Fifteen year surveillance of echinococcal heart disease from a referral hospital in Greece

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Ten cases of hydatid heart disease were treated over a 15-year period (1980–1995). Cysts were located in the left ventricular wall (four patients), right ventricular wall (one patient), interventricular septum (one patient), interatrial septum (one patient), right atrium (one patient), pericardial cavity (one patient) and in multiple loci (one patient). Apart from two asymptomatic cases, clinical manifestations included chest pain (four patients), anaphylactic shock (one patient), constrictive pericarditis (one patient), congestive heart failure (one patient) and arterial embolism (one patient). Computed tomography was found useful in the detection of hydatid cysts and also in the determination of their morphology. Magnetic resonance was performed in three patients, with satisfactory imaging. Three out of the 10 patients died: rupture of pulmonary echinococcal cyst (one patient), massive pulmonary hydatid embolism (one patient) and rupture of an undiagnosed hydatid cyst of the right atrium during cannulation for cardiopulmonary bypass (one patient). One other patient experienced recurrent systemic embolism and became hemiplegic. Six patients were successfully treated. In five patients, the cysts were excised by open heart surgery, while in one by pericardectomy. In addition, antiparasitic drugs were successfully used in two patients with long-term satisfactory results.

In conclusion, cardiac echinococcosis is associated with an increased risk of potentially lethal complications. Newer techniques of cardiac imaging have helped locate the cysts while surgical removal may offer cure. Some patients responded to specific long-term drug treatment.

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Key Words: Echinococcosis, echocardiography, computed tomography.

Introduction

Echinococcus infestation is known to be endemic in some parts of the world[1,2]. This parasitic disease, from the time of ingestion of the ova, or larvae by man, until clinical manifestations begin, may evolve insidiously over several decades. In some cases, it may lead to conditions which constitute a medical emergency, again without any previous history of the disease, or any symptoms. The endemic nature of the disease is due largely to poor hygienic measures in these areas. Man is infected from contaminated dogs (intermediate host), which have been infected by eating sheep’s liver containing hydatid cysts. Another source of contamination is the abundant consumption of raw vegetables growing in naturally fertilized (contaminated) soil. After ingestion by man the worms and/or ova of the parasite reach the liver, evolve to hexacanthic embryos and enter the right circulation through either the venous or lymphatic route. Some pass the lung barrier and enter the systemic circulation. The involvement of the heart itself is reported to range between 0-5 and 2% of cases of echinococcosis[3]. Today, the incidence and prevalence of hydatid cyst disease in Greece, although not accurately known because it is not reportable, are estimated to be high. The disease will hopefully be completely eradicated with improved hygiene measures, which are coming into force. Nevertheless, Greek hospitals, as well as those of other European countries, may admit patients with echinococcosis presenting as difficult cardiological diagnostic or therapeutic problems.

We report all the clinical, laboratory, diagnostic and therapeutic data of 10 cases of echinococcal infestation involving the circulatory system. It is a retrospective study extending back about 15 years in the hospital’s records and hence not every case will have been studied on the basis of contemporary knowledge, equipment, and diagnostic and therapeutic modalities.
Cocciosis. It consists in injecting intradermally a minute quantity of hydatid cyst fluid, which as an antigen produces redness of the skin where it is injected. Despite the simplicity, however, in performing the test, there are many disadvantages. Its major limitation is that the test is not specific. False-positive reactions are very common in sufferers from other parasitic and even non-parasitic diseases.

For the above reasons, several tests which aim at searching for titres of anti-echinococcal antibodies in the serum of these patients have been developed. These include: compliment fixation (CF), indirect haemagglutination (IHA), indirect immunofluorescence (IFA), latex agglutination, immunoelectrophoresis, ELISA, etc. The sensitivity and specificity of these tests vary and it is suggested that in suspected cases, more than one test should be performed. It is reported, however, by some that these tests may be used in patients following surgical, or pharmacological treatment. The positivity of the tests is influenced mainly by the location and the condition of the cyst. Pulmonary cysts very frequently give false-negative results. Also, the immuno-antigenic titres are low when the cysts contain clear liquid, or when they are calcified. However, they are very high if

Materials and methods

This report concerns a retrospective clinico-laboratory study of 10 patients (six male and four female), with a mean age of 46.5 years (range 22–72), who were admitted to our hospital between 1980 and 1995. Depending on the year of admission, some of the patients presented here lack the more detailed imaging investigations available nowadays. All the patients were handled in a standard and classical way by the specific department to which they were admitted. Because most of them constituted difficult diagnostic problems, all the available diagnostic means and modalities were freely utilized: imaging techniques, biochemical, haematological, skin and serological tests, as well as, interventional diagnostic methods (see Table 1). A short description of the specific tests follows.

Table 1 Substantial data of the ten patients studied

<table>
<thead>
<tr>
<th>Serial no.</th>
<th>Sex</th>
<th>Age (years)</th>
<th>Year admitted</th>
<th>Specific tests</th>
<th>Clinical presentation</th>
<th>Location of hydatid cyst</th>
<th>Cardiac catheterization</th>
<th>Survival</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>M</td>
<td>50</td>
<td>1980</td>
<td>Not available</td>
<td>Asymptomatic</td>
<td>Right atrial myocardium</td>
<td>Not performed</td>
<td>Died of 'vomique'</td>
</tr>
<tr>
<td>2</td>
<td>F</td>
<td>72</td>
<td>1983</td>
<td>Casoni (+)</td>
<td>Asymptomatic</td>
<td>Left ventricular myocardium</td>
<td>Filling defect at the apex of the left ventricle</td>
<td>Died of massive pulmonary embolism during cannulation for TCB</td>
</tr>
<tr>
<td>3</td>
<td>M</td>
<td>22</td>
<td>1984</td>
<td>Casoni (+)</td>
<td>Anaphylactic episodes</td>
<td>Interatrial septum</td>
<td>Not performed</td>
<td>Died of massive pulmonary embolism (necropsy finding)</td>
</tr>
<tr>
<td>4</td>
<td>M</td>
<td>30</td>
<td>1986</td>
<td>Casoni (+)</td>
<td>Chest pain</td>
<td>Left ventricular myocardium</td>
<td>Filling defect at the base of the left ventricle</td>
<td>Surviving and well till 31.6.1995</td>
</tr>
<tr>
<td>5</td>
<td>M</td>
<td>52</td>
<td>1986</td>
<td>Casoni (+)</td>
<td>Chest pain</td>
<td>Left ventricular myocardium</td>
<td>Obstructive compression of circumflex artery</td>
<td>Surviving and well till 31.6.1995</td>
</tr>
<tr>
<td>6</td>
<td>F</td>
<td>31</td>
<td>1987</td>
<td>IHA titre 102 000</td>
<td>Multiple arterial emboli</td>
<td>Left ventricular myocardium</td>
<td>Ruptured hydatid cyst of the left ventricle</td>
<td>Surviving till 31.6.1995 (hemiplegic)</td>
</tr>
<tr>
<td>7</td>
<td>F</td>
<td>45</td>
<td>1991</td>
<td>Casoni (+)</td>
<td>Chest pain, dyspnoea</td>
<td>Multi foci in the myocardium</td>
<td>Not performed</td>
<td>Surviving and well till 31.6.1995</td>
</tr>
<tr>
<td>8</td>
<td>M</td>
<td>36</td>
<td>1993</td>
<td>IHA titre 320</td>
<td>Chest pain</td>
<td>Right ventricular myocardium</td>
<td>Filling defect at the apex of right ventricle</td>
<td>Surviving and well till 31.6.1995</td>
</tr>
<tr>
<td>9</td>
<td>F</td>
<td>63</td>
<td>1994</td>
<td>ELISA &gt;5000</td>
<td>Chest pain, congestive heart failure</td>
<td>Constrictive pericarditis and anaphylactic episodes</td>
<td>Pericardium</td>
<td>Equalization of diastolic pressures in all cardiac chambers</td>
</tr>
<tr>
<td>10</td>
<td>M</td>
<td>64</td>
<td>1994</td>
<td>ELISA &gt;5000</td>
<td>Chest pain</td>
<td>Constrictive pericarditis and anaphylactic episodes</td>
<td>Not performed</td>
<td></td>
</tr>
</tbody>
</table>

*IHA = indirect haemagglutination assay; †TCB = total cardiopulmonary bypass.

Casoni's test

This is one of the oldest diagnostic tests for echinococcosis. It consists in injecting intradermally a minute quantity of hydatid cyst fluid, which as an antigen produces redness of the skin where it is injected. Despite the simplicity, however, in performing the test, there are many disadvantages. Its major limitation is that the test is not specific. False-positive reactions are very common in sufferers from other parasitic and even non-parasitic diseases.

For the above reasons, several tests which aim at searching for titres of anti-echinococcal antibodies in the serum of these patients have been developed. These include: compliment fixation (CF), indirect haemagglutination (IHA), indirect immunofluorescence (IFA), latex agglutination, immunoelectrophoresis, ELISA, etc. The sensitivity and specificity of these tests vary and it is suggested that in suspected cases, more than one test should be performed. It is reported, however, by some that these tests may be used in patients following surgical, or pharmacological treatment. The positivity of the tests is influenced mainly by the location and the condition of the cyst. Pulmonary cysts very frequently give false-negative results. Also, the immuno-antigenic titres are low when the cysts contain clear liquid, or when they are calcified. However, they are very high if
Discussion

Echinococcosis is a human parasitic disease, caused by the larval stage of *Echinococcus granulosus*. Echinococcus cysts were known to Hippocrates, who mentioned the serious manifestations and consequences of rupture of hydatid cysts of the liver. Human infection with echinococcus is not so rare, when one takes into consideration that, in certain areas, this parasitic disease exists in endemic form. The disease is common in the sheep-raising countries primarily Uruguay, Australia, New Zealand, Greece, the Middle East, North Africa and the Balkans. In some of these places, those affected may be carriers of the parasite in a 'quiescent clinical form' for several years, even decades. This is understandable, since the cysts of the parasite, even if they reach a significant size (5–10 cm diameter), and if they do not exert pressure on organs and structures, or do not rupture, may be present without any symptoms at all. The larvae survive within the cyst for 4–5 years. About this time, their capsule becomes calcified, the larval die and they may be found incidentally during life, or at post-mortem. The usual foci where cysts may develop are the parenchymal organs (liver, kidney, spleen) as well as the lungs, the omentum, the peritoneal cavity etc. Of course, if a cyst ruptures, or even if it leaks, the most common symptoms are anaphylactic reactions, with chills and fever, skin exanthem, bronchospasm and dyspnoea, and sometimes circulatory collapse with death.

A great variety of symptoms, depending mainly on the location of the parasitic cyst, have been described. It is understandable that, if the cyst ruptures suddenly, the patient may present as a medical emergency. Besides all the anaphylactic symptoms, which are the consequence of the foreign protein molecules entering into the circulation of the patient, severe and fatal circulatory collapse may occur. In the first three patients of this series the cysts ruptured and insufficient time was given for therapeutic interventions. The first patient died of massive pulmonary embolism during cannulation for cardiopulmonary bypass for replacement of the mitral valve. The second patient developed 'vomique'. This French word is derived from the word vomitus and describes the situation where a pulmonary cyst is ruptured within the bronchial tree and a torrent of liquid containing parts of the echinococcus is expelled like vomitus. The third patient died suddenly after becoming cyanotic and dyspnoeic and was found at autopsy to having suffered massive pulmonary embolism from hydatid cyst remnants. It is clear that rupture of the cyst is a real medical emergency, frequently killing the patient.

The involvement of the pericardium may be manifested by: (1) silent rupture and the appearance of the echinococcus cyst some months later; (2) acute pericarditis with or without cardiac tamponade; (3) constrictive pericarditis.

Disturbances in rhythm are attributed to interference with the conduction system. Murmurs
may be caused by valve insufficiency, when the echino-
coccus invades the papillary muscles or by partial
obstruction of the right ventricular outflow tract, by an
intruding cyst. Clinical pictures mimicking subaortic
stenosis, pulmonary stenosis, mitral stenosis, and
involvement of the tricuspid valve, have all
been described. Sometimes the first manifestations are
destructive and may even be fatal.

Biochemical and haematological investigations
may not reveal anything else except an abnormal dif-
ferential white blood count with an impressive increase of
the eosinophilic series of the white cells. In this series, all
the patients exhibited an increased eosinophilic count as
an indication of the anaphylactic reaction to the parasite.

Whenever a case is met, which does not fit into
the symptomatology and the general manifestations of a
known disease, particularly if the patient originates from
a geographical area where echinococcosis is common, the
physician has to think about the possibility of echino-
coccal disease. If, within the frame of the initial routine
blood tests, eosinophilia above 6–7% is noted, then the
physician should proceed with specific tests. We are
fortunate today to live in the era of several diagnostic
imaging techniques. Even if one performs 'blindly'
ecocardiograms, computed tomography (CT) scans,
Magnetic Resonance Imaging (MRI) scans, or if one
applies various radionuclide imaging techniques it is
quite probable that one might come across findings
which will illuminate the diagnosis. However, these
techniques should not encourage the clinician to omit, or
pay scant attention to the history. On the contrary, the
very careful and detailed recording of the patient's
symptoms frequently render some very important and
essential information which may lead to the correct
diagnosis. Only when the patient is asymptomatic, or the
symptoms are very mild and not specific, or they vary in
different time periods, may the physician decide to use
'blindly' the above mentioned diagnostic modalities, but
again, with moderation and scientific reasoning.

The myocardium is affected mainly via the cor-

nary and arterial circulation. A few weeks or months
after the implantation of the hexacanthic embryo, the
hydatid cyst develops. The latter is encapsulated by a
fibrous structure, which is often becoming calcified.
Within 1–5 years the cyst may degenerate (die), unless it
ruptures, with all the consequences.

In this series, the hydatid cyst was found in the
left ventricle (four times) or other myocardial site (four
times) and in the pericardial sac (once). Cysts in multiple
loci were encountered in one case. The locations of the
cysts indicate the possibility of the disease appearing
with a great variety of symptoms.

All five patients of this series who complained
of chest pain gave a rather vague and undetermined
description of their symptoms. Even in the patient in
whom the coronary arteriogram demonstrated obstruc-
tive compression of the circumflex artery (case no. 5),
the pain did not resemble that of angina. According to
Murphy et al., the precordial pains are usually

the result of the partial rupture of the cyst into the
pericardial sac, while Rivera and Delcan attribute
the chest pain to coronary insufficiency, as a result of
compression of coronary arteries. As a matter of fact,
they suggest angiographic examination of all cardiac
echinococcosis suspects. It is advisable to submit every
case of cardiac echinococcosis to coronary arterio-


ography, particularly if they are above the age of 40, and
possible candidates for open heart surgery.

Routine haematological examinations may re-

veal some degree of eosinophilia, which was a stable and
common finding in this series, and impressively high in
case nos 2 and 3 (38 and 27%, respectively).

Certain changes in the electrocardiogram, although neither the electrocardiogram, nor
the chest film contributed any specific findings in the
cases reported here. While the chest film was entirely
negative, quite often the electrocardiogram would
exhibit non-specific (or pathognomonic) changes.

The contribution of echocardiography, particu-
larly in two dimensions, has to be underlined.
In this series, echocardiography proved more sensitive and
more specific than computed tomography. However, the
sensitivity of the method is reduced in relation to:
(1) the size and number of cysts (difficulty in the location
of cysts less than 0.5 cm in diameter); (2) the contents of
the cyst, when these have degenerated and include
remains of membranes and necrotic material (case no.
5); and (3) in distinguishing between a single and a
multilocular cyst.

Computed tomography, is a sensitive modality in
the diagnosis, but lags behind the echocardiogram
in the precise location of a hydatid cyst. In case nos 2
and 3 the echocardiogram accurately determined the
location of the cardiac cysts, but the computed tomog-


raphy scan did not. Also, in case no. 9 in whom the right
pulmonary artery was occluded, the computed tomog-
raphy scan could not locate the cyst, while with the
magnetic resonance imaging scan its intraluminal
location was ascertained. These peculiarities are illus-
strated in Figs 1 and 2.

We conclude from these last observations that a
history of non-specific symptoms (mainly atypical chest
pain), combined with a lack of physical findings and
eosinophilia, should raise the suspicion of echino-
coccal involvement of the heart, and lead to meticulous
and careful echocardiographic investigation. If, from
such an approach, enough information is gained indica-
tive of such involvement, then (and only then) should
one proceed to further studies such as magnetic resonance imaging
and cardiac catheterization and angiography, aiming at more detailed information,
which will contribute to a more accurate and precise
diagnosis and better planning of surgical treatment for
the condition.

Concerning treatment of cardiac echinococcosis,

it is evident that this must always be immediate and
aggressive, in order to prevent morbid complications
from ruptured cysts.

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Figure 1 Echocardiogram (a), computerized axial tomo-
gram (b) and magnetic resonance imaging (c) of a patient
(case no. 9), showing a cyst of 3 cm diameter in the
interventricular septum.

In this series, the outcome of all four patients
who were not operated on, was very poor (three died,
one became hemiplegic); note that patient no. 1 died of
massive pulmonary embolism from the rupture of an
undiagnosed cyst of the right atrium during cannulation
for total cardiopulmonary bypass for mitral valve
replacement.

Treatment with albendazole (Zentel 400 mg)
should be applied in patients with low life expectancy,
such as those with multiple cerebral lesions, or cases
with frequent relapses. The result of albendazole treat-
ment in our patients (case no. 6 with a cerebral echino-
coccus cyst and case no. 8 with multiple small cysts in
both lung fields) was spectacular and the cysts dis-
appeared after treatment. However, in patient no. 8,
there was a relapse 3 years later and the drug had to be
administered again. The usual way this drug is given is
as follows: Zentel 800 mg daily, in two equal divided
doses during meals, for 28 days. This 28-day treatment
can be repeated after an interval of 14 days, for a total of
three cycles.

In conclusion, to diagnose hydatid disease of the
heart, a high degree of suspicion is necessary. However,
if the patient comes from an area where the disease is
endemic, if the symptomatology is not quite 'typical',
and most importantly, if significant eosinophilia is
present, the patient should be submitted to a most
careful and meticulous investigation with all the avail-
able diagnostic modalities, to rule out the presence of
hydatid disease infestation of the circulatory system. If
the disease is diagnosed with certainty, its surgical
management may be curative and life saving.

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