

Acta Medica Okayama

Volume 62, Issue 5

2008

Article 9

OCTOBER 2008

Cystic Echinococcosis of the Heart and Brain: A Case Report

Anka Aleksic'-Shihabia*

Edvin Paro Vidolin†

*Department of Neurology, Šibenik General Hospital,

†Department of Internal Medicine, Šibenik General Hospital,

Cystic Echinococcosis of the Heart and Brain: A Case Report*

Anka Aleksic'-Shihabia and Edvin Paro Vidolin

Abstract

A very rare case of echinococcus cysts of the heart and brain in a 27-year-old man who worked as a butcher, lived on a farm, and had a dog before disease onset. The initial manifestation of hydatid disease was anaphylactic shock, the etiology of which remained unknown on initial hospitalization. On rehospitalization, the diagnosis of cardiac hydatidosis was made and the patient underwent surgery. Two years later, reoperation was required for hydatid cyst of the brain and cardiac cyst recurrence. There was no other organ involvement from the disease onset, which is rarely reported. Based on this case, we suggest that echocardiography be performed as a standard method in the diagnosis of anaphylactic reaction of obscure etiology in the areas endemic for hydatidosis, even in the absence of symptoms indicative of cardiac involvement. This especially applies to individuals with occupational or epidemiological exposure to the infection.

KEYWORDS: hydatidosis, cerebral hydatid cyst, cardiac hydatid cyst, diagnosis, treatment

Case Report

Cystic Echinococcosis of the Heart and Brain: A Case Report

Anka Aleksić-Shihabi^{a*} and Edvin Paro Vidolin^bDepartments of ^aNeurology, and ^bInternal Medicine, Šibenik General Hospital, Šibenik 22000, Croatia

A very rare case of echinococcus cysts of the heart and brain in a 27-year-old man who worked as a butcher, lived on a farm, and had a dog before disease onset. The initial manifestation of hydatid disease was anaphylactic shock, the etiology of which remained unknown on initial hospitalization. On rehospitalization, the diagnosis of cardiac hydatidosis was made and the patient underwent surgery. Two years later, reoperation was required for hydatid cyst of the brain and cardiac cyst recurrence. There was no other organ involvement from the disease onset, which is rarely reported. Based on this case, we suggest that echocardiography be performed as a standard method in the diagnosis of anaphylactic reaction of obscure etiology in the areas endemic for hydatidosis, even in the absence of symptoms indicative of cardiac involvement. This especially applies to individuals with occupational or epidemiological exposure to the infection.

Key words: hydatidosis, cerebral hydatid cyst, cardiac hydatid cyst, diagnosis, treatment

Human cystic echinococcosis (hydatid disease) is a zoonosis caused by *Echinococcus (E.) granulosus* occurring endemically in sheep-breeding and cattle-raising areas of southern Europe, the Mediterranean, South America, Africa, Turkey, Australia, New Zealand and India [1]. In our region, it is most common in Dalmatia, and in the neighboring countries of Bosnia and Herzegovina, Montenegro and Macedonia. *E. granulosus* parasitizes dogs, jackals and wolves. Human infection develops upon ingestion of the eggs excreted in animal feces. The embryo develops from the egg and migrates to the circulation via intestinal mucosa. Any organ can be involved by the cyst formation, most frequently the liver (50%–70%) and lungs (20%–30%); however, other organs may also be involved, e.g., the eye, heart, brain or muscle (less than 10%) [2]. Because the cyst grows slowly, the

disease may be asymptomatic for a long period of time. Discharge of the cyst content or cyst rupture can induce an allergic reaction to echinococcal antigen, including anaphylactic shock. In addition, dissemination of echinococcosis may occur. The heart and brain are quite infrequently affected (0.5%–2% and 2% of all hydatid cysts, respectively) [3, 4]. Echinococcosis is usually detected incidentally on x-ray taken for other reasons. A definitive diagnosis is made by use of computed tomography (CT), magnetic resonance imaging (MRI), and ultrasonography (US), in addition to serologic tests that are of limited value alone [5]. The disease is treated by a combination of preoperative chemotherapy and postoperative albendazole, operative treatment with aspiration of the cyst content and injection of antiscolecolid solution.

Case Report

A 27-year-old man affected with hydatid disease presented with severe anaphylactic reaction, the eti-

Received November 12, 2007; accepted June 23, 2008.

*Corresponding author. Phone: +385-22-334713; Fax: +385-22-888392
E-mail: analeksi@inet.hr (A. Aleksić-Shihabi)

ology of which remained unknown on initial hospitalization. He was a butcher, living on a farm, and had a dog before the disease onset. His past medical history revealed exudative pericarditis at the age of 12, and no other serious diseases.

In April 2005, the patient was first hospitalized at the Intensive Care Unit (ICU), Šibenik General Hospital, for severe anaphylactic reaction manifesting with hypotension, generalized urticaria and angioedema, and with normal findings of electrocardiography (ECG), chest x-ray and laboratory tests (except for a nonspecifically elevated IgE of 858 IU/mL). The patient showed no signs or symptoms indicative of heart involvement and echocardiography was not done. On the 4th day after admission, the patient completed his therapy and was discharged for home care in good condition.

One month later, the patient was rehospitalized for anaphylactic shock, this time associated with an apical systolic murmur of Levine grade 2/6 and negative T-waves in leads III, aVF, and V6 on ECG. Transthoracic echocardiography (TTE) showed a partially hyperechoic formation of 50×36 mm in size, localized in the myocardium-pericardium region along the atrioventricular margin on the left, without communication with the circulation, which caused traction of the posterior segment of the mitral annulus with grade III mitral regurgitation. CT of the thorax confirmed the finding, visualizing a cyst of 50×30 mm in size (Fig. 1). Pathologic laboratory findings included eosinophilia of 46% (in 7 days 56%), IgE 2,500 IU/mL and α_1 -globulins 3.8%. Serologic tests for echinococcosis (fluorescence test 1: 600+; com-

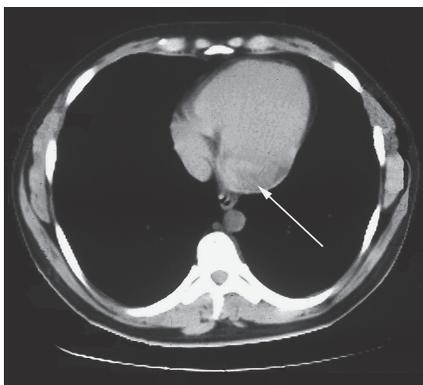


Fig. 1 CT scan of the heart showing an echinococcal cyst along the posterior part of the mitral annulus (arrow).

plement fixation reaction 1: 200+) were positive. CT of the abdomen and pelvis produced normal findings. Therapy with albendazole, 3×400 mg, was introduced as recommended by an infectologist, and the patient was transferred to the University Department of Cardiac Surgery, where he was operated on in July 2005. The hydatid cyst was accessed from the posterior aspect of the left ventricle, where the cyst adhered firmly to the pericardium. The cyst was prepared and 20 mL hypertonic NaCl solution was instilled in the area, followed by cystostomy and evacuation of the cyst content, which consisted of several minor connective sheaths filled with transparent gelatinous content and numerous small white round cystic formations. The material was referred for histopathology and microbiology, which confirmed the diagnosis of echinococcosis. Histologically, the cyst wall was composed of laminated layers of chitinous material. Therapy with albendazole at the same dosage was continued for 10 days postoperatively, when it had to be discontinued due to the elevated level of liver enzymes. The patient regularly presented for infectologic and cardiologic follow up. US of the heart (TTE and transesophageal echocardiography (TEE)) revealed a minor formation of 7×10 mm in size in the area of the mitral annulus. It may have corresponded to annulus enlargement, but recurrence in the area could not be ruled out. Reoperation was not indicated, but further echocardiographic monitoring was recommended. Upon discontinuation of the first course of albendazole, the liver enzymes normalized and the patient received 6 courses of albendazole from November 2005 till April 2006, free from any side effects.

In March 2007, the patient was admitted to the Department of Neurology for severe headaches. On admission, pronounced left-sided hemiparesis, left-sided homonymous hemianopsia, left-sided supranuclear facial paresis and positive Babinski sign on the left were present. An emergency brain CT revealed a round, sharply demarcated hypodense lesion, a capsule of 63×87×60 mm in size, supratentorially parieto-occipitally on the right, causing impression of the ipsilateral ventricular system, pushing it across the medial line by about 10 mm. The septate formation was filled with calcifications indicative of echinococcal cyst (Fig. 2). The patient was transferred to the Department of Neurosurgery at another hospital,

where he underwent right-sided parietotemporal craniotomy through the temporal lobe. The cyst was removed *in toto* and follow up CT yielded normal findings. Antiepileptic therapy (methyl phenobarbitone $tbl \acute{a} 200 \text{ mg } 2 \times 1$) was postoperatively introduced and therapy with albendazole resumed. On the TTE and TEE obtained in May 2007, the cardiologist observed exacerbation of mitral insufficiency (grade III-IV) from previous findings obtained a year before, with a visible cluster of small cysts sized $15 \times 10 \text{ mm}$ in the posterior area of the mitral annulus (Fig. 3). There were no signs of recurrence in the cavity left after the first operation or of echinococcal cysts on some other segment of the heart. US of the abdomen and brain CT showed no signs of echinococcosis. Albendazole was reintroduced in the therapeutic regimen and the patient underwent cardiac surgery in May 2007, with extirpation of the cysts and mitral valve replacement

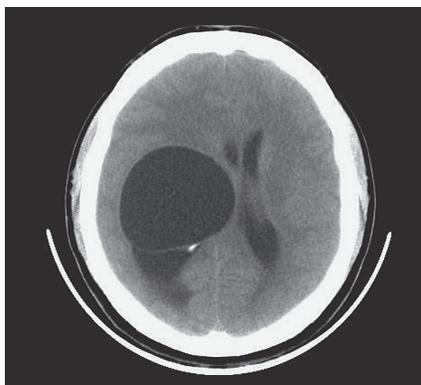


Fig. 2 CT scan of the brain showing a large echinococcal cyst in the right hemisphere of the brain.



Fig. 3 Transthoracic ultrasonography of the heart showing a cluster of small cysts in the posterior part of the mitral annulus (arrows).

by a mechanic valve. After the surgery, anticoagulant therapy with warfarin was introduced and the dose of albendazole increased to 600 mg in the morning and in the evening, and 400 mg at noon. The patient had a normal postoperative recovery.

Discussion

In cases of heart involvement, the cyst is mostly located in the left ventricle (60%), followed by the right ventricle (10%), pericardium (7%), pulmonary artery (6%), left atrium (6%) and interventricular septum (4%) [6]. The embryo is fully grown in 1–5 years of its implantation in the heart. During this stage, the disease usually proceeds asymptotically and can only be accidentally detected. Enlargement in the cyst volume can compromise adjacent structures, thus causing symptoms. In the case of cardiac echinococcosis, the disease can manifest with multiple symptoms, most commonly chest pain and cough, hemophthisis, breathlessness, elevated body temperature, cardiac syncope, anaphylactic shock, arrhythmias and conduction disorders, acute myocardial infarction, pericarditis, valvular dysfunction, pulmonary hypertension and pulmonary embolism, or even sudden death [7]. In the case of cyst rupture, sudden death or anaphylactic shock may be the initial manifestation of the disease [8], as in the case presented. ECG lesions are not always present and depend on the cyst localization. Echocardiography remains the most efficient method in the diagnosis of cardiac hydatid cyst [8]. Radiography of thoracic organs can visualize pulmonary cysts, altered heart contours or calcifications that suggest the diagnosis of hydatid cyst.

Additional diagnostic work-ups include CT and MRI of thoracic organs and examination of other organs for echinococcal cysts by use of computer-assisted imaging techniques, US and serologic testing [5]. Serologic tests are useful in the diagnosis of hydatid disease, but in some patients they may yield false-negative results due to inadequate immune response. About 10%–20% of

patients with hepatic cyst and 40% of patients with pulmonary cyst do not produce a detectable level of specific serum antibodies, thus yielding false-negative test results [9]. In our patient, serology produced positive findings.

Cerebral hydatid cyst is a rare finding, accounting for 1%–2% of all *E. granulosus* infections, with 50%–75% of these being recorded in children [2]. It can be primary, caused by direct infestation via the circulation, or secondary due to spontaneous rupture or an operative procedure for a primary cyst. The symptoms develop due to intracranial pressure elevation and depend on the cyst localization. They are most commonly located supratentorially, in the parietal lobe of the middle cerebral artery territory. Headache and vomiting are the most common initial signs of cerebral hydatid disease, followed by papillary edema, hemiparesis, epileptic seizures, visual impairment, speech difficulties, cranial nerve defects and ataxia [4]. In the diagnosis of cerebral hydatid cyst, CT shows > 90% accuracy. Currently, brain MRI is also used [10]. Operative treatment is used in the management of hydatid disease; however, it may occasionally be contraindicated due to the adjacent vital structures [11]. It is recommended that the cyst be sterilized by the injection of a parasitocidal solution to prevent intraoperative dissemination [12]. For this purpose, the use of hypertonic NaCl, cetrimide or ethanol solution is recommended by the World Health Organization. Pharmacological therapy consists of albendazole (10–15 mg/kg/day) or mebendazole (40–50 mg/kg/day) administered peri- and postoperatively to prevent dissemination or recurrence of the disease [13].

In our 27-year-old male patient, severe anaphylactic reaction of unrecognized cause was the initial manifestation of hydatid disease. Hydatid cysts were detected in the heart and brain, without other organ involvement, which has rarely been reported in the literature. The MEDLINE database search yielded only one case of the heart and brain being affected without other organ involvement [4].

We believe that echocardiography should be performed as a standard method in the diagnosis of anaphylactic reaction of obscure etiology in the areas endemic for hydatidosis, even in the absence of symptoms indicative of cardiac involvement and normal

findings of standard studies (ECG, chest x-ray). This should apply to individuals with occupational or epidemiological exposure to the infection. Upon completion of therapy, the patient should be regularly followed up in order to recognize possible recurrence, dissemination or complications of the disease in a timely fashion.

Acknowledgments. The valuable advice offered to the author by Professor Matko Marušić on preparing this report is greatly appreciated.

References

1. Karabay O, Önen A, Yildiz F, Yilmaz E, Erdal CA, Sanli A, Kilci G, Alqin I, Ital O and Acikel U: The case of a cyst hydatid localized within the interatrial septum. *Jpn Heart J* (2004) 45: 703–707.
2. Iyigun O, Uysal S, Sancak R, Hokelek M, Uyar Y, Bernay F and Ariturk E: Multiple organ involvement hydatid cysts in a 2-year-old boy. *J Trop Pediatr* (2004) 50: 374–376.
3. Kurtoglu N, Ermeydan C, Akdemir R, Basaran Y and Dindar I: Interventricular septal hydatid cyst. *Echocardiography* (2000) 17: 693–695.
4. Bükte Y, Kemaloğlu S, Nazaroğlu H, Ozkan U, Ceviz A and Simsek M: Cerebral hydatid disease: CT and MR imaging findings. *Swiss Med Wkly* (2004) 134: 459–467.
5. King CH: Cestodes; in Mandell, Douglas and Bennett's Principles and Practice of Infectious Diseases, Mandell GL, Bennett JE and Dolin R eds, 6th Ed, Elsevier Churchill Livingstone, Philadelphia (2005) pp 3290–3292.
6. Ben Ismail M, Fourati M, Bousnina A, Zouari F and Lacronique J: Hydatid cyst of the heart. Apropos of 9 cases. *Arch Mal Coeur Vaiss* (1977) 70: 119–127.
7. Laglera S, Garcia-Enquita MA, Martinez-Gutierrez F, Ortega JP, Gatierrvez-Rodriguez A and Urieta A: A case of cardiac hydatidosis. *Br J Anaesth* (1997) 79: 671–673.
8. Zobel C, Kuhn-Reqnier F, Krüger K, Gerharz M, Schneider CA, Müller-Ehmesen J and Erdmann E: Echinococcus cyst located in the interventricular septum. *Clin Res Cardiol* (2006) 95: 600–604.
9. Ammann RW and Eckert J: Cestodes. *Echinococcus*. *Gastroenterol Clin North Am* (1996) 25: 655–689.
10. Onal C, Barlas O, Orakdöğen M, Hepçül K, İzqi N and Unal F: Three unusual cases of intracranial hydatid cyst in the pediatric age group. *Pediatr Neurosurg* (1997) 26: 208–213.
11. Miralles A, Bracamonte L, Pavie A, Bors V, Rabaço G, Gandjbakhch I and Cabrol C: Cardiac echinococcosis. Surgical treatment and results. *J Thorac Cardiovasc Surg* (1994) 107: 184–190.
12. Rossouw GJ, Knott-Craiq CJ and Erasmus PE: Cardiac echinococcosis: cyst removal in a beating heart. *Ann Thorac Surg* (1992) 53: 328–329.
13. Morris DL, Dykes PW, Marriner S, Boqan J, Burrows F, Skeene-Smith H and Clarkson MJ: Albendazole—objective evidence of response in human hydatid disease. *JAMA* (1985) 253: 2053–2057.