been surgical. According to Al-Naaman and Al-Omeri (1970), the total number of surgically treated cases was 69, including three personal cases. In a recent review of the literature Heyat, Mokhtari, Hajaliloo, and Shakibi (1971) reported 118 cases of cardiac echinococcosis treated surgically. An additional case was described by Murphy, Kean, Venturini, and Lillehei (1971); another case was described by Hazan, Leblanc, Aobillard, and Mathey (1970), three cases by Urquia, Perez Leon, de los Arcos, and Madurga (1972), one case by Dodek, De Mots, Antonovic, and Hodam (1972), and three cases by Romanoff (1973). Between 1959 and 1969, five cases of hydatid cyst of the heart were successfully operated upon at the Institute of Surgery of Rome University. Our paper is concerned with the description of these five cases which bring to a total of 133 the number of such cases described in the world literature.

Hydatid cyst of the heart is an ominous disease which, in the absence of surgical treatment, is usually fatal (Peters, Dexter, and Weiss, 1945). An improvement in the prognosis of these patients follows surgery, especially since open-heart techniques allow radical treatment (Larghero, 1964; Di Bello and Menéndez, 1963; Dodek *et al.*, 1972; de los Arcos *et al.*, 1971).

CASE REPORTS

CASE 1 P.A., a 25-year-old man, was admitted on 16 February 1959 to the Surgical Department of Rome University. Four years before admission he had had an episode of crushing precordial pain, radiating to the neck and to the left shoulder and lasting only a few minutes. A few days later he presented with fever, shivering, cough, and bloodstained sputum; two

months later fainting and loss of consciousness occurred. For the next two years the patient was in no distress. In 1957 he presented with a further episode of precordial pain followed by fever and shivering. A chest radiograph disclosed cardiac enlargement.

Physical examination was negative on admission. The electrocardiogram showed normal sinus rhythm with left ventricular strain. A chest tomogram and diagnostic pneumothorax showed a lesion thought to be an hydatid cyst of the pericardium. Blood tests were normal, and the Casoni intradermal skin test was positive. The patient was explored on 25 March 1959. Through a left thoracotomy in the fourth intercostal space, a hydatid cyst was found covered by the pleura which was adherent to the pericardium. The pericardium was opened and it was found to be incorporated in the wall of the cyst and adherent to the myocardium. The cyst was then aspirated and irrigated with iodine solution. The capsule was partially resected, leaving a portion adhering to the underlying myocardium.

CASE 2 C.G., a 13-year-old boy, was admitted on 10 June 1968. Two years before admission he had presented with an episode of fever (40°C), widespread oedema, itching, dyspnoea, precordial pain, and cough. Chest films showed an enlarged cardiac shadow with a number of small masses along the left and right borders (Fig. 1a and b); on the lateral projection a bilobed mass was seen anteriorly. A loud split second sound over the pulmonary area, and a third heart sound were audible over a wide area.

Blood sugar, haemoglobin, and haematocrit values were within normal limits. A Casoni intradermal skin test was positive and a Weinberg complement fixation test was positive. A full blood count was normal except for 13% eosinophils in the peripheral blood smear.

The electrocardiogram showed normal sinus rhythm and inverted T waves in leads II, III, aVF and the left precordial leads; Q waves were present in leads III and aVF.

The preoperative diagnosis was echinococcosis of the pericardium.

On 19 July 1968 an exploratory operation was performed through an anterior bilateral thoracotomy in the fourth intercostal space with transverse sternal transection. The pericardium was completely adherent to the heart. Two cysts, 3 cm in diameter, were removed from the anterior surface of the right ventricle. Smaller cysts were isolated and removed from the right atrium. Four infected cysts were removed from the mediastinal aspect of the heart following irrigation with iodine into the cyst cavities. The cysts were adherent to the surface of the left ventricular wall. Other cysts were removed (following irrigation) from the left atrium. The left phrenic nerve, involved by the hydatid disease, was transected. At the conclusion of surgery the heart showed normal contractability. The patient died six months after operation from chronic heart failure due to extensive disease of the myocardium.

CASE 3 C.G., a 45-year-old man, was admitted in November 1968. His principal complaint was of precordial pain for two months. A midsystolic murmur in the second intercostal space at the right sternal edge was found. An early diastolic murmur was audible along the left sternal edge. The electrocardiogram showed normal sinus rhythm and evidence of previous anterolateral myocardial infarction. Chest films showed a round shadow with marginal calcification projecting on the left ventricle (Fig. 2a and b). There were 5% eosinophils in the peripheral blood smear; all other blood tests were within normal limits.

On 22 May 1969 an exploratory thoracotomy was performed through the fourth left intercostal space. The pericardium was found to be adherent to a bulging mass on the left ventricle. Aspiration of the mass produced fresh blood. The patient was placed on total cardiopulmonary bypass. The mass was then excised. The left ventricle was widely opened, the papillary muscles and mitral valve being left intact. The ventriculotomy was then closed with multiple mattress sutures buttressed with strips of Teflon felt. The patient regained a normal cardiac output as soon as cardiopulmonary bypass was discontinued.

CASE 4 B.G.D., a 9-year-old boy, was admitted on 10 June 1969 complaining of intermittent precordial pain of short duration. In April 1969 a chest radiograph had shown a round shadow along the left heart border. A loud and split pulmonary sound and a midsystolic murmur in the second and third intercostal space along the left sternal edge were audible. The electrocardiogram showed normal sinus rhythm and signs of anterolateral subepicardial myocardial ischaemia.

Blood tests were within normal limits.

The patient was operated upon on 16 July 1969. A hydatid cyst was found adherent to the left ventricular apex. The pericardium was packed with iodized gauze and the cyst was aspirated. Ten millilitres of transparent fiuid was obtained. The cyst was then opened and the cyst membrane was enucleated intact from the underlying myocardium. The left ventricular wall was approximated by interrupted silk sutures.

CASE 5 G.A., a 43-year-old woman, was admitted in July 1969. Her principal complaint was intermittent crushing precordial pain, radiating to the left shoulder, of approximately six years' duration. A mass in the left lower pulmonary field was first noted in November 1968 on routine pre-employment radiographs. A diagnosis of echinococcosis of the heart was made following electrocardiogram, chest radiographs (Fig. 3a and b), a positive Weinberg complement fixation test, and a positive Casoni intradermal

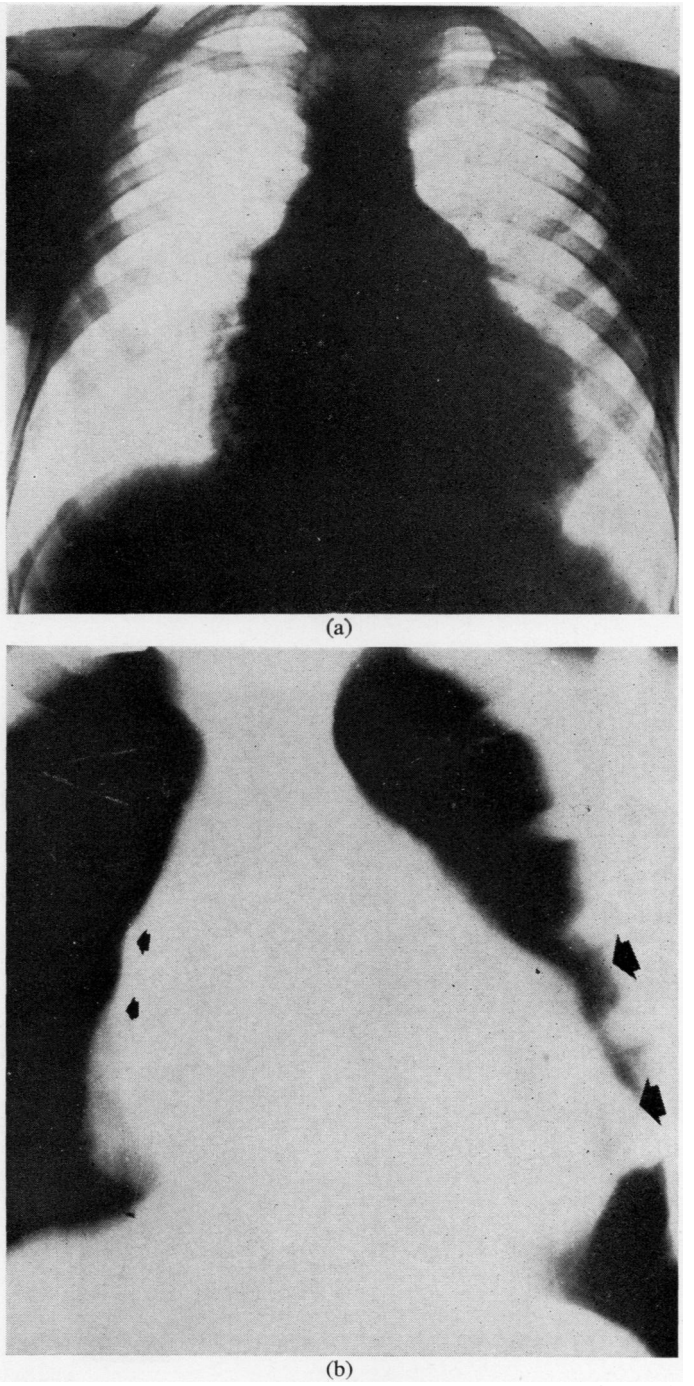
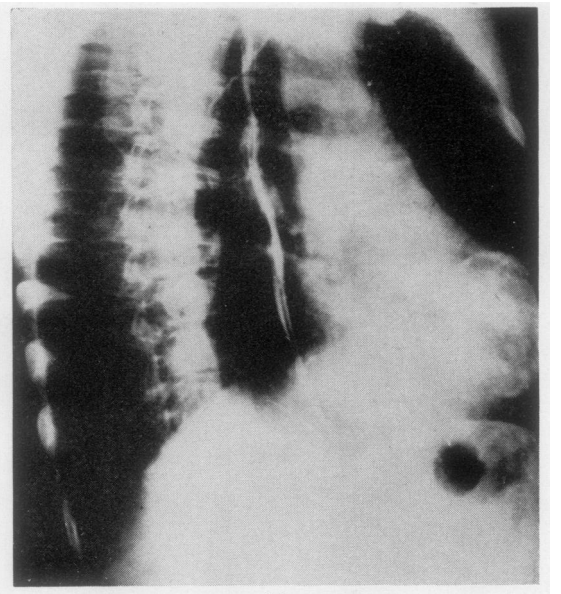
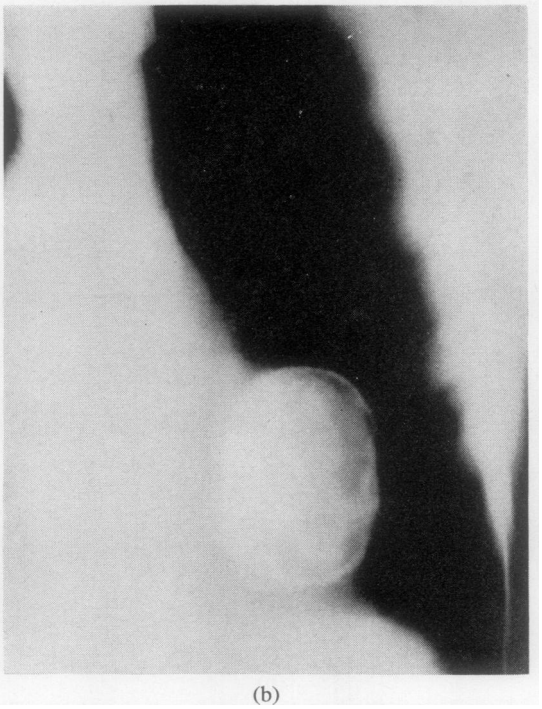


FIG. 1. *Case 2. (a) Preoperative posteroanterior chest radiograph showing an enlarged cardiac shadow with multiple small masses on both sides of the heart. (b) Anteroposterior tomogram shows masses on the left and right borders of the cardiac shadow.*



(a)



(b)

FIG. 2. *Case* 3. *(a) Oblique radiograph showing a round shadow with marginal calcification. (b) Antero-posterior tomogram shows an oval calcified cyst.*

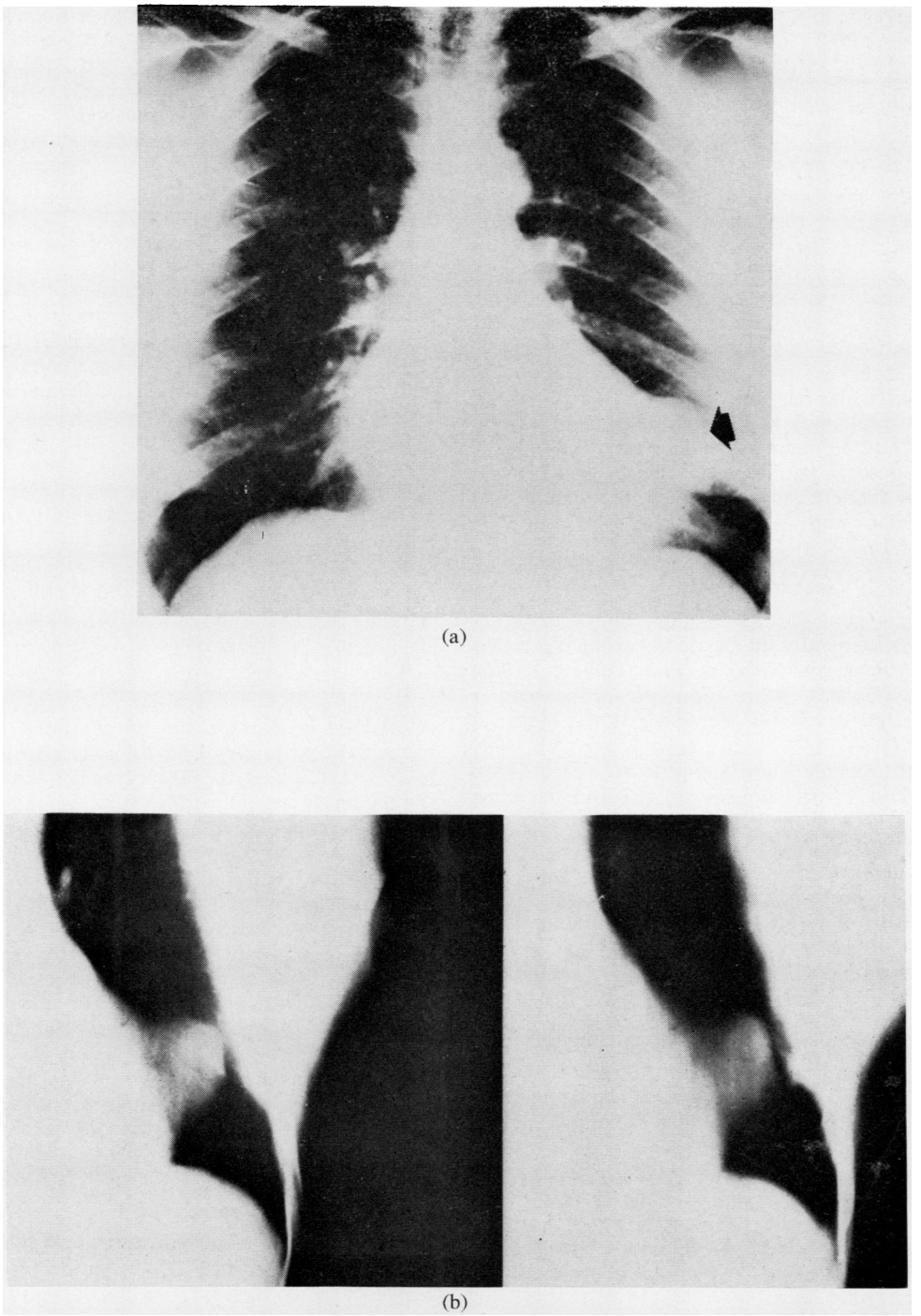


FIG. 3. *Case 5. (a) Preoperative posteroanterior chest film showing a round shadow on the left cardiac border. (b) Tomograms showing the mass protruding from the left ventricular wall.*

skin reaction. The peripheral blood smear revealed 4% eosinophils.

The patient was operated upon and the mass was excised through a left thoracotomy according to the previously described techniques. She made an uneventful recovery.

DISTRIBUTION OF HYDATID DISEASE OF THE HEART

Once the hexacanth embryo has embolized to a coronary artery the following distribution within the heart may be found. Ivanissevich and Rivas (1962), in a series of 194 cases of hydatid disease of the heart, reported the following distribution: in 116 (60%) cases the disease occurred in the left ventricular myocardium, and in 33 (17%) in the right ventricle; in 18 (9%) cases in the interventricular septum; in 16 (8%) cases in the right atrium; in 8 (4%) cases in the left atrium; and in 3 (2%) cases in the interatrial septum.

In our series of five cases the distribution was three in the left ventricular wall, one in the right ventricular wall, and one case with multiple localization.

SYMPTOMS AND SIGNS

Hydatid cyst of the myocardium can be asymptomatic; occasionally the first clinical manifestation is sudden death (Guarini and Torraco, 1962; Comakov, 1965). More often precordial pain is present (Di Bello and Menéndez, 1963; Heyat *et al.*, 1971).

Other possible signs are due to compression by the cyst. Mitral stenosis can be simulated when the cyst lies in the left atrium obstructing the cardiac outflow.

Angina is a main complaint if the cyst compresses a coronary artery.

Electrocardiographic changes simulating myocardial infarction can be caused by the hydatid cyst (Vestri and Tardio, 1968) or conduction defects due to compression of the bundle of His (Heimann, 1928; Corkill, 1929; Ghanem and Darwish, 1951; de los Arcos *et al.*, 1971).

When the cyst is located in the right heart the most common features are hepatomegaly, oliguria, ascites, or chronic cor pulmonale due to repeated pulmonary emboli (Aguirre *et al.*, 1956; Al-Naaman and Al-Omeri, 1970).

From rupture of a cyst into the cardiac cavity a variety of allergic phenomena result, ranging from wheals to anaphylactic shock. Acute pericarditis may be caused by rupture of the cyst into the pericardial sac.

DIAGNOSIS

In spite of many symptoms the diagnosis during life may be difficult. If a patient comes from an endemic area the possibility of hydatid disease should be kept in mind. A chest radiograph is often the first clue to the diagnosis, showing an abnormality of the cardiac outline.

Fluoroscopy adds information about motility or absence of normal pulsation, suggesting the possibility of a hydatid cyst.

Occasionally the presence of calcification is an aid to diagnosis (Blondeau, Lauprètre, and Miramond de la Roquette, 1938).

Angiography must be considered the most specific of all diagnostic procedures if a filling defect in the cardiac cavity is revealed.

The electrocardiogram shows a thinner cardiac wall in the area of localization of the parasite and low-voltage R waves, QRS notching, and T-wave inversion and increase of R waves in adjacent areas, or absence of anomalous Q waves.

When the cyst is in the interventricular septum arrhythmia may be present, and the QRS appears notched.

The laboratory tests are of little help: the Casoni intradermal skin test and Weinberg complement fixation test are often negative and eosinophilia is not a constant finding.

DIFFERENTIAL DIAGNOSIS

A hydatid cyst must be differentiated from a bronchial carcinoma originating close to the lung hilum, tumours of the heart, benign tumours of the mediastinum and pericardium, and aneurysms of the heart.

Ventricular aneurysms are the most common and can be easily differentiated by means of angiography.

According to Murphy *et al.* (1971), the consequences of cardiac hydatid cyst are: death of the cyst with calcification; rupture, either intrapericardial, causing pericarditis, or intracardiac, leading to anaphylactic shock or embolic phenomena.

Cor pulmonale or metastatic pulmonary echinococcosis, systemic emboli or systemic metastasis of the hydatid cyst can be caused by rupture of the cyst into the right or left heart.

TREATMENT

There is no known medical treatment of cardiac echinococcosis. In view of the serious complications of this disease surgical treatment is mandatory.

The chest should be entered through a left, right or bilateral thoracotomy, depending on the

site of the cyst. Once the cyst has been freed from the surrounding structures its content should be aspirated and then irrigated with hypertonic (33%) saline in order to achieve sterilization of the parasite wall and viable elements (Vaglio and Guarini, 1965). When the cyst is located inside the wall of the left ventricle, removal may be difficult because of the thinness of the surrounding myocardium (Guarini and Torraco, 1962). In our opinion, bypass facilities should be available every time the ventricular walls are involved by a hydatid cyst. Once the cyst has been removed the area should again be irrigated with hypertonic saline.

The mortality of this procedure is low and the postoperative course is usually uneventful; the patient soon resumes normal life.

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