Echinococcal Cyst of the Interventricular Septum with Right Ventricular Protrusion

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Although echinococcosis (echinococcal hydatidosis) is common in sheep-raising countries such as Turkey, cardiac involvement is rare; the presence of a hydatid cyst in the interventricular septum is rarer still. We report a case of hydatid cyst of the interventricular septum that was first revealed by 2-dimensional echocardiography and then confirmed by right ventricular angiography. The cyst was removed surgically under cardiopulmonary bypass.

Within the context of the medical literature concerning this rare lesion, we discuss this case and 10 other cases of cardiac hydatidosis, previously unreported in the world literature, that we have treated from January 1967 through January 1987. ***(Texas Heart Institute Journal 1989;16:292-5)***

According to Ottino and associates,[1] cardiac involvement in echinococcosis is found in from 0.5% to 3% of all cases. In about 9% of these cases, the cyst is located in the interventricular septum. The first removal of a septal cyst was performed by Artucio and associates[2] in 1962.

In a 1977 update[3] of an earlier review[4] of the world literature, Shakibi and associates added 60 cases of surgical treatment of cardiac hydatidosis to the 118 they had reported in 1971. To those 178 cases and others reported in the world literature since the last Shakibi review, we add 11 previously unreported* cases of cardiac echinococcosis that we have treated surgically within a period of approximately 20 years. One of these involved the successful removal of a septal echinococcal cyst detected by means of 2-dimensional echocardiography.

Case Report

In January of 1987, an 18-year-old man was admitted to our hospital for evaluation of palpitations and shortness of breath upon exertion. Physical examination revealed a grade 3/6 systolic ejection murmur. The patient's heart rate was 60 beats/min, and his blood pressure was 120/80 mmHg. Electrocardiography indicated a normal sinus rhythm and a right bundle-branch block. Chest radiography revealed no abnormalities. Two-dimensional echocardiography showed a round, cyst-like structure in the interventricular septum that bulged toward the pulmonary infundibulum (Fig. 1). Cardiac catheterization and angiography confirmed the presence of a cyst in the interventricular septum, protruding into the right ventricle (Fig. 2).

The patient was operated on with the aid of cardiopulmonary bypass. After cardiac arrest had been instituted with cold potassium cardioplegic solution, a transverse right ventriculotomy was performed and the echinococcal cyst was found in the muscular septum. The white, ovoid lesion measured 4 x 6 cm, and its surface was partly covered by ventricular septal myocardium. We aspirated the cyst before sterilizing its contents by instilling absolute alcohol. When the cystic cavity was entered, several daughter cysts (Figs. 3 and 4) of various sizes were encountered. These we removed individually, before extirpating the contents of the large ectocyst. Septal continuity was kept intact throughout, and the edges of the ectocyst were excluded with multiple 3-0 Ethibond purse-string sutures.

*Some of these cases have been reported locally in Istanbul, but not in the world literature.

Key words: *Cysts, hydatid; echinococcosis*

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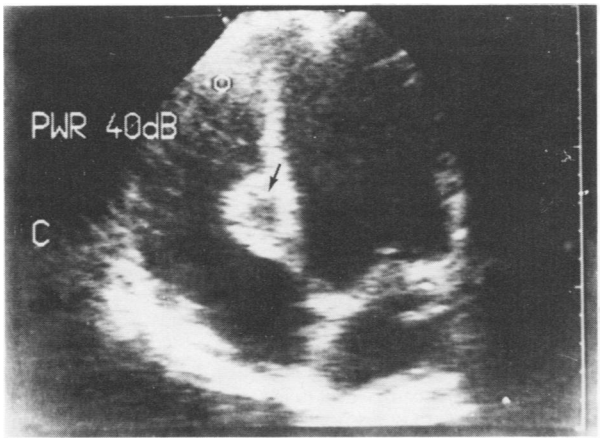

Fig. 1 *Two-dimensional echocardiography, not definitive for a diagnosis of cardiac hydatidosis, nevertheless revealed a round, cyst-like structure (dark spot indicated by arrow) in the interventricular septum that bulged toward the pulmonary infundibulum.*

The patient recovered uneventfully. A year later, he was doing well, and the evidence of right bundle-branch block had disappeared from his electrocardiogram.

Discussion

The definitive host of *Echinococcus granulosus* is the dog, and most often the sheep is the intermediate host. Man is a common accidental host, and echinococcosis is endemic to many sheep-raising areas of the world—particularly Argentina, Uruguay, New Zealand, North Africa, Turkey, and the countries of the Balkan peninsula.

During a period of approximately 20 years (January 1967 through January 1987), 2000 cases of pulmonary echinococcosis were treated at our thoracic and cardiovascular surgery center in Istanbul. In contrast, we treated only 11 cases of cardiac echinococcosis during this same period (Table I): in 5 of these cardiac cases, we observed a large number of cysts within the pericardial sac; 5 cases involved the right ventricle, 1 involved the left ventricle, and 1 involved the interventricular septum. Although, in other series, the left ventricle has been the most common site of cardiac involvement, partly because of its rich coronary circulation,[5] our cases more often involved the right ventricle.

The clinical picture associated with cardiac echinococcosis is nonspecific, because the condition can mimic various cardiac diseases, even in instances of septal localization. The differential diagnosis includes all other cardiac tumors and cysts, as well as mediastinal tumor, pericardial cyst, and ventricular aneurysm. Of these, the most common finding is ventricular aneurysm, a condition easily differentiated by angiography.[6,7] Often, the first symptoms are caused by rupture of the cyst and pulmonary embolization. In 269 postmortem examinations, Di Bello and Menendez[8] observed that 10% of the myocardial cysts had burst toward the pericardial sac and 38% had ruptured toward the intracardiac cavities. Rupture into the heart chambers occurs most frequently with cysts located in the right ventricular wall, probably due to the relative thinness of this structure.

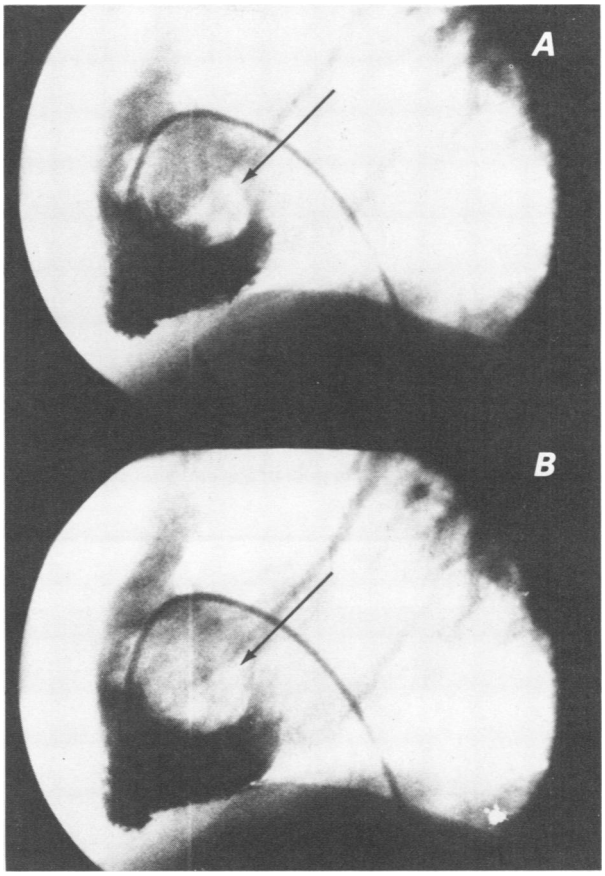

Fig. 2 *Cardiac angiograms confirmed the presence of a cyst in the interventricular septum, protruding into the right ventricle. Arrows indicate the cyst as it appeared during systole (**A**) and diastole (**B**).*

Even if cardiac hydatidosis is not suspected upon presentation, a routine echocardiographic investigation can lead to a correct diagnosis. Indeed, a 2-dimensional echocardiographic result indicative of hydatid cyst may aid in the early detection of cardiac involvement and is considered the best noninvasive diagnostic method when it is confirmed by computed tomographic scanning.[1,9] A definitive preoperative diagnosis is provided only by cardiac catheterization and angiography, but some investigators[1] feel this invasive procedure is unnecessary and yields little additional information.

In instances of septal lesion, cardiac rhythm is often disturbed because the conduction system has

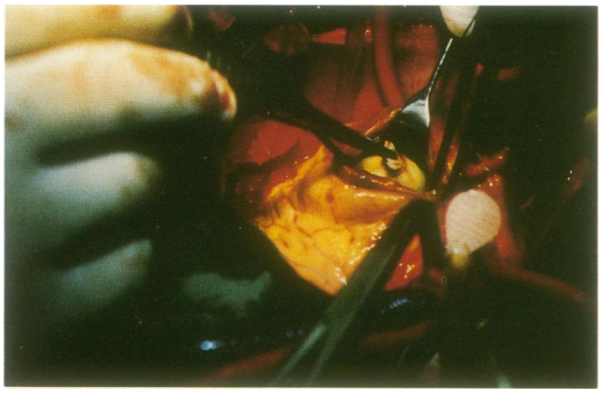

Fig. 3 *The top of the ectocyst is seen through the ventriculotomy, as the 2nd daughter cyst is prepared for removal. To the right is the 1st and largest of the daughter cysts, immediately after its removal.*

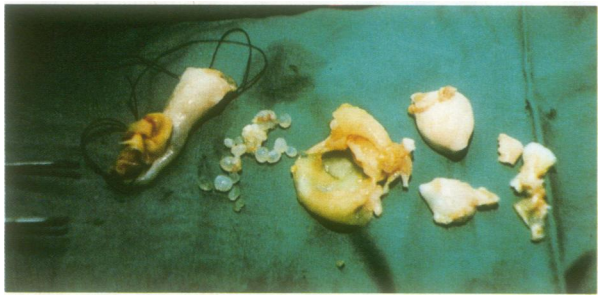

Fig. 4 *Small daughter cysts (left center of photograph), surrounded by fragments of the ectocyst and (right center, top) the large daughter cyst in Figure 3.*

been compromised. Atrioventricular block and syncopal attacks have been reported in at least 7 patients with hydatid cyst of the interventricular septum,[1,8,10] and restoration of atrioventricular conduction after the surgical removal of such a cyst has also been reported.[1]

Since the first successful cardiac echinococcal cystectomy, by Long[11] in 1932, many steps have been taken toward perfecting the treatment of cardiac hydatidosis. Although there is still no medical treatment for cardiac cysts, the results of cystectomy are excellent, with complete recovery the most frequent outcome.[5] This disease should always be treated immediately upon diagnosis, even in asymptomatic patients, because of the significant risk of cystic rupture and its serious — quite possibly fatal — sequelae.

Because the cyst wall is highly fragile and prone to rupture, the cyst must be handled with great care, and the surgical field should be protected throughout with pads soaked in an aqueous sodium chloride solution, or in another helminthicide not toxic to the patient. When feasible, the best treatment is total excision of all cysts. If total excision is inadvisable or impossible, the cyst should be aspirated with a trocar (a syringe with a large-bore needle will do) and its remaining contents sterilized with an appropriate anthelmintic agent.* All contents, including the germi-

TABLE I. Cases of Cardiac Echinococcal Cyst Treated at the Istanbul Thoracic and Cardiovascular Surgery Center from January 1967 through January 1987

Pt. No.	Age, Sex	Surgical Pathology	Operation
1	16, M	Pericardial cyst (7 cm in diameter)	Pericardial resection and cystectomy
2	29, M	Pericardial cyst (huge)	Pericardial resection and cystectomy
3	38, F	Myocardial cyst in RV (6 cm in diameter)	Partial cystectomy under full CPB
4	19, F	Multiple cysts in myocardium, pericardium, and pulmonary artery	Exploration under partial CPB
5	49, F	Pericardial and lung cysts	Pericardial resection and cystectomy; lingula resection
6	30, M	Pulmonary cysts with rupture	Pericardial resection into pericardial cavity and cystectomy; left lower lobectomy
7	18, M	Cyst in the interventricular septum, protruding into the RV	Cystectomy and exclusion, under full CPB
8	23, M	Pericardial cyst (5 cm in diameter)	Partial pericardial resection and cystotomy
9	61, M	External myocardial cyst on LV, compressing LAD (10 cm in diameter)	Cystectomy and exclusion under full CPB; aorto-coronary bypass to LAD
10	39, F	Myocardial cyst in the RV outflow tract (5 cm in diameter)	Cystectomy and exclusion under partial CPB
11	12, M	Myocardial cyst in the AV groove (3 cm in diameter)	Cystectomy and exclusion under full CPB

AV = atrioventricular; CPB = cardiopulmonary bypass; LAD = left anterior descending coronary artery; LV = left ventricle; RV = right ventricle

*Formalin has long been used for this purpose, and we used absolute alcohol; but for obvious reasons these substances should not be applied in areas where thay can easily enter the bloodstream.[12] Some clinicians[3,13,14] have recommended killing the scoleces by instilling a 0.5% silver nitrate solution, because it is fast (due to its low viscosity), nonabsorbable, and comparatively nontoxic.

native membrane, should then be extirpated. When a cyst is not amenable to total excision, its cut edges may be excluded as in the case presented here. If very small, a cystic cavity may be left open, provided its location does not cause interference with blood flow. When the cyst occurs on a thin diaphragmatic wall, however, its edges should be excluded to strengthen the tissue at the wound site. Seldom should it be necessary to create a septal defect in treating septal cysts (but should one occur, it can be repaired with a Dacron patch).

In our series, cardiopulmonary bypass support was used not only in treatment of intracardiac cysts, but routinely when an extracardiac cyst was attached to the epicardium or myocardium. Bypass affords, at minimal operative risk, greater mobilization of the heart and an additional safeguard against fatal accident, should the ventricular wall be perforated during cystectomy or the cyst rupture toward a cardiac cavity.[15] We recommend that a pump be available on standby even during surgical treatment of cardiac echinococcosis confined to the pericardium.

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