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Case report

LEFT VENTRICULAR APICAL HYDATID CYST - A RARE CASE REPORT

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ABSTRACT

Cardiac hydatid cyst is an extremely rare manifestation of cystic echinoccococis, but without prompt diagnosis and management, it can be fatal. Despite the absence of standardized guidelines, an early, complete surgical removal in conjunction with anthelmintic therapy plays a central role and is associated with an excellent prognosis. We report an indigenous case of left ventricular apical hydatid cyst in a 75 - year - old female who presented with atypical symptoms including abnormal fatigue and increasing exercise intolerance. Although serological tests were negative, imaging modalities revealed a 36 x 30 mm oval structure in the left ventricular apex, suggesting a type I hydatid cyst. A single encapsulated cyst was surgically removed through a median sternotomy under cardiopulmonary bypass. Histopathological analysis confirmed a hydatid cyst, and Echinococcus ortleppi was identified. The patient was discharged without incident and was prescribed 3 cycles of Albedazole.

Keywords: *Cardiac hydatid cyst, Echinococcosis, Echinococcus ortleppi, Cardiac surgery.*

I. INTRODUCTION

Hydatid disease (HD) is an uncommon parasitic disease caused by larval forms (metacestodes) of tapeworms of the genus Echinococcus. It remains a major health concern in many parts of the world, especially in less developed regions where animal husbandry is common but there is no veterinary control and where people have close contact with certain domestic carnivores. Echinococcosis is found almost worldwide and is particularly prevalent in Australia, Tasmania, New Zealand, Southern and Northern Africa, South America, the Mediterranean region, the Middle East, Alaska, and Canada [1, 2]. It has also been reported in Asia, with the highest prevalence in Thailand, followed by very low incidence in Indonesia, Malaysia, and Vietnam [3].

E. granulosus and *E. multilocularis* are two of the nine species of Echinococcus with particular medical significance [1, 4, 5]. *E. canadensis* and *E. ortleppi* are also infectious to humans, albeit to a much lesser extent [6]. Their definitive hosts are dogs and other canids, whereas sheep and other ruminants are intermediate hosts. With the consumption of ova in vegetables or water contaminated with dog feces, humans may act as incidental hosts [4, 7]. After the outer capsule of the eggs has been ingested, the freed embryos (oncospheres) pass through the duodenal mucosa and enter the branches of the portal vein. Most of these embryos become entrapped in the hepatic capillaries, where they perish or develop into HC. Some enter the lungs and other organs when passing through the capillary sieve [4]. These embryos may

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reach the myocardium via the coronary circulation from the vascular beds of the liver and lungs [7, 8]. As a result, HD can occur almost anywhere in the body and demonstrates various clinical features that vary according to the number of cysts, their size, growth stage, associated complications, and affected tissue [4]. The liver is the most commonly infected organ (65 - 75%), followed by the lung (25%). Even in countries where HD is endemic, only isolated, sporadic cases of cardiac involvement (0.05% to 2%) have been reported [4, 7]. Any component of the heart may be susceptible, including the left ventricle (50 - 60%), interventricular septum (10 - 20%), right ventricle (5 - 15%), pericardium (10 - 15%), and right or left atrium (5 - 8%) [4, 8, 9].

Theoretically, a Hydatid cyst (HC) has three layers: (1) The outer layer (pericyst), representing the reaction of the host to the parasite, consists of modified host cells, fibroblasts, giant cells, and eosinophils, which together form a rigid protective shell only a few millimeters thick; (2) The middle - laminated membrane resembles the white (albumen) of a hard - boiled egg and is easily ruptured. This membrane is acellular and is approximately 2 mm in thickness. It is impenetrable to bacteria, but nutrients can still pass through. Disruption of the laminated membrane predisposes to infection; (3) The inner germinal (or germinative) layer is thin and translucent. The cyst fluid is crystal clear and contains the scolices - infectious embryonic tapeworms. If it is released into the circulation of the host, it can cause eosinophilia or anaphylaxis [4].

On the basis of their appearance, radiologic findings of an HC are classified into four types: Type I - Simple cyst with no internal architecture; Type II - Cyst with daughter cyst (s) and matrix; Type III - Calcified cyst; and Type IV - Complicated HC [4]. It is emphasized that calcification is more common in the liver (20 - 30%), spleen, and kidney than in the heart [4].

When there is a high rate of rupture (> 25%), septic shock or embolic complications kill 75% of the patients [4, 9]. Despite the absence of standard guidelines for management made variations in treatment strategies, early surgical intervention plays a central role in the management of cardiac hydatid cyst (CHC) and can sometimes be life - saving [7, 8]. CHC surgical excision yields complete recovery and an excellent prognosis [7, 10]. It should not be delayed.

In Vietnam, HD is uncommon given that we are not in the reported endemic areas. Before 2013, this disease had never been reported to be infecting humans in Vietnam, not to mention the CHC [1]. Our objective is to increase the awareness of this diagnosis among physicians and discuss the diagnostic and therapeutic considerations, along with a brief literature review. This case report has been presented in line with the SCARE Criteria [11].

II. CASE REPORT

Our case involved a 75 - year - old female who had never left the country. She is a farmer who raised numerous cattle and dogs. She had led a normal life with no significant medical history until the middle of 2022, when she began to experience some vague symptoms, including abnormal fatigue and exercise intolerance, which progressively worsened and rendered the farm work impossible. Over a course of 7 months, she visited several hospitals and was discharged with the same diagnosis of left ventricular cyst with non - specific oral medications. Due to ongoing symptoms, the patient visited our center.

Admission clinical examinations showed nothing unusual. Several hematological, biochemical, and immunological laboratories were evaluated, which finally gave normal results, including the infectious markers (white blood cells, eosinophils, C-reactive protein and pro - calcitonin) and serial serologies of parasites *(Strongyloides stercoralis, Gnathostoma, Cysticercus cellulosae, and Echinococcus granulosus)*. On cardiac multislice computerized tomography (MSCT), a 36 x 30 mm oval structure in the left ventricular apex appears as a well - defined, water - attenuation mass without hydatid sand and septa, highly suggestive of type I HC (figure 1A).

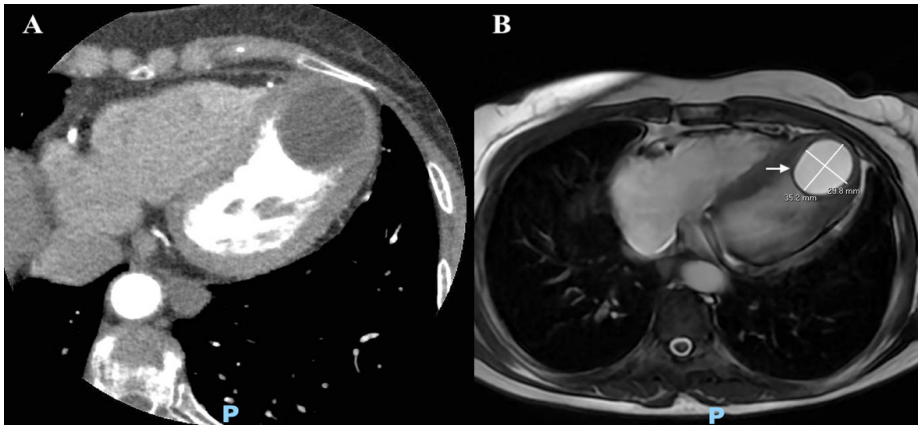

Figure 1: Cardiac multislice computerized tomography images

(A) MSCT of a 36 x 30 mm in size left ventricular cyst quite suggestive of type I HC; (B) Marked hyperintensity of the cyst on T2-weighted MRI of this structure with the "rim sign" (white arrow)

The diagnosis was supported by magnetic resonance imaging (MRI), which revealed hypointensity on T1-weighted images and marked hyperintensity on T2-weighted images of this structure. A low - signal - intensity rim ("rim sign"), which is more evident on T2-weighted MR images, is also observed (Figure 1B). This finding represents the parasitic membranes and pericyst, so it is a characteristic of HC as compared to nonparasitic cyststidosis. Echocardiography showed a left ventricular apical cyst that restricted the surrounding myocardium's motion; the left ventricular ejection fraction was at 47.6%. A full - body MSCT revealed no additional lesions. The patient's electrocardiogram revealed T-wave alterations resembling ischemia, for which she underwent a preoperative coronary angiography indicating insignificant stenoses.

Concerning the risk of rupture and the impairment of the heart's function, we decided immediately to perform surgery. The operation was performed through a median sternotomy under general anesthesia and cardiopulmonary bypass. The cyst was found bulging from the left ventricular apex (Figure 2A).

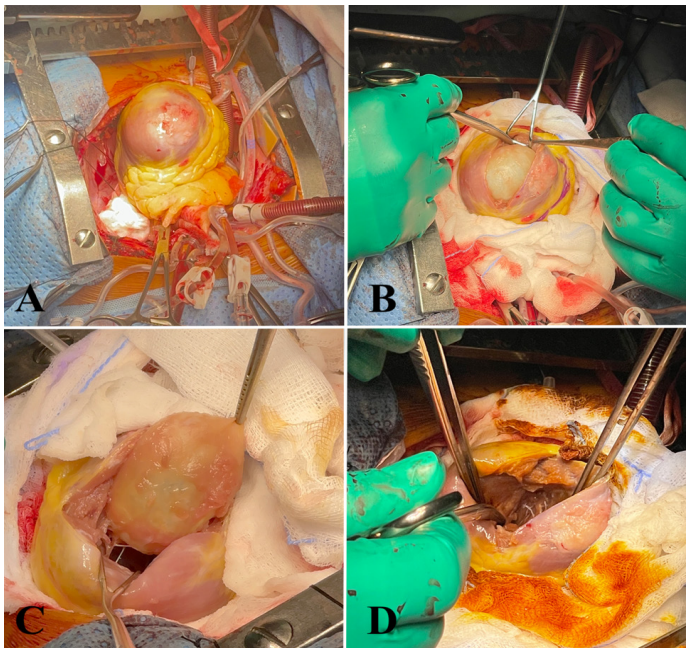

Figure 2: Intra operative images.

(A) Intact hydatid cyst bulging from the apex of the left ventricle; (B) Longitudinal left ventriculotomy right on the cyst; (C) Cyst's intact removal; (D) Residual cavity after cystectomy

The operative field was wrapped with sponges moistened with glutaraldehyde solution to avert embolism and the possible introduction of free scolices to other structures. A longitudinal left ventriculotomy was performed where the cyst was closest to the surface, followed by a meticulous and delicate incision of the intact cyst (Figure 2B, C). Communication between the residual cavity and the left ventricular chamber indicated that this region's ventricular myocardium was completely altered (Figure 2D). Following sterilization with a hypertonic saline solution, the residual cavity was plicated with pledgeted sutures and closed via capitonnage.

The cyst surgically excised contained milky, viscous, and fairly homogeneous fluid.. Microscopic observation of the cyst fluid found a large number of pathogens existing in 2 forms: spherical shape with 2 rows of thorny hooks inward and segmented shape with 1 protruding head, with 2 rows of thorny hooks arranged in an even circle in the peak. (Figure 3).

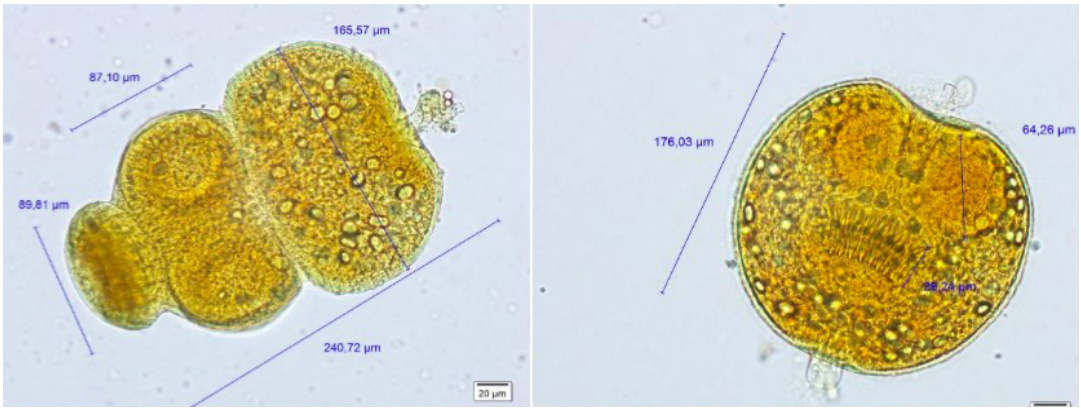

Figure 3: The protoscolices and hooklets of *E. ortleppi* from the reported case

Characteristic images allow us to identify the pathogens belonging to the genus *Echinococcus* spp. Molecular diagnostics: applied PCR technique and sequencing of isolates with specific gene fragments for *Echinococcus* spp. with sizes of 1608 bp in the cox1 gene and 894 bp in the nad1 gene and those in the cox1 gene fragments of 875 bp size and the nad1 gene with a size of 520 bp specific for *E. ortleppi*, the pathogen was identified as *Echinococcus* ortleppi infecting humans and has been reported from China (MZ190836.1, 2021), Egypt (MK492617.1, 2019) and Estonia (KY766908.1, 2017) [12, 13].

The postoperative period was uneventful, and the patient was discharged ten days after the surgery. She has been prescribed Albendazole 400 mg twice daily for 3 cycles of 4 weeks with 2-week breaks between the cycles. At her first 1-month follow-up, echocardiography revealed no evidence of cardiac abnormalities.

III. DISCUSSION

Human HC is still significant globally and poses medical challenges for developing countries. It is difficult to estimate the real number of echinococcosis cases in Vietnam, which may be more prevalent but were underdiagnosed and unreported. This case of cardiac *Echinococcosis* had an atypical clinical presentation, but it was clearly diagnosed as the larvae were detected in the fluid of the surgically removed cyst and identified as *E. ortleppi*.

E. ortleppi (G5) along with *Echinococcus granulosus sensu stricto* (G1) is a hapto type of *Echinococcus granulosus sensu lato* whose larval stage causes HC but to a much lesser extent than that of the latter [6, 14]. There have been only a few cases of HC due to *E. ortleppi* reported in Vietnam, including one CHC [1, 14].

No specific clinical picture could lead to the correct preoperative diagnosis. Our patient was in a similar circumstance. It is estimated that 10% of patients remain asymptomatic for many years, despite the ongoing risk of rupture [7, 10]. Further, it has been estimated that about 20% of fatal cases involve sudden death with no preceding signs or symptoms [7]. For this reason, the diagnosis of HD,

especially that of a CHC, is difficult and usually arises from suspicion, which should be handled with care. The results of serologic tests may be inconclusive due to their variability [10]. Serologic testing is reported to be positive in approximately 80-94% of liver cysts and 65% of lung cysts [15]; however, the rates of seropositivity decline significantly for other organs, and sensitivity may be even lower for cases involving the heart. We emphasize the importance of imaging modalities in rapid diagnosis, screening of adjunctive lesions and complications, and management planning.

The growth of CHCs leads them to be pushed toward a weaker side of the cardiac wall, either the epicardium or the endocardium [7, 8]. Subepicardial rupture is uncommon (approximately 10% of cases), but when it occurs, it may be silent or cause acute pericardial tamponade, constrictive pericarditis, or secondary pericardial cysts [7, 16]. On the other hand, a subendocardial cyst rupture can occur spontaneously in about 40% of patients, causing anaphylactic shock, peripheral, systemic, or pulmonary embolization due to their germinative membrane, and even sudden death [7, 16]. As CHC can simulate a silent bomb, it must be managed urgently to prevent catastrophic events. Multiple reports of sudden deaths relating to HC rupture demonstrate the serious nature of this disease [2, 17, 18].

Initially, our patient was presumed to have an echinococcal infection with a large, isolated cardiac cyst. Based on imaging studies, its appearance and location did not match those of a tumor, vegetation, or thrombus. Because diagnostic biopsy may be associated with the risk of infection spreading to other organs, we decided to proceed directly to the operative bloc to reduce the likelihood of CHC complications.

The method of CHCs' resection does not follow official guidelines but must be individually determined for each patient by the medical team. However, according to Birincioglu and colleagues, superficially located cysts can successfully be removed with a beating heart (off - pump) technique if they are not connected to the ventricular cavity [19]. Resection under cardiopulmonary bypass, since 1962, has been considered the safest method, with the lowest risk of cyst contents leaking during surgery [7, 9, 10, 19]. There are concerns regarding the treatment of residual cavities after cyst removal. After cystectomy, our patient underwent a capitonnage, and no regional contractility and relaxation abnormalities were observed. Postoperative sequelae may include myocardial tearing, atrioventricular block requiring pacemaker implantation, and ventricular arrhythmias caused by the scar. Our patient survived without any of these.

Albendazole is central to this disease's medical therapy [20]. Dehkordi et al. demonstrated that treating hydatid cysts with albendazole may be associated with preventing recurrence and reducing the size and death of the hydatid cysts [20]. However, the use of preoperative anthelmintic therapy for CHC remains controversial [7, 9, 19, 21].

Once the larvae reach the myocardium via the coronary arteries, cysts form in 1 - 5 years [10]. The follow-up extending beyond at least 5 years after the operation using IgG ELISA and echocardiography seems reasonable [22]. This is our follow-up plan for the patient.

IV. CONCLUSION

CHC may remain asymptomatic for years, but there is always a risk of fatal complications so long as the hydatidosis remains undiagnosed and untreated. This report adds to the limited HC database in Vietnam. Our finding expands the geographical distribution of this parasite. This case report emphasizes that this diagnosis should still be considered whenever a cystic lesion is found anywhere in the body, especially the heart, regardless of whether it is typical or epidemiological. Due to the high risk of associated complications, cardiac hydatid cysts should be removed surgically, even in asymptomatic patients. In general, surgery is safe, and the outcomes are satisfactory. Serial follow-up examinations by IgG ELISA and echocardiography as well as other imaging methods should be considered to detect recurrences.

Informed Consent: The patient provided written consent to have their data and images published.

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