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Case Report

Endoscopic transgastric drainage of pancreatic pseudocyst in hereditary pancreatitis. A case report

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Abstract

Hereditary pancreatitis (HP) is a rare genetic disorder characterized by acute recurrent pancreatitis (ARP) and chronic pancreatitis (CP) that runs in families. It' s symptoms are usually typical ones pancreatitis but HP is more amenable to treatment, especially when complication presents. Recent single-center studies have identified several genetic risk factors, including cystc fibrosis transmembrane conductance regulator (CFTR), cationic trypsinogen (PRSS1), pancreatic secretory trypsin inhibitor (SPINK1), chymotrypsin (CFTP) and carboxypeptidase 1 (CPA1) genes. Other risk factors include obstructive, traumatic, infectious and systemic causes. Our case report presents a 9-years old boy, with a pancreatic pseudocyst (8cm in diameter) as a consequence of recurrent episodes of pancreatitis. The diagnostic investigation (MRCP, Cystic Fibrosis test) had proved no obvious aetiology. An enterocystic roux en Y anastomosis was performed, but the boy continued o develop episodes of pancreatitis and after 2 years, he presented with a new pancreatic pseudocyst (PPC) of the same dimensions, which was attributed to genetic factors. A more conservative approach was decided: endoscopic transgastric drainage of the pseudocyst. In gastroscopy, the cyst was protruded on the posterior wall of the stomach and it was drained into it via a pig tail catheter, which was removed 6 weeks later. After 3 months abdominal ultrasonography follow up confirms the successful drainage of the pseudocyst.

Keywords: Hereditary pancreatitis (HP), pancreatic pseudocyst (PPC), endoscopic transgastric drainage (ETD), acute recurrent pancreatitis (ARP)

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Abbreviations:

HP: Hereditary pancreatitis

CFTR: Cystc Fibrosis Transmembrane Conductance Regulator

PRSS: cationic trypsinogen

SPINK1: Pancreatic Secretory Trypsin Inhibitor

CFTP: chymotrypsin

CPA1: carboxypeptidase 1

PPC: Pancreatic Pseudocyst

CF: Cystic Fibrosis

MRCP: Magnetic Resonance Cholangiopancreatography

Introduction

Pancreatitis in children has been diagnosed more frequently in the past few decades. (Skuhla-Udawatta et al, 2017),(Parniczky et al,2018) and it differs from that of adults in causes and treatment as as well in prognosis. The international, multi-center INSPPIRE (International Study Group of Pediatric Pancreatitis: In Search for a CuRE) consortium was created to address this issue by collecting data on the largest group of pediatric patients with ARP or CP to date. (Soma Kumar et al.2016).

Leading causes of pancretitis n children are 1) biliary system abnormalities, 2) systemic diseases (cystic fibrosis, sepsis) and 3) blunt abdominal trauma. (Pediatric Surgery 6th edtion), (JAMA Pediatr. 2016), (Hebra A. et al, 2016).

In cases of recurrent pancreatitis in children without any obvious cause, with early onset and in families with multiple affected individuals, hereditary pancreatitis should be suspected (Solomon S et al, 2012). Since it was first described in 1952, multiple genetic defects that affect the action of digestive enzymes in the pancreas have been implicated. The most common mutations involve the PRSS1, CFTR, SPINK1, and CTRC genes. New mutations in these genes and previously unrecognized mutations in other genes are being discovered due to the increasing use of next-generation genomic sequencing. (Raphael K, Willingham F,2016)(Oracz et al,2016).

Pancreatic pseudocyst in children till recently was treated surgically with enterocystic anastomosis R-n-Y. Indication for surgical management are: I. size, II. symptomatic, III. long-lasting, IV. or when comlications occur. (Almaihan A et al.2018).

Today with the progress of medical equipment and the increased experience in children, endoscopic transgastric drainage of pancreatic pseudocysts, though it's slow going seems to gain ground. (Scheers I et al.2015). The first case of pediatric pseudocyst endoscopic drainage was reported by Wiersena and associates in 1996 (Evagelista M. 2017). In cases with PPCs, endoscopic transgastric drainage, may be the most indicated treatment because is less interventional approach and can be repeated in cases of recurrent pseudocyst or creation of new ones (Yi Jia et al, 2015).

Case Report

A six years old boy was referred to our clinic with a large pancreatic pseudocyst, 8cm in diameter. The boy had ARPs at least 3 episodes, which were treated conservatively. These episodes started at the age of 4 and till then was a healthy boy.

During his control check up wih abdominal US, a large pancreatic cyst was revealed, and further examination with MRCP took place (Figures 1 and 2).

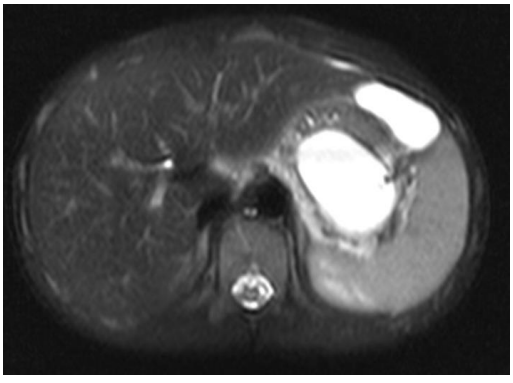

Figure 1

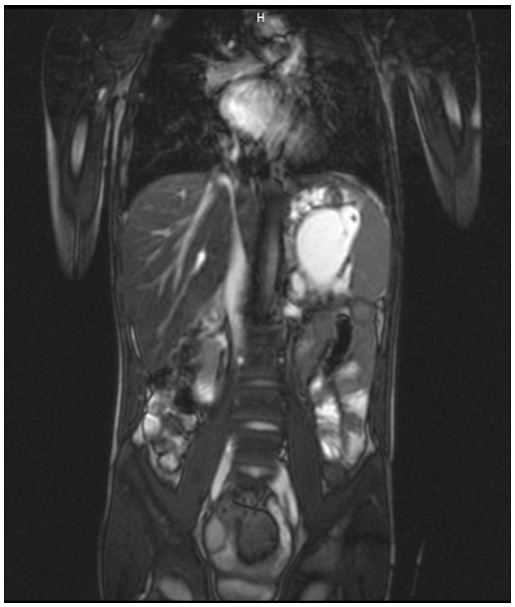

Figure 2

A genetic examination was performed and the results were negative for Cystic Fibrosis (CF) mutation (99% of the recorded mutations). A cysteo-jejunum R-n-Y anastomosis was performed (Figure 3). The patient was discharged from the clinic 13 days after the operation.

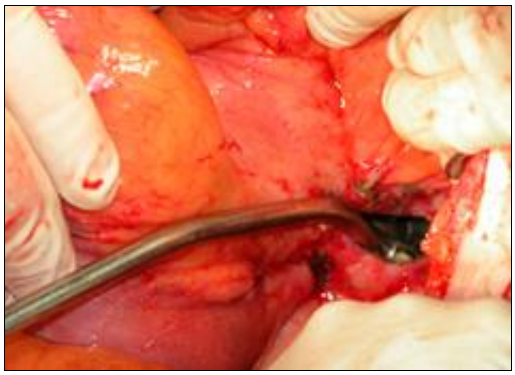

Figure 3

Three months after surgery, he had several episodes of pancreatitis, which were also treated conservativelly and his abdominal US was without any pathological finding. From his family a new information revealed, that a 2nd degree relative was referred to had had also RAP from childhood. Parents were advised to make a further gene control for mutations that are responsible for recurrent pancreatitis, but that option was denied due to social-economical issues.

A year following a new episode, dilatation of the pancreatic duct (5.55mm) was detected and 3 years later and many RAPs, a new pancreatic pseudocyst developed (Figures 4 and 5).

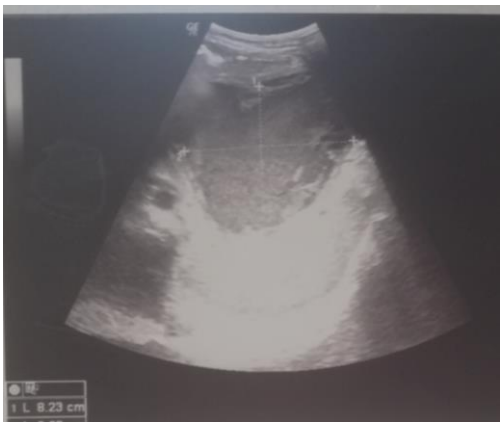

Figure 4

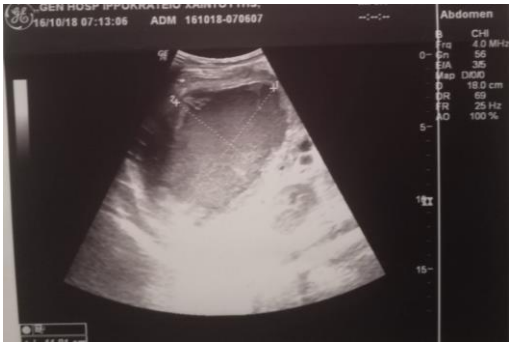

Figure 5

The MRCP findings were that the cyst was located at the body and tail of the pancreas and protruded to the posterior wall of the stomach.

After thorough discussion with the gastroenterologists a genetic aetiology was suspected and an endoscopic transgastric drainage of the pseudocyst was performed, as a less invasive and repeatable method. (Figures 6-9)

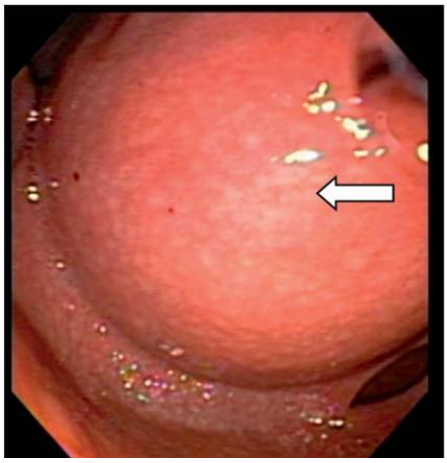

Figure 6 : During the gastroscopy, see the bulging on the gastric wall caused by the pseudocyst

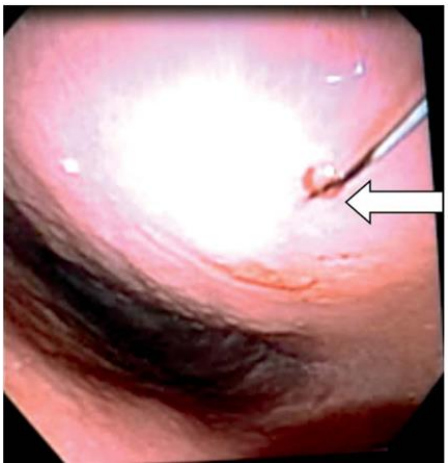

Figure 7: Through a small incision, a guidewire is passed after cyst puncture and balloon dilatation of the tract

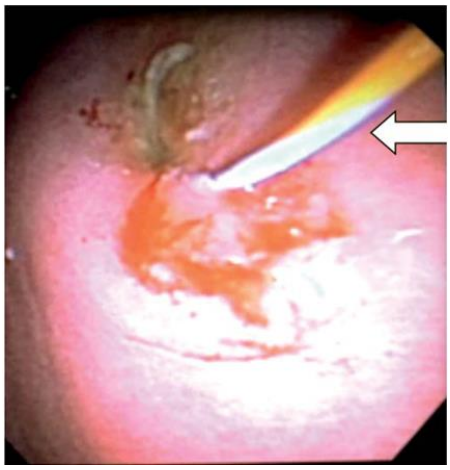

Figure 8

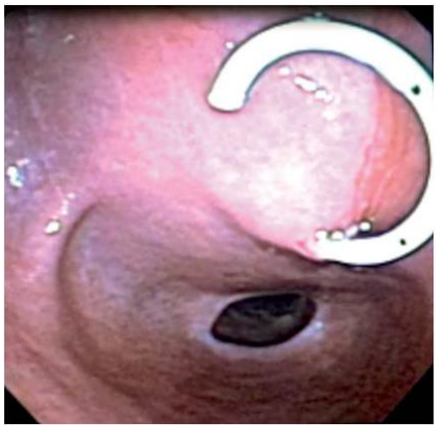

Figure 9: A pigtail stent was placed to drain the cyst

Ten days after, the abdominal distension insisted, and an abdominal x-ray showed that the pigtail had been replaced (Figure 10).

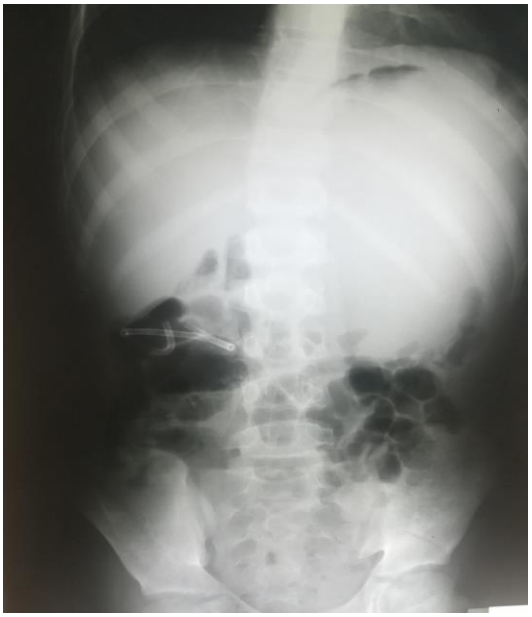

Figure 10

Another stent was placed endoscopically and was left there for 6 weeks. (Figure 11)

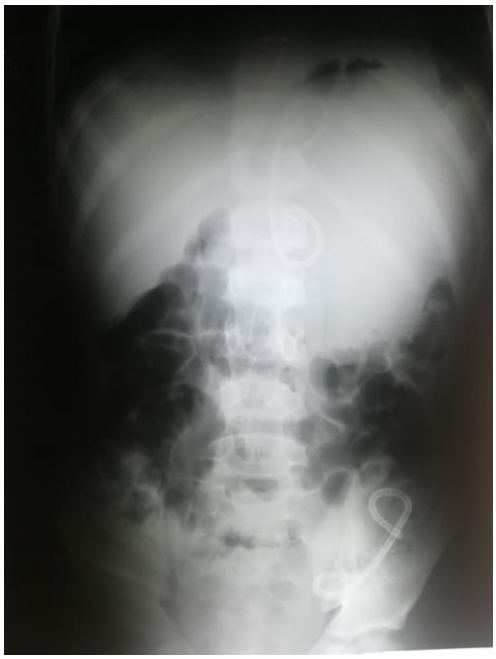

Figure 11

Till now the patient is without any new episode of pancreatitis and his blood tests and abdominal US are normal.

Discussion

HP is a rare autosomal dominant genetic disorder of the pancreas and typically presents in childhood with multiple ARP. (Celeste A., Whitcomb D,2016)In cases of ARP, CP or idiopathic pancreatitis, especially if the child is younger than 3 years old, certain hereditary disorders should be included in the diagnostic evaluation including PRSS1 gene mutation, SPINK 1 gene mutation, cystic fibrosis transmembrane regulator gene abnormalities, chymotrypsin C and calcium-sensing receptor genes. Those conditions previously described as idiopathic (Whitcomb D,2019)(Suzuki et al,2014). Patients with PRSS1 or SPINK1 gene mutation are at higher risk of developing pancreatic exocrine insufficiency, diabetes melitus type 1 and pancreatic cancer. (Schmitt F et al,2009).

Pancreatic pseudocysts are rare in pediatric population. (Kumar S,2016) Most PPCs resolve spontaneous, with conservative treatment, especially those smaller than 6 cm. (Chaurasiya O. 2015). The most common causes of pseudocysts in children are trauma and infections (Takeshi Miyano.2006). Other causes are biliary tract diseases and abnormalities (pancreas divisum), medications and toxins, systemic diseases (cystic fibrosis) or it could be idiopathic. (Mitsuyoshi S et al,2014),(Hebra A et al,2016).

Pancreatic pseudocysts are managed with Roux-n-Y cystojejunostomy, which is the most widely used internal drainage procedure for this problem and is associated with the lowest rate of complications and recurrence. External drainage of the pseudocysts carries a higher risk for complications such as fistula formation and a higher, recurrence rate than internal drainage does. (Takeshi Miyano,2006).

In recent years endoscopic transgastric drainage of pancreatic pseudocysts and especially the ultra sound-guided, seems to be considered the most preferable choice for children too. (Scheers I et al,2015).

A systematic review that compared the drainage of pancreatic pseudocysts by endoscopic, percutaneous and surgical routes concluded that endoscopic and surgical drainage are equally effective, but the endoscopic drainage requires shorter hospital stays and lower costs and results in better quality of life (Párniczky et al, 2018). Surgical or percutaneous drainage should be considered in patients who have anatomical reasons for avoiding endoscopy (Premal A. Patel et al,2019), (Luis Augusto Zarate et al,2018).

In our case we managed the pancreatic pseudocyst surgically because of the large size of the cyst and not knowing that a heredity was hidden. After the recurrency of the cyst and through further inspection, a hereditary background was revealed and with the contribution of the gastroenterologists, endoscopic transgastric drainage was performed, which was successful.

Conclusion

In children with ARP, CP and idiopathic pancreatitis a thorough check for gene mutations should be performed and if PPCs presents we should consider to perform endoscopic drainage of the cysts, as a better way of treatment. As the specific forms of HP are more precisely identified and our understanding of their particular manifestations grows, it is hoped that we can offer more timely triage to the best care for each patient.

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