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Congenital true cystic mass of the pancreas in a young woman: which treatment?

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Aim. **We report a case of congenital true pancreatic cyst with high level of enzymatic activity, rarely seen in young and children which, to the best of our knowledge, rarely has previously been reported. A young girl was admitted to our clinic with a history of abdominal swelling and pain during exercise for many months. A mobile, smooth, non-tender mass was palpated on the left side of the abdomen during physical examination. Ultrasonography and computed tomography imaging revealed a not subdivide cystic mass sized 11.51x8.2 cm.**

Methods. **Surgical treatment was considered, but given the young age of the patient, we declined for fear of complications. Therefore, US-guided percutaneous drainage of cystic was performed. After a week, at the ultrasound check, thanks to obstruction of drainage, we decided to intervene surgically. During abdominal surgical exploration, the pancreatic tail was larger than normal, and a pancreatic cyst arising from the tail of the pancreas was observed. Total cystectomy was performed with distal pancreatectomy.**

Results. **The postoperative period was complication free and the patient was discharged on the seventh postoperative day. No complications occurred during the six-month follow-up period.**

Conclusion. **Early diagnosis and timely intervention have improved the results of surgical therapy in this disease. Although it is extremely rare in children, congenital true pancreatic cyst should be diagnostically considered in cases involving a cystic mass neighbouring the pancreas. Ultrasonography, computed tomography and magnetic resonance imaging is helpful in differential diagnosis of other cysts originating from neighbouring organs. Total excision with distal pancreatectomy and splenic preservation are advised for distal pancreatic cyst.**

Key words: **Pancreatic neoplasms - Congenital abnormalities - Cysts - Ultrasonography.**

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Developmental anomalies of the pancreaticobiliary tree are usually detected in children; congenital abnormalities may be first found later in life.

Less than 1% of pancreatic cysts seen in young and children are congenital true pancreatic cysts and just over 25 cases were reported as congenital true pancreatic cysts in the literature.[1]

In young and children (under the age of two years),[1] the symptoms and signs are often nonspecific (intermittent abdominal pain, nausea and occasionally vomiting) and diagnosis can be difficult and frequently, an extensive work-up is undertaken before the correct diagnosis is made.

Persistent but unexplained upper abdominal symptoms, an underlying congenital abnormality of the pancreatic body, duct or bile duct should be considered.

In true cyst, enzymatic activity of cystic fluid is at normal or low levels;[1] we report a rare case of congenital true pancreatic cyst with high enzyme activity of cystic content.

Although the results of ultrasonography (US), computed tomography (CT), and magnetic resonance imaging may suggest the abnormality in some cases, cholangiopancreatography is often needed to reach a definitive diagnosis and more clearly outline duetal anatomy.

Recognition of congenital anomalies may aid in surgical planning and prevent inadvertent injury.

Case report

A sixteen years old girl (48 kg) was admitted to our clinic with a history of abdominal swelling, intermittent abdominal pain, mild nausea and vomiting for four months.

On physical examination, a mobile, smooth, nontender mass was palpated at the left upper quadrant of the abdomen.

The laboratory showed that routine blood, liver function tests and urine biochemistry profiles were at normal levels.

Laboratory studies including tests for serum lipase and amylase showed no abnormalities; IgG- and IgM-type antibodies against toxoplasma, rubella, cytomegalovirus, and herpes simplex virus infections were negative.

Outliers were detected in some biochemical markers such as CEA (8.49 ng/mL), CA19.9 (460 U/mL) and CA125 (65 U/mL); normal values for LDH and NSE.

Abdominal ultrasonography demonstrated a not subdivide cystic lesion sized 11.51x8.2 cm (volume: 49.43 cm³), anechoic, low-attenuating mass typical of simple cyst (Figure 1), located medial to the spleen, originating from the left midline lying towards the pelvis, near by the left kidney and spleen.

Abdominal MR images revealed that the left kidney and the spleen were displaced towards the posteriolateral and superior, respectively, by a not subdivide cystic mass. The body and tail of the pancreas and tissue interfaces could not be observed.

CT noted voluminous pancreatic cyst (size 11x11x17 cm), hyperdense and calcified walls, likely to be congenital nature; liver, bile ducts, gallbladder, splenic, adrenals, kidneys and bladder of normal appearance (Figure 2).

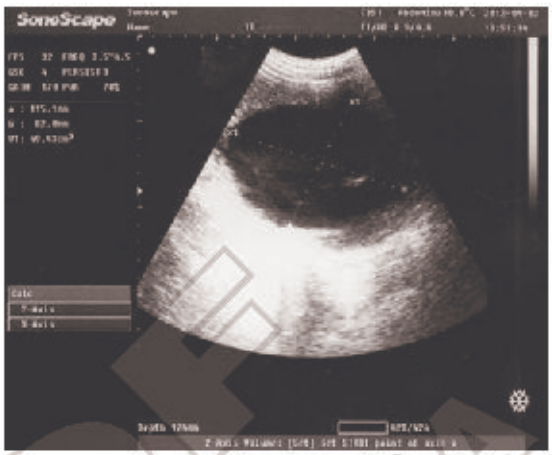

Figure 1.—Abdominal ultrasonography control showed not subdivide cystic lesion sized 11.51x8.2 cm (volume: 49.43 cm³) causing abdominal swelling and pain.

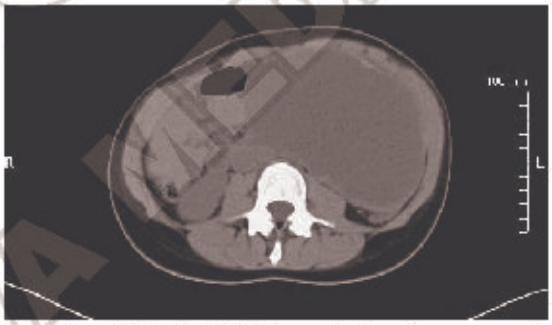

Figure 2.—Abdominal CT showed voluminous not subdivide pancreatic cyst with hyperdense and calcified walls, likely to be congenital nature (circle); cystic lesion sized 11x11x17 cm.

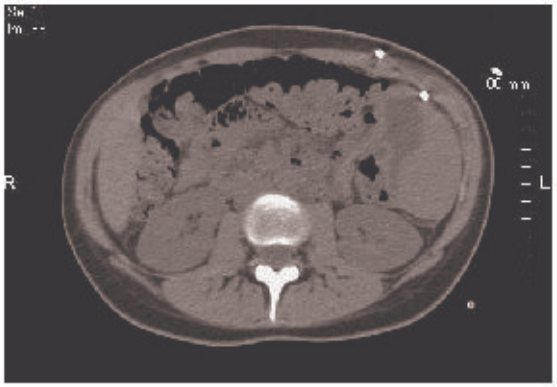

Figure 3.—Postdrainage-abdominal CT revealed cyst collapsed with partially calcified walls (circle); the arrows indicate site of entry and exit of "pigtail" percutaneous US-guided drainage.

Surgical treatment was considered, but given the young age of the patient, we declined for US-guided percutaneous drainage of cystic (external drainage pig-tail ultrasound-guided, inserted after local anesthesia in abdominal lumbar region), prior antibiotic prophylaxis (intravenous ceftriaxone 2 g).

About 850 mL of clear fluids was drained via catheter, in the absence of endoscopic or clinical complications.

Examination of the cystic contents, was: sediment consisting of red blood cells, few lymphocytes and some histiocytes; biochemical analysis of the cystic fluid revealed an amylase activity of 2300 U/L.

After the percutaneous drainage, the pain subsided dramatically.

Postdrainage-CT revealed cyst collapsed with partially calcified walls and modest free fluid under-mesocolico; no enlarged lymph nodes (Figure 3).

The catheter was removed one week after the procedure, due to obstruction of the same, and the self-sustaining of cyst; there were no complications or symptoms after removal of the catheter.

After removal, thanks to the return of mild symptoms, we decided to intervene surgically, through surgical exploration of the patient.

This procedure revealed one major cystic masse originating from the tail of the pancreas.

This mass was filled with a yellow-colored cystic fluid and was slightly adherent to adjacent structures; the tail of the pancreas was observed to be wider than usual.

Distal pancreatectomy with splenic preservation and total cystectomy was performed.

The postoperative period was complication free and the patient was discharged on the seventh post-operative day.

No complications occurred during the six-month follow up period.

Histopathological examination revealed congenital cystic mass from the tail of the pancreas (normal cytologic and histologic figures); this mass was embedded in the pancreatic tissue and coated with cuboidal epithelial tissue.

The follow-up (on-going), up to now, showed no recurrence of the pathological and no complications, making sure the authors of the efficacy of the surgical treatment.

Discussion

Important factors in the development-promoting congenital anomalies of the pancreaticobiliary ductal system are familiarity, embryological and environmental factors; to date, the exact etiology for development of a cyst is unknown.

True congenital pancreatic cyst (derived from an abnormal segmentation of the primitive ducts of the pancreas) frequently appears as solitary, unilocular, nonenzymatic, sterile and fluid-filled, usually from tail, as in our case report, or neck of the pancreas in 62% of cases, head of the pancreas in 32% of cases in literature[1].

Pancreatic cysts can be either of six types: congenital-developmental cysts, retention cysts, duplication cysts, pseudocysts, neoplastic cysts (further subdivided into serous cystadenomas, mucinous cystic neoplasms, intraductal papillary mucinous neoplasms or lymphoma), and parasitic cysts (echinococcus cyst).

The first three types are also known as true cysts and these cysts have true epithelial components on their walls; in the most frequently seen post-traumatic pancreatic pseudocysts, the walls of the cysts do not contain epithelial components.

Isolated-single congenital true cyst of the pancreas is extremely rare event (as in our patient); in many cases the congenital true cysts of the pancreas are multiple, unilocular or multilocular, usually range in size from microscopic to 5 cm, and are associated with genetic disorders that involve cystic disease in other organs, including adult polycystic kidney disease, von Hippel-Lindau disease and cystic fibrosis or also associate with tubular ectasia, polydactyly, anorectal malformation, and asphyxiating thoracic dystrophy.

These cysts may also occur as solitary lesions, and are usually small and may either diffusely involve the pancreas or localize to one region; the number of cysts varies, but, typically, pancreatic involvement is less than the cystic change seen in the kidneys and liver.

Rarely, pancreatic involvement may predominate and concomitant hepatic and renal involvement facilitates diagnosis when cysts also are seen in the pancreas.

In our case, the presence of cyst, showing continuity towards the tail of the pancreas was evaluated as indicators of ductal developmental anomaly. Furthermore, the epithelial component on the cystic wall was indicative of a true cyst.

Generally, symptoms of congenital pancreatic cysts are seen in patients under the age of two years, in opposition with symptoms of pseudocysts, most common during the fourth and fifth decades corresponding to the peak incidence of pancreatitis.

Congenital pancreatic cysts are generally asymptomatic; symptoms of abdominal pain, nausea and vomiting, weight loss, jaundice (secondary to pancreatitis and cyst compression of the common bile duct and peri-ampullary region), fever, diarrhea, hematemesis, melena, perceived abdominal mass, physical findings of tenderness and an palpable abdominal mass are the common hallmarks of the diagnosis.

In our patient, evident symptoms related to the cystic mass had not been observed, except for abdominal distention and pain during exercise.

It is difficult to distinguish post-traumatic cyst from congenital true cyst only with the clinical, diagnostic and histopathological features; intervenes to help the enzymatic activity of the cyst.

In congenital cystic fluid low enzymatic activity is observed generally[1], whereas enzymatic activity of pseudocysts is considerably high (1000-3000 U/L).

In our patient, cystic fluid had an amylase activity of 2300 U/L, higher than the normal limits; this activity is the value expected for a retention cyst.

For an accurate localization of the abdominal masses (occasional findings of pancreatic cystic lesions are found in approximately 1% of all abdominal scans), there is necessity of computed tomography (CT), magnetic resonance (MR) imaging or ultrasound (with EUS guided cyst aspiration); these techniques show detailed information, including the characteristics of the wall, septum, internal structure and surrounding tissues; the same techniques in cystic fibrosis, sequent calcification and chronic disease show progressive dilatation of the acini, fatty replacement of the pancreatic parenchyma and dilated remnants of the pancreatic ducts with secondary fibrosis and pancreatic atrophy.

The relationship between a cyst and surrounding tissues, and extensions of a cyst can be demonstrated with MR imaging (due to its multiplanar capacity) than with CT. We believe that for evaluating extensions and the nature of cysts in pediatric patients, MR imaging is a good choice.

However, the authors believe in ultrasonography (EUS) as rapid and reliable technique used for cystic evaluation, its differentiation in the abdomen and other side; the ultrasound method is very helpful in evaluating the patient who has an congenital abdominal mass, elevated amylase, trauma or episode of pancreatitis.

The only limitation of the endoscopic ultrasound technique is that it is only indicative of a nonsolid mass.

EUS technique-guided fine needle aspiration (FNA) of these cystic lesions has led to their better cytology detection, molecular analysis and characterization.

Cyst fluid can be further studied after aspiration in order to analyze cytology, viscosity, extracellular mucin, tumour markers (for example (CEA, CA 19-9, CA 15-3, Ca 72-4), enzymes (amylase, lipase), DNA analysis (quality/content or mutational).

The diagnostic accuracy of EUS alone for detection of malignant or premalignant cysts reaches 95%[2], although it has important limitations for the differential diagnosis of benign and malignant cysts with overall accuracy rates of 40 to 93% cyst contents and sampling of the cyst wall or septa, as well as mural nodules[2].

Rare disease that is commonly misdiagnosed preoperatively is "epithelial cyst in an intrapancreatic accessory spleen" (often mistaken as congenital pancreatic cyst).

Serious complications secondary to rupture, inflammation/infection, or hemorrhage into the true congenital pancreatic cyst are rare, but may occur and they bring difficulty in diagnosis; studies indicate that rupture was associated with a 60% mortality, hemorrhage with a 17% mortality, and inflammation/infection with a 21% mortality.[3]

The literature emphasizes several points in the treatment of congenital cyst.

External drainage procedures appear to be related to an increasing awareness of the

criteria for individualization of treatment; the same may be definitive, or show need for additional approaches.

Internal drainage methods (cyst-gastrostomy, cyst-jejunostomy or cyst-duodenostomy) should be the preferred treatment for small to medium sized cyst in the pancreatic head, body or tail not drainable externally so resolutive (a well formed cyst membrane is necessary for internal drainage); if it is not possible, the surgical procedure is total excision, particularly if the cyst is located in the distal pancreas and is small cyst.

Close relationship to the duodenum or stomach and contiguous association to these organs is a prerequisite for anastomosis at these sites.

A Roux-en-Y jejunal anastomosis can be used if the cyst is not attached to the duodenum or stomach, or if this is the more easily accomplished procedure, provided that the cyst is not infected and that it does not have a thin friable capsule.

Other procedures are often associated with the internal drainage, or other type of drainage, such as cholecystectomy or choledochoduodenostomy.

In our patient, total excision with distal pancreatectomy and splenic preservation was performed for the distal pancreatic cyst.

The cost effectiveness of different strategies of management was studied in asymptomatic pancreatic cystic neoplasms[2]; the study has further emphasized the utility of EUS-guided FNA with cyst fluid analysis.

Literature compared three approaches in solitary asymptomatic pancreatic cystic neoplasms.

The approaches were: 1) no specific intervention; 2) an aggressive surgical intervention; 3) EUS-guided FNA with cyst fluid analysis approach; the results showed that the latter approach yielded the highest quality adjusted life years with an acceptable incremental cost effectiveness ratio[2].

Conclusions

The authors believe in rarity of the presentation of this congenital disease.

Young and children with symptoms may undergo extensive diagnostic evaluation and computed tomography, ultrasound and magnetic resonance imaging studies can help, in combination with biochemical tests (such as amylase assay).

Because a cystic mass of the pancreas is not always a pseudocyst, percutaneous drainage should be followed by cytologic analysis of the aspirate.

A biopsy of the wall of the cystic mass may also be necessary.

Early diagnosis and timely intervention have improved the results of therapy in this disease.

The authors believe strongly in the surgical treatment of congenital cyst of the pancreas, that if done accurately can lead to cure, without forgetting the alternative treatments, such as percutaneous drainage EUS-guided, especially in young patients.

In condusion, with true pancreatic cyst, enzymatic activity of cystic fluid can be higher than would be expected (like in our patient).

For surgical treatment, total excision with distal pancreatectomy and splenic preservation is preferred.

Riassunto

Cisti pancreatica congenita in una giovane donna: quale trattamento?

Obiettivo. Riportiamo un caso di cisti pancreatica congenita con alto livello di attività enzimatica, rara nei giovani e nei bambini, raramente riportata in letteratura.

La nostra giovane paziente è stata ricoverata nella nostra clinica con storia di gonfiore addominale e dolore durante l'esercizio fisico, della durata di molti mesi.

All'esame obiettivo era evidente una massa liscia sul lato sinistro dell'addome.

L'ecografia e la tomografia computerizzata hanno rivelato la sovracitata massa cistica, non settorializzata, di dimensioni 11,51 x 8,2 cm.

Metodo. Il trattamento chirurgico è stato considerato, ma data la giovane età della paziente, abbiamo optato per un trattamento meno cruento, per paura di complicazioni.

Pertanto, è stata eseguita una tecnica drenante ecoguidata percutanea.

Dopo una settimana, al controllo ecografico, a

causa dell'ostruzione del drenaggio, abbiamo deciso di intervenire chirurgicamente.

Durante l'esplorazione chirurgica dell'addome, abbiamo notato una coda pancreatica più grande del normale, ed è stata notata l'origine cistica nella coda pancreatica. Abbiamo eseguito una cistectomia totale associata a pancreasectomia distale.

Risultati. Il periodo postoperatorio è stato libero da complicanze e il paziente è stato dimesso in settima giornata post-operatoria.

Nessuna complicazione è intervenuta durante i sei mesi di follow-up.

Conclusione. La diagnosi precoce e il tempestivo intervento hanno migliorato il risultato terapeutico chirurgico di questa malattia.

Ecografia, TC e RM sono metodiche utili nella diagnosi differenziale delle cisti addominali.

L'escissione totale della cisti, la pancreasectomia distale e la conservazione splenica sono da considerarsi come l'ideale trattamento.

Parole chiave: Tumori pancreatici - Anomalie congenite - Cisti - Ultrasonografia.

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