

ABSTRACT Echinococcosis is a zoonosis caused by adult or larval stages of Echinococcus granulosus. It involves mainly liver and lungs. Pancreatic involvement is found in less than 1%. Clinical feature depends on size and location of the cyst. The main symptoms are pain in left upper quadrant, epigastric or right upper quadrant, jaundice and fever. Its preoperative diagnosis is challenging, as its radiologic findings are often mistaken for other cystic lesions of the pancreas. Endoscopic ultrasound provides more detailed morphological information. Beside this biochemical and cytological sampling helps in differentiating hydatid cysts from other cystic pancreatic lesions. Surgical treatment is considered whenever possible. A definitive diagnosis of hydatid disease of the pancreas can be made only at surgery and, during surgical treatment of hydatid cysts, extreme caution must be taken to avoid rupture of the cysts. A hydatid cyst in the tail of the pancreas can be successfully treated with a distal pancreatectomy.

KEYWORDS :Echinococcosis, ELISA, Hydatid cyst, Pericystectomy, Pseudocyst

Introduction

Hydatid disease is caused by the cystic stage of Echinococcus granulosus and is endemic in many parts of world, it is more common in sheep and cattle raising countries [1]. Echinococcus granulosus is the most common species and is responsible for 95% of the human cystic echinococcosis cases reported [2].

The annual incidence of hydatid disease has been reported to be 18 to 20 cases per 100000 inhabitants [3]. Most hydatid cysts occur in the liver (70%), followed in frequency by the lung (10%), muscles (4.7%). Involvement of the spleen (2.1%), bone and brain (1.4%) are commonly occurring. Other sites, such as the heart, spleen, voluntary muscle, and pancrease are very rarely involved. The reported incidence of hydatid cyst of the pancreas is less than 1% [4]. Even if rare the diagnosis should be considered in any pancreatic cystic mass specially in endemic countries [4].

Etiology and Pathogenesis

Adult tapeworm lives in the upper small bowel of the domestic dogs, a definitive host. Other definitive hosts are wolves, jackals, domestic cats, and reindeer etc. Sheep, cattle, pigs and humans contain larval stage and are intermediate hosts. They are infected feco-orally by echinococcus eggs shed in the environment with feces of infected dogs [1].

Eggs hatch in intestinal mucosa and transform into oncospheres, which penetrate bowel wall, enter portal venous blood, and are carried to the liver. The embryos become encysted and grow at the rate of about 1 cm/yr. The hydatid cyst (figure-1) is composed of a fibrous and inflammatory outer layer of host origin, a laminated cuticle and a germinal membrane where protoscoleces grow and secrete clear fluid. From germinal layer, daughter cysts may be formed by invaginations. The lifecycle (figure-2) is completed when the definitive host is infected by viable cyst containing organs of intermediate host. In the intestine of the dog, protoscoleces develop into adult tapeworms. Man is a dead end host and becomes feco-orally infected by echinococcal eggs [1].



Figure-1 Hydatid cyst Source:www.googleimages.com/H. cyst



Figure-2 Life cycle of Echinococcus granulosus. Source:www.googleimages.com/E.granulosus.

Clinical features

Due to slow growth of the cyst, most patients have no symptoms. Large cysts (usually >10 cm in diameter) may cause right upper quadrant pain, which is the most characteristic symptom. Pain may be intermittent or continuous and gradually increasing over a long period of time. Due to increased travel and population migration, all clinicians should be familiar with this disease that used to be endemic only in certain areas of the world.

Infestation of the pancrease occurs by arterial route after passage of hepatic and pulmonary filter. Localization may be in the head of pancrease (57%), body (24%) and tail (19%).High frequency in head region because it is more vascularized [5]. The clinical presentation of the pancreatic cyst depends on their size and anatomic locations. Pancreatic hydatid cyst located in the head may present with obstructive jaundice due to compression of common bile duct or and spillage of scolices into the biliary tree and it may lead to recurrent attack of acute pancreatitis. While cyst located into body or tail presents with less specific clinical features like abdominal pain, discomfort, vomitting, and feeling of fullness [6].

Complications:

 Cyst may rupture into the biliary system (leading to cholangitis with or without obstructive Jaundice. and marked eosinophilia), into the peritoneum (leading to anaphylaxis /peritoneal dissemination) or into the pleura or lung (causing pleural hydatidosis or bronchial fistula).

- There may be compression of splenic vein by the cyst cause portal hypertension.
- There may be recurrent attack of acute pancreatitis.
- Thrombosis of superior mesenteric artery.
- Pancreatic abscess and pancreatic fistula are unusual complications.

Diagnosis:

The diagnosis of pancreatic cystic lesion can be performed by clinical examination and imaging like Ultrsonography, CT scan, MRI and Endoscopic Ultrasound. Inspite of these it is often difficult to differentiate this from pancreatic pseudo cyst or cystic neoplasm. Aspiration of cyst fluid for analysis or biopsy of cyst wall has been recommended for this situation. Microscopic examination of the cyst content confirms the diagnosis. The preoperative diagnosis of hydatid cysts is very difficult. Puncture of primary hydatid cyst is avoided if diagnosis is suspected. When CT and MRI failed to provide diagnosis of cystic lesion of the pancreas when positive; however with negative serology the hydatid cyst cannot be rule out.

Cystic lesions of the pancreas (CLP) are relatively common, because of rarity of Primary hydatid cyst and the presenting symptoms clinical features may be similar to some other more commonly encountered cystic lesions of the pancreas [7]. Differentiating among these cysts is challenging.

CLPs include pseudocysts, retention cysts, duplication lymphoepithelial cysts, congenital epithelial cysts, cysts which occur in association with polycystic disease or von Hippel-Lindau disease, serous cystic adenomas, mucinous cystic neoplasms, intraductal papillary mucinous tumors, solid pseudopapillary tumors, cystic metastases from cancer of the lung, ovary, or melanoma, vascular tumours (haemangioma or lymphangioma).

Treatment

Surgery:

Surgery remains the treatment of choice in hydatid disease. Many surgical procedure reported for panceatic cyst like partial or total cystectomy, marsupialization and external drainage. Surgical treatment can be managed either by laproscopy or laprotomy. Because of the delicate nature of the laproscopic procedure and the importance of avoiding spillage of the cyst contents. Open resection has been the standard of care for the treatment of pancreatic echinococcosis. If cyst is present near vascular structure like biliary or pancreatic duct and high risk of pancreatic fistula then partial cystectmy is preferred in such case. Distal pancreatoromy if cyst is in tail of pancrease. If cyst is in head a cystojejunal anastomosis can be performed to prevent a postoperative pancreatic fistula.

Mainly two types of surgeries are performed:

- Radical
- Evacuation of cyst content

Radical surgical resection includes liver resection, pericystectomy and cystectomy and has the advantage of prevention of intraabdominal spillage. The cyst is entirely removed and its opening is avoided. Liver resection is the most expensive technique.Pericystectomy involves a non-anatomical resection of the entire hydatid cyst including endocyst and pericyst. While in cystectomy only laminal layer, germinal layer and cyst content are removed pericyst is not resected.

Surgical evacuation of cyst is performed when radical surgery is not possible. Preoperative use of Scolicidals like 20% hypertonic saline, 95% alcohol or cetrimide prevent intraabdominal seedling of proctoscoleces.

Medical treatment:

Chemotherapy is effective in small cyst with diameter having < 4 cm, those with thin walls and in younger patients. It is also indicated in patients who are at high risk for surgery, in patients with multiple peritoneal cysts, cysts in multiple organs, bone , in brain, to prevent secondary echinococcosis after spillage during surgery and as a concomitant therpy with percutaneous drainge [8,9].

Albendazole (10 mg/kg/day) is the most common drug used in medical treatment of hydatid cyst. Albendazole kill the parasite by impairing the glucose uptake and effective against the larval stages of E. granulosus. Multivesicular cysts, hepatic cysts & cysts in older patients respond less well to treatment than univesicular, pulmonary cysts and cysts in younger patients <30 years of age[10].

Percutneous treatment:

It is minimally invasive and very effective in the treatment of hepatic hydatidosis.

Technique is called PAIR (Puncture-Aspiration-Injection- Re-aspiration). Usually Albendazole prophylaxis is started 1 week before PAIR and continued for 3-4 weeks there after. Cyst is punctured under ultrasonographic guidance. Cyst fluid is aspirated; 20% hypertonic saline or 95% alcohol is injected. Scolecidals are re-aspirated after 10 minutes. Following intracystic injection of scolecidals, both germinal layer and protoscoleces become instantaneously non-viable.Success of PAIR is defined as detachment of endocyst,rupture of daughter cysts, and non-viable protoscoleces at microscopy of cyst fluid. Ultrasonography may show heterogeneous reflection of cyst contents at 3 months, obliteration and pseudo tumor aspect at 5 month or loss of echogenicity and disappearance of cyst a 9 months.

Indications to PAIR : An echoic lesion > 5cm in diameter with no cyst biliary communication, patient refusing surgery or unfit, or has relapsed after surgery, pregnant patients, children > 3 years of age and multiple cysts > 5cm in different liver segments.

Contra indication to PAIR: Cystobiliary fistula, cysts at risky locations, inaccessible cysts, multiple septal divisions, echogenic lesions, and inactive or calcified cysts.

Conclusion.

we can conclude that primary hydatid cyst even it is very rare, yet it should be considered in the differential diagnosis of the cystic lesion of the pancrease. It occurs most commonly in liver and lung but pancreatic hydatid cyst has been rarely reported. The location of the cyst in the pancreas has different distributions. Clinical presentation is variable and insidious, depending on the location and the size of the cyst. Epigastric pain is the most frequent symptom. Cysts in the corpus and tail rarely cause any symptoms and abdominal mass may be found on examination. A definitive diagnosis of hydatid disease of the pancreas can be made only at surgery. Surgical treatment is to be considered whenever it is possible and depends on the location of hydatid cyst. In pancreatic disease, it's essential to maintain the organ's exocrine and endocrine functions. Many surgical techniques are available to remove the cyst; however pericystectomy with drainage of the residual cavity is the technique of choice. Because of the localization of the cyst near a major biliary or vascular structures and high risk of pancreatic fistula_total pericystectomy cannot be done in such cases.

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