Hydatid cyst of interventricular septum with pulmonary artery embolization: Case report and review of literature.

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Abstract:
Echinococcosis is an infection of human caused by the larval stage of Echinococcus and this species is endemic in Iran.
Cardiac involvement in hydatid disease is uncommon (0.5-2%) (2) And usually is intramyocardial.symptom depends on the location, size and integrity of cyst .We present two cases of cardiac hydatic cyst with embolization to pulmonary artery.
Key Words: Interventricular septum, Pulmonary artery embolization.

Echinococcosis is an infection of human caused by larval stage of Echinococcus. cardiac hydatic cysts are rare and represent 0.5 to2% of all hydatic cyst in humans. They are usually found in the left or right ventricle, and exceptionally in the interventricular septum. An exclusively pericardial hydatid cyst is excessively and embolisation to pulmonary artery or systemic arteries is also very rare.
Case report:
An 18 year-old male suffered from dyspnea and fatigue .symptoms was progressed in last 4 months. Other symptoms were orthopnea, PND, palpitation and anorexia. He had two previous operations, a brain cyst was operated 11 years ago and lung hydatid cyst was operated 2 years ago.
BP=120/80 mmhg PR=100
RR=24 Temp=37
In physical examination patient was pale JVP was distended and a 3/6 systolic murmur in left sternal border with paradoxical splitting of S2 was heard.
In ECG RAD and RVH (RV strain pattern) was seen. in chest X-ray cardiomegally and cephalization in lungs were seen .CBC and electrolytes were normal and Casoni test was 2+.
In TEE a well-defined cyst (2 in 1.8 cm)in basal portion of septum with normal size LV and sever RV enlargement and dysfunction, sever RVH and sever TR and sever pulmonary hypertension (100 mmhg) moderate PI with suspicions of emboli in proximal RPA and large PFO were seen.

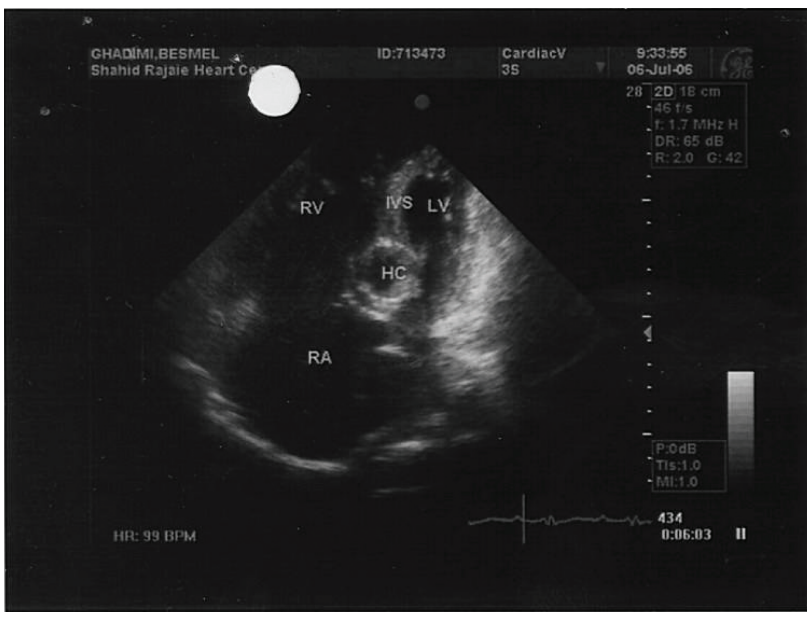

Fig 1: *Echocardiography showed septal cyst*

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In CT Angiography massive pulmonary emboli in both lungs prominently in lower zones and hydatid cyst was seen in interventricular septum of the heart.

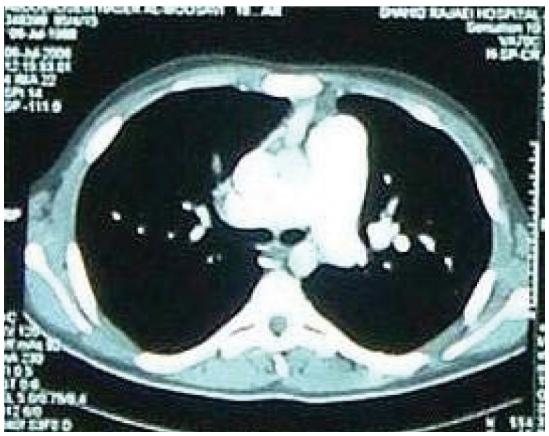

Fig 2: CT- Angiography showed cyst.

In abdominal sonography congestive hepatomegally was seen.

Brain CT scan revealed extensive and hypo dense area in left occipital lobe without any significant edema with the diagnosis of chronic sub cortical infarcts was seen.

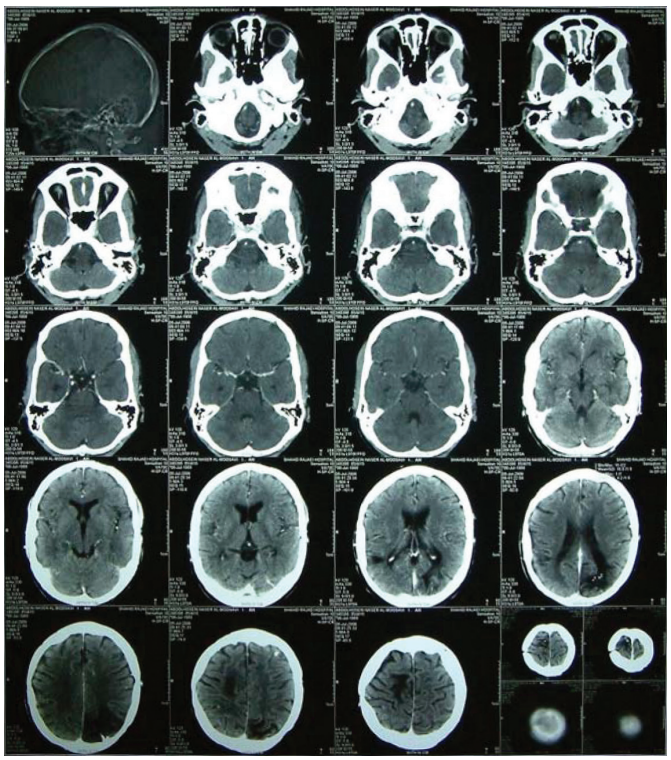

Fig 3: Brain CT –Scan of the patient

In lungs perfusion scan vascular supplying impairment was seen in the lower zones of both lungs.

This patient was operated with the diagnosis of hydatid cyst of cardiac septum with distal embolisation to pulmonary arteries .under CPB first the cyst in the septum was injected with hypertonic saline solution from right atrium and then it was enucleated.

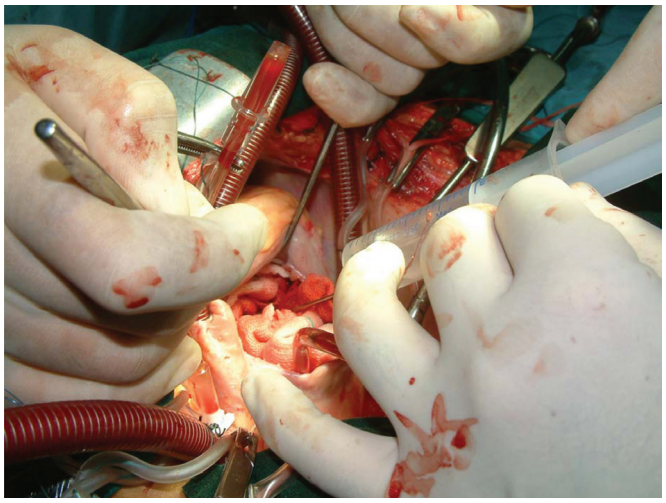

Fig 4: Injection of saline solution to cyst

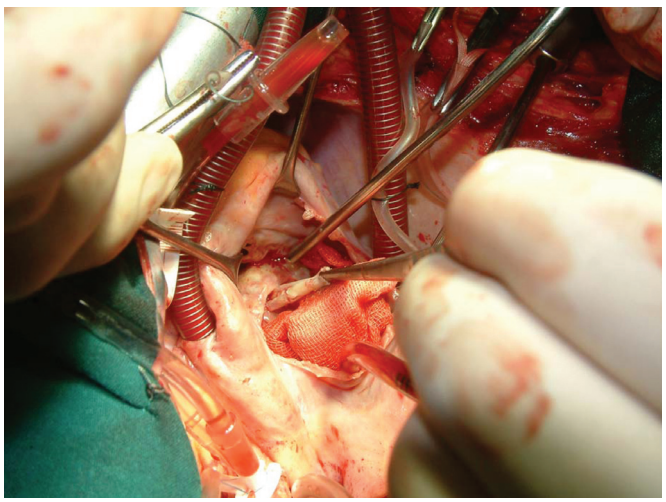

Fig 5: Enucleated cyst.

Then total circulatory arrest performed at 24 degree centigrade temperature and main PA opened .there was two cysts, one in right pulmonary artery and the other was in the left that both were enucleated.

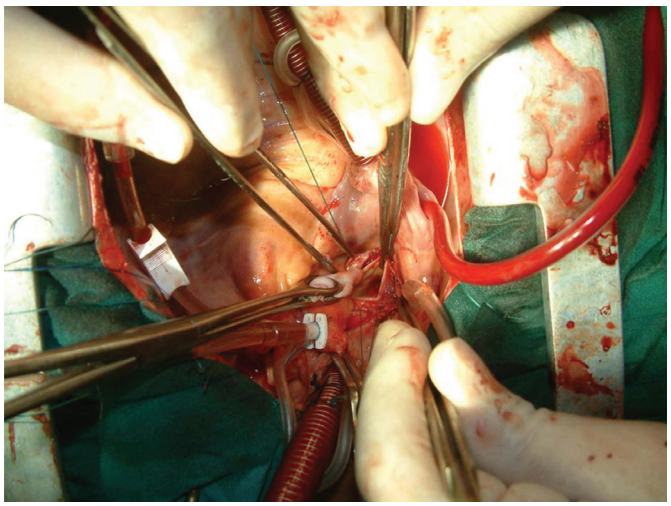

Fig 6: Cyst in left pulmonary artery.

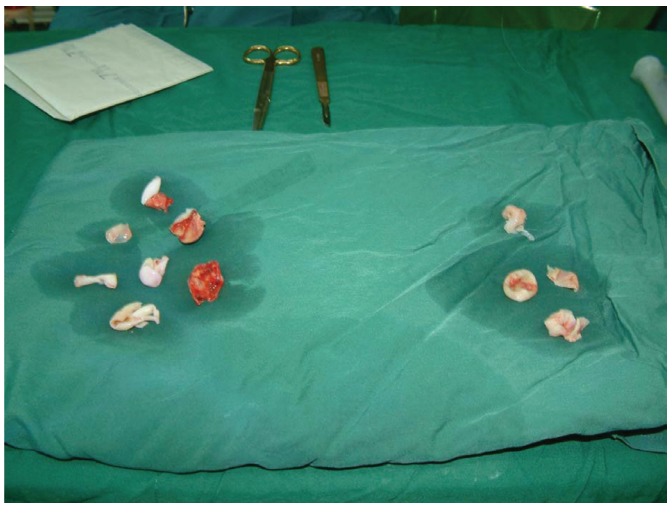

Fig 7: Hydatid Cysts

CPB was discontinued after rewarming with medium dose of Milrinon.

Mean PAP after discontinuation of CPB was 50 mm Hg. This patient was extubated 4 days later in ICU and discharged from hospital 25 days later. The complication was icter with total serum billirobin of 10(direct 5) mg% and hepatic enzymes were raised that responded to conservative management.

The Second case was a 30 year's old woman that was admitted with dyspnea and the history of previous surgery for pericardial effusion six years ago.

Breathing sound in the base of chest was absents bilaterally. in ECG inverted T wave was seen in V5 andV6 and echocardiograph images was those of large echo free space in AV Septum and large mobile mass inferior to SVC in right atrium . After CPB right atrium was opened and cysts were enucleated after saline injection .then main pulmonary artery was opened and the last cyst was enucleated without circulatory arrest.

CPB was discontinued without inotropic agents.ICU stay was 2 days and she discharged from hospital after 14 days. Medical treatment with Albendazol also started for these two patients after surgery but was discontinued in first one because of hepatic enzymes rising.

Discussion:

The tapeworms or cestods are ribbon-shaped segmented hermaphroditic worms which inhabit the intestinal tract of many vertebrates .the term hydatis is the Greek word for a drop of water ,which refers to the fluid filled cysts formed by the echinococcus species .larvae in humans hydatid disease is endemic in most sheep raising countries in Asia, Europe ,south America,newzealand and Australia .the main form is due to Echinococcus granulosus ,most case in Europe and north America occur from immigrants from highly endemic countries .(2)

Like other cestodes, echinococcal species have both intermediate and definitive hosts. The definitive host is a carnivore mainly dogs that harbors the adult tape worm in the small intestine. The carnivore becomes infected by ingesting the larval form in tissue of the intermediate host .the intermediate hosts, chiefly herbivorous mammals and also humans become infected by ingesting tapeworm eggs passed from carnivore faeces.the larval stage is referred to as a hydatid cyst. Human consumption is mostly inadvertent via hand to mouth transmission occurring after close contact such as petting with infected animals. (2)

The ingested parasitic larvae migrate through intestinal mucosa and are carried to the liver by portal venous circulation and lymphatic where most of the larvae are filtered out they migrate to the hosts viscera where they develop into matural larval cysts

If embryos bypass the liver they reach the lungs and other organs via systemic circulation or lymphatic(3).it is well documented that the majority (52-77%) of hydatid cysts are located in the liver followed by lungs (9-44%)(2). Larvae reach the left side of the heart from the coronary circulation, PFO, the lymphatic, or through the pulmonary veins. The host dense fibrous response to the presence of parasite creates an adventitial pericyst layer.(1),(2)

Cardiac hydatid disease is more common in those 20 to 40 years of age .in previous studies the most common locations of cardiac echinococci cyst were the left ventricle(60%) and the ventricular septum (9to 20%) but the right ventricle and right atrium (4to 17%) and sinus valsalva (1.6%) can also be involved .

In three quarters of all cases, the hydatid cyst is enlarged subendocardially in the right heart and subepicardially in the left heart.

There is no uniform clinical presentation of cardiac hydatid cysts, patients may present with symptoms due to mechanical interference with cardiac function, simulating coronary artery disease, arrhythmia, conduction disturbances, pericarditis, and peripheral emboli or as an abnormality of cardiac silhouette on chest x-ray(1),(4). the most frequent symptom was Dyspnea followed by palpitation, angina, and syncope episodes according to the data collected.

Because cardiac hydatid cysts can cause life threatening complications such as cardiac failure, cyst rupture, embolization, etc.the establishment of an early diagnosis and the performance of a timely potentially curative surgical intervention are of paramount importance .the negative serology cannot rule out the diagnosis .echocardiogram provided definitive diagnosis in 94% of cases whereas angiogram was an essential diagnostic modality in patient with symptoms of acute coronary syndrome.

Information in the literature on medical treatment of cardiac hydatid disease is limited .benzimidazol carbamate is a viable option for symptomatic cysts that are inoperable. Albendazol which has good GI absorption is the choice treatment .response to this treatment is apparently related to the thickness of the cyst wall which drug must penetrate.

To reach the germinal layer .therapy is usually in cycles of 28 days on treatment and 14 days off with the dosage of 10 to 15 mg/kg or 400 mg twice a day.

The side effects from the drugs are considerable including the rupture of the cyst or abscess formation, common side effects of Albendazole are abdominal pain, diarrhea, elevated liver enzymes and allergic reactions.

Conclusions:

The differential diagnosis of an intracardiac cyst should include cardiac echinococcal (hydatid) cyst in patients from ship raising countries where echinococcal infestation is endemic.

This study suggests that surgical resection of cardiac hydatic cysts offers a good chance of cure with acceptable operative mortality even in rare cases of pulmonary emboli.

Cystectomy alone or with capitonage appears to be effective in preventing recurrence in the absence of multiorgan involvement. In view of the lack of efficient alternative treatment options, we recommend surgical intervention even in asymptomatic patients in order to prevent the occurrence of lethal complications (2)

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