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ORIGINAL ARTICLE

How to differentiate congenital pancreatic cysts and cystic pancreatic teratomas?

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Abstract

Purpose The differential diagnosis of congenital pancreatic cysts and mature cystic pancreatic teratomas is challenging due to the lack of reliable diagnostic tests or pathognomonic imaging findings. This study highlights the differences between these extremely rare entities with few published case reports.

Methods Based on our recent experience with two cases, we conducted an extensive review of literature including all reported cases of congenital pancreatic cysts and cystic pancreatic teratomas in children.

Results We report two cases of large cystic abdominal mass detected by prenatal ultrasonography. Both neonates were asymptomatic after birth and were operated on by laparoscopy at the age of 1 year and 3 months, respectively. Histopathology demonstrated a congenital pancreatic cyst in the first case and a mature cystic teratoma in the second case. The postoperative course was uneventful for both children. We describe the clinical and imaging data of the 33 congenital pancreatic cysts and 10 mature cystic teratomas of the pancreas previously reported in the literature, and discuss diagnosis criteria and treatment strategies.

Conclusion Congenital pancreatic cysts and pancreatic teratomas are rare lesions in children. Clinical presentation is not specific and these lesions are often asymptomatic. Although associated malformations are evocative of congenital pancreatic cyst, the final diagnosis can only be confirmed by histopathological examination. Available treatments include ultrasound-guided puncture, enucleation, distal pancreatectomy, cystogastrostomy, and cysto-jejunostomy by the Roux-en-Y technique. Laparoscopic excision can be performed shortly after birth and is our preferred option.

Keywords Congenital pancreatic cyst · Pancreatic cystic teratoma · Dermoid cyst · Prenatal diagnosis · Laparoscopy

Introduction

Dermoid cysts of the pancreas, also called mature cystic teratomas, are benign extragonadal germ cell cysts derived from all three germinal layers. They are lined by stratified squamous epithelium and usually contain a variety of tissues. On the other hand, congenital pancreatic cysts are lesions lined by a cuboidal epithelium with a deeper layer of acinar tissue. Both lesions are extremely rare entities with few published case reports. Prenatal diagnosis is challenging due to the lack of reliable diagnostic tests or pathognomonic imaging findings. Common differential diagnoses include other cystic lesions of the upper abdomen such as intestinal duplication and choledocal cyst. The largest pancreatic cysts may be confused with choledocal, mesenteric, adrenal, omental, hepatic, splenic, renal, ovarian, or urachal cysts [1].

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The diagnosis of abdominal cystic pulmonary sequestration can also be discussed.

The exact etiology of congenital pancreatic cyst is unclear. They may occur as a result of developmental anomalies of the pancreatic ductal system and result from sequestration of primitive pancreatic ducts [2]. Cysts that develop secondarily to obstruction of the ductal system are sometimes called retention cysts [2, 3]. Pancreatic cystic dysplasia can also be encountered in a variety of congenital syndromes including von Hippel–Lindau and Ivemark II syndromes [4–8]. In this case, the pancreas usually contains multiple cystic lesions associated with other congenital malformations. Mature cystic teratoma is a congenital developmental abnormality of germ cell origin that may be derived from any of the three germinal layers (ectoderm, mesoderm, and endoderm). These tumors are commonly detected in ovary, but can also occur in testes, cranium, mediastinum, retroperitoneum, and sacrococcygeal regions [9]. Pancreas is the rarest site of presentation and very few cases of pancreatic dermoid cysts were reported in children [10–12].

We report two cases of large cystic abdominal lesions detected by prenatal ultrasonography. Both children were operated on by laparoscopy with uneventful follow-up. Histopathology demonstrated a congenital pancreatic cyst in the first case and a mature cystic teratoma in the second case. We conducted a systematic literature review and identified 33 congenital pancreatic cysts reported in children and 11 pancreatic dermoid cysts. Diagnosis criteria and treatment strategies are discussed.

Case 1

A 44-year-old women gravida 3 para 2 was referred at 22 weeks of gestation, because a 12 mm abdominal cyst was detected by prenatal ultrasonography. This cyst was located below the stomach, in contact with the inferior vena cava. It was regular and delimited by a thin wall. No other anomaly was detected. Follow-up ultrasounds demonstrated normal fetal growth. A male neonate was born at 38 weeks of gestation by vaginal delivery, weighting 3980 g. He was asymptomatic and we underwent clinical and radiological follow-up. Postnatal ultrasonography showed a progressive growth of the cyst with a diameter of 19 mm after birth and 36 mm 3 months later. Its size remained stable afterward. An MRI was performed at the age of 3 months. The cyst showed high signal intensity on T2-weighted images. There was no biliary dilatation and the gallbladder was normal. The boy remained asymptomatic and the mass was not palpable. Routine blood biochemistry profiles showed an increase of lipase at 127 U/L at 6 months. Surgical treatment was undergone by laparoscopy at the age of 1 year. The cyst was originating from the back of the pancreatic head and was approached by Kocher maneuver. A few mL of clear fluid were aspirated from the cyst, containing 259,160 U/L lipase. A large posterior portion of the cyst wall was excised with the whole mucosa. The post-operative period was uneventful. The boy was discharged 2 days after surgery. Histopathologic evaluation revealed the diagnosis of congenital pancreatic cyst, as the cyst wall was lined by a single layer of cuboidal epithelium. The patient was totally asymptomatic with no sign of recurrence on abdominal ultrasonography after 2 months of follow-up.

Case 2

A 30-year-old woman gravida 1 para 2 was referred at 20 weeks of gestation, because prenatal ultrasonography showed a double bubble sign. A duodenal atresia was evocated. No other malformation was seen and karyotype was 46, XX. The end of pregnancy was uneventful and a female neonate was born by vaginal delivery at 38 weeks of gestation weighing 3120 g. She was completely asymptomatic and there was no palpable abdominal mass. No jaundice or stool color changes were observed. Plain abdominal X-ray excluded the diagnosis of duodenal atresia. Ultrasonography confirmed the persistence of the cystic abdominal mass and showed a well-defined wall without calcifications. It was 3 cm diameter, located in the right hypochondria in contact with the hepatic hilum. As the patient was completely asymptomatic, she was discharged and MRI was programmed 3 weeks later. It revealed the persistence of the cystic mass measuring $35 \times 16 \times 22$ mm, located in the retroperitoneal area just below the hepatic hilum and in front of the inferior vena cava. Diagnosis of duodenal duplication was suspected. Although unlikely, the diagnosis of choledochal cyst was also mentioned. Surgery was performed at 3 months of age by laparoscopic approach (Fig. 1). The retroperitoneal mass was in direct contact with the posterior wall of the pancreatic head. The cyst was opened and contained a whitish sebaceous secretion with debris. The mucosa was completely resected with the extrapancreatic part of the cyst wall. The patient was discharged 3 day post-operative after removal of the abdominal drainage. Lipase level remained normal in the post-operative period. Histologic examination revealed a cyst lined by stratified squamous epithelium with overlying lamellar keratins. The underlying connective tissues were composed of collagen fibers containing multiple tissues (cartilage, lymphoid nodules, pancreatic tissue, colonic epithelium, and scattered neural tissue), suggesting a mature teratoma. No immature foci were identified. At 33 months, the girl was doing well and alpha-fetoprotein (AFP) and US were normal.

To review the literature on these topics, articles on true congenital pancreatic cysts and pancreatic teratomas

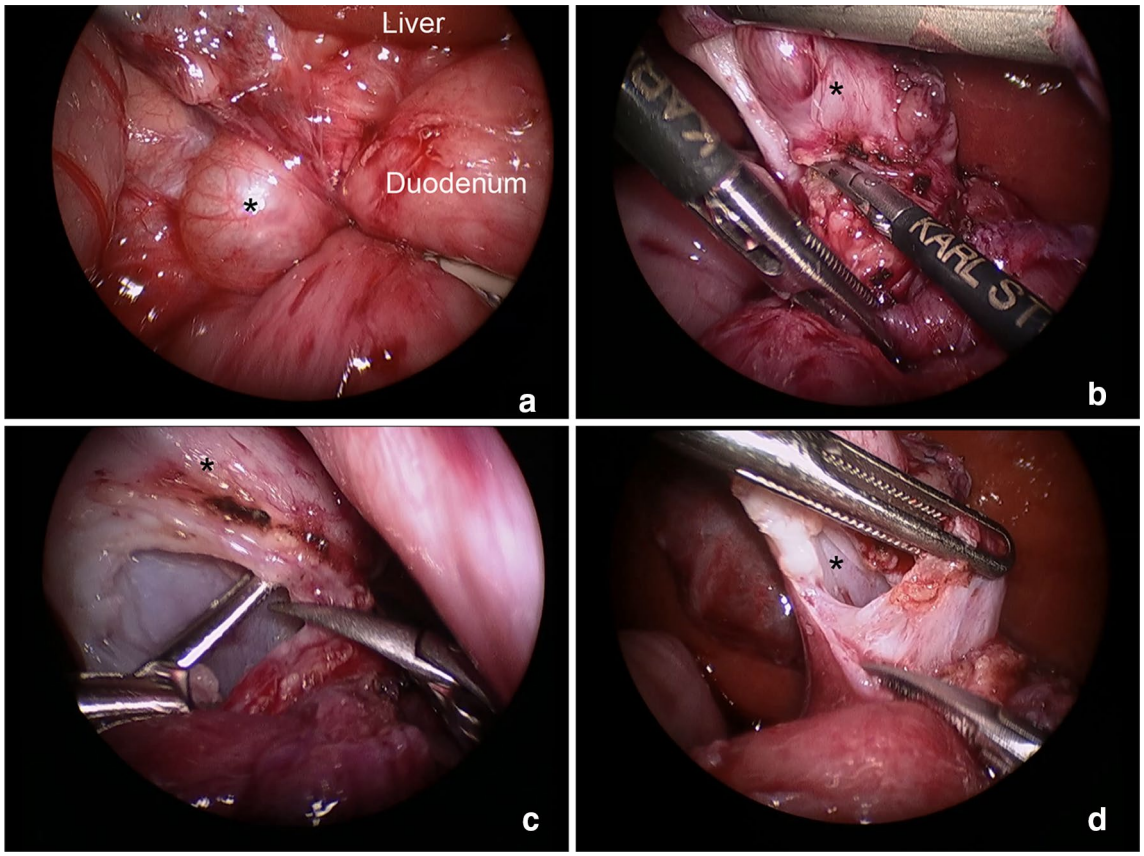

Fig. 1 Laparoscopic excision of a mature pancreatic teratoma (asterisk) in a 3-month-old girl. **a** Aspect of the teratoma located behind the duodenum, **b** opening of the cystic teratoma, **c** resection of the extrapancreatic part of the cyst wall, **d** resection of the mucosa

published in English language were identified through a MEDLINE computerized search. Additional sources were identified through cross-referencing.

Clinical, imaging findings, treatment, and outcomes of 33 other congenital pancreatic cysts reported in the literature are summarized in Table 1 [1, 2, 4, 7, 13–36]. Eleven of them were diagnosed prenatally [1, 13–20, 36]. Clinical, imaging findings, treatment, and outcomes of 11 pancreatic teratomas in children are summarized in Table 2 [12, 37–45].

Discussion

Congenital pancreatic cysts and teratoma are extremely rare diagnoses in children. Including the two present cases, we identified 34 cases of congenital pancreatic cysts and 11 cases of pancreatic teratomas reported in the pediatric population. 11 congenital pancreatic cysts were detected by prenatal ultrasonography. As illustrated by case no. 2, a large abdominal cyst can also correspond to a pancreatic teratoma. Differential diagnosis between these two entities is challenging, as no clinical or radiological sign is pathognomonic.

Congenital pancreatic cysts are revealed by the presence of a large abdominal cyst in the upper part of the abdomen, with no identified parenchymal tissue or calcification. The cyst wall is usually thin, but can be difficult to differentiate from the duodenum wall, making pancreatic cyst difficult to differentiate from enteric cysts. On the other hand, pancreatic teratomas often contain calcifications considered as pathognomonic. The presence of fat/fluid levels, or air/fluid level is another precious sign. However, pancreatic teratoma can appear exclusively cystic on ultrasound and, therefore, be difficult to differentiate from a congenital pancreatic cyst.

Differential diagnoses include duodenal duplication, duodenal atresia, lymphangioma, mesenteric, adrenal, omental, hepatic, splenic, renal, ovarian, and urachal cysts. It may be difficult to localize the site of origin of largest cysts. Ultrasonography must, therefore, detail which intra-abdominal organs are in contact with the cyst. The biliary tract must be described to eliminate the diagnosis of choledocal cyst. Abdominal computed tomography and magnetic resonance imaging can also provide additional information regarding the aspect and localization of the cyst, which is important in planning the operative procedure.

Table 1 Clinical, imaging findings, treatment, and outcomes of 34 cases of congenital pancreatic cysts

References	Age at diagnosis	Gender	Associated malformation	Symptoms	Maximum diameter	Location	Age at time of treatment	Treatment	Follow-up
McPherson [21]	6 months	Male	None	Abdominal distension and vomiting	16 cm	Pancreas body	6 months	Laparotomy, distal pancreatectomy	Uneventful after 2 weeks
Miles [22]	2 days	Male	None	Abdominal distension	23 cm	Pancreas head and body	8 days	Laparotomy, complete resection	Uneventful after 3 years
Pilot 1964 [23]	NA	NA	NA	NA	NA	NA	NA	NA	NA
Gundersen [24]	16 months	Male	None	Abdominal distension	35 cm	Pancreas head and body	16 months	Laparotomy, gastrocystostomy	Recurrence 6 months later, treated by laparotomy and complete resection
Olurin [25]	10 months	Female	None	Abdominal distension and vomiting	NA	NA	NA	Laparotomy, complete resection	NA
Olurin [25]	3 years	Male	None	Abdominal distension and vomiting	NA	NA	NA	Laparotomy, gastrocystostomy	NA
Mares [26]	20 months	Female	None	Abdominal distension	NA	Pancreas head, multilocular	20 months	Laparostomy, cystoduodenostomy	Uneventfull after 18 months
Baker [13]	Antenatal	Female	None	None	3 cm	Pancreas tail	2 months	Laparotomy, distal pancreatectomy	Uneventful after 17 months
Fleet [14]	Antenatal	Male	None	None	5 cm	Pancreas body	4 days	Laparotomy, cystojejunostomy by the Roux-en-Y technique	Uneventful after 2 weeks
Auringer [27]	4 months	Female	None	None	7 cm	Pancreas tail	4 months	Laparotomy, complete resection	NR
Shieh [28]	14 years	Female	None	Pancreatitis	6.1 cm	Pancreas tail	14 years	Laparotomy, distal pancreatectomy	Uneventful
Daher [15]	Antenatal second trimester	Female	None	None	5.5 cm	Pancreas tail	Neonatal	Laparotomy, distal pancreatectomy	Uneventful after 1 week
Fremond [16]	Antenatal, second trimester	male	Omphalocele, Beckwith–Wiedemann syndrome	None	7.5 cm	Pancreas head	7 months	Laparotomy, cystojejunostomy by the Roux-en-Y technique	Uneventful after 2 years,
Agarwala [29]	6 years	NA	NA	NA	NA	NA	NA	NA	NA
Kebapci [17]	Antenatal, second trimester	Female	None	Abdominal distension	13 cm	Pancreas tail	7 days	Laparotomy, distal pancreatectomy	Uneventfull, 5 days

Table 1 (continued)

References	Age at diagnosis	Gender	Associated malformation	Symptoms	Maximum diameter	Location	Age at time of treatment	Treatment	Follow-up
Sepulveda [36]	Antenatal, second trimester	Male	Right hemihypertrophy	Abdominal distension	12 cm	Pancreas head	8 days	Antenatal drainage at 30 weeks of gestation, laparotomy, complete resection	Metabolic and hepatic dysfunction after surgery, asymptomatic at 1 year of age
Liao [18]	Antenatal 33 weeks of gestation	NA	NA	NA	NA	Pancreas tail	10 days	NA	NA
Boulanger [4]	11 years	Female	None	None	NA	Pancreas head	11 years	Laparoscopy converted to laparotomy, resection of the mucosa and part of the cyst wall	NA
Matta [30]	4 months	Female	None	Abdominal distension, vomiting	13.5 cm	Pancreas head and body	4 months	Laparotomy, cystojejunostomy by the Roux-en-Y technique.	Uneventful after 15 months
Kazez [31]	4 months	Male	None	Abdominal distension	9.5 cm	Pancreas tail	NA	Laparotomy, distal pancreatectomy	Uneventful after 6 months
Chung [19]	Antenatal 22 weeks of gestation	Female	None	Abdominal distension	5.8 cm	NA	3 days	Laparotomy, complete resection	Uneventfull after 7 days
Choi [48]	Antenatal	Female	None	None	5.6 cm	Pancreas tail	Neonatal	Laparotomy, distal pancreatectomy	Uneventful
Boulanger [32]	7 weeks	NA	None	Abdominal distension	NA	Pancreas head and body	7 weeks	Laparotomy, complete resection	Uneventful after 16 months
Castellani [20]	Antenatal	Female	None	Abdominal distension, pain, and anorexia	NA	Pancreas body	10 days	Two ultrasound-guided punctures, then laparotomy for cystojejunostomy by the Roux-en-Y technique	Uneventful after 30 days
Gerscovich [1]	Antenatal 26 weeks of gestation	Female	None	Abdominal distension	10 cm	Pancreas tail	11 days	Laparotomy	NA
Chahed [7]	1 month	Male	Ivemark II syndrome with situs inversus, asplenia, heart disease, dysplasic liver	Abdominal distension and vomiting	7 cm	NA	1 month	Laparotomy, cystoduodenostomy	Died of heart failure at two and a half months of age
Al Salem [33]	18 months	Male	None	Abdominal distension	9 cm	Pancreas body	18 months	Laparotomy, complete resection	Uneventful after 8 days

Table 1 (continued)

References	Age at diagnosis	Gender	Associated malformation	Symptoms	Maximum diameter	Location	Age at time of treatment	Treatment	Follow-up
Al Salem [33]	4 months	Female	None	Abdominal distension and vomiting	12 cm	Pancreas head and body	4 months	Laparotomy, cysto-jejunostomy by the Roux-en-Y technique.	Uneventful after 2 weeks
Nasher [34]	18 months	Male	None	Abdominal distension	NA	NA	18 months	Laparotomy, complete resection	NA
Warnock [35]	5 weeks	Female	None	NA	NA	NA	NA	Ultrasound-guided punctures, laparoscopic resection, and a second surgery after relapse	NA
Bawazir [2]	4 months	Female	None	Abdominal distension and vomiting	12 cm	Pancreas head and body	4 months	Laparotomy, cysto-jejunostomy by the Roux-en-Y technique.	Uneventful after 2 weeks
Bawazir [2]	16 months	Male	None	Abdominal distension	9 cm	Pancreas body	16 months	Laparotomy, complete resection	Uneventful after 8 days
Bawazir [2]	4 years	Female	None	Abdominal distension	NA	Pancreas head	4 years	Laparotomy for a Wilms tumor, cysto-jejunostomy during the same procedure	NA
Present case no. 1	Antenatal	Male	None	None	3.6 cm	Pancreas head	1 year	Laparoscopic enucleation	Uneventful after 2 months

NA not available

Table 2 Clinical, imaging findings, treatment, and outcomes of 11 cases of pancreatic teratomas in children

References	Age at diagnosis	Gender	Associated malformation	Symptoms	Maximum diameter	Location	Age at time of treatment	Treatment	Follow-up
DeCourcy [37]	2 years	Female	None	Vomiting epigastric mass	5 cm	Pancreas body	2 years	Laparotomy, complete resection	NA
Bittner [38]	2 years	Female	None	Liver failure epigastric mass	NA	Pancreas head	2 years	Laparotomy, complete resection	Uneventful after 6 months
Iovchev [39]	8 years	Male	None	Vomiting fever abdominal pain abdominal mass	NA	Pancreas body	8 years	External drainage	Uneventful after 7 months
Pomosov [40]	6 years	Male	None	Vomiting fever abdominal pain abdominal mass	NA	Pancreas tail	6 years	Laparotomy, distal pancreatectomy	Uneventful after 6 months
Komarov [41]	4 years	Female	None	Vomiting pain epigastric mass	NA	NA	4 years	External drainage	NA
Assawamatiyanont [42]	11 years	Female	None	Abdominal mass	9 cm	Pancreas body	11 years	Laparotomy, complete resection	Uneventful
Das [12]	4 months	Female	None	Abdominal mass	10 cm	Pancreas body and tail	4 months	Laparotomy, complete resection	Uneventful
Yu [43]	2 years	Male	None	Epigastric mass	11 cm	Pancreas head	2 years	Laparotomy, complete resection	NA
Kela [44]	5 months	Male	None	Abdominal pain central mass	8 cm	Pancreas tail	5 months	Laparotomy, complete resection	Immature teratoma Chemotherapy Lost to follow-up after the 1st chemotherapy
Wang [45]	12 months	Female	None	Vomiting Central mass	18 cm	Pancreas body	12 months	Laparotomy, complete resection	Recurrence 19 months after Distal pancreatectomy Well 2 years later
Present case no. 2	Antenatal 20 weeks of gestation	Female	None	Prenatal diagnosis no palpable mass	3.5 cm	Pancreas head	3 months	Laparoscopy, complete resection	Uneventful after 30 months

NA not available

Congenital pancreatic cyst can also be associated with other congenital malformations. Two cases were associated with Beckwith–Wiedemann syndrome [16] and hemihypertrophy [36]. A few cases of pancreatic dysplasia associated with von Hippel–Lindau and Ivemark II syndrome were reported [6–8]. In 1979, Hopper et al. reported a case of pancreatic dysplasia associated with asphyxiating thoracic dysplasia and short limbs. The infant survived for only 9 h [46]. Balci et al. also reported three cases of sib fetuses with situs inversus totalis, renal and pancreatic dysplasia, bowing of the lower limbs and clavicles, severe

intrauterine growth retardation, and oligohydramnios [5]. Pregnancy was terminated in all cases.

We did not identify any case of pancreatic teratoma associated with congenital malformations.

After birth, small pancreatic cysts are usually asymptomatic. In the present case (case no. 1), the cyst size increased throughout the first year of life, while the patient remained asymptomatic. Abdominal distension and vomiting seems to appear when the diameter of the cyst is over 6 cm in infants.

There is no reliable biological marker that can help for the diagnosis of congenital pancreatic cyst or pancreatic teratoma. Immature teratoma can be associated with elevated α-fetoprotein levels, but this is not reliable in infancy. Moreover, Kela et al. reported the only case of immature teratoma in a 5-month-old child and α-fetoprotein level was normal [44]. Conversely, Asai et al. presented a case of retroperitoneal immature teratoma diagnosed at 33 weeks of gestation, with elevated postnatal α-fetoprotein level [47]. Amylase and lipase levels in serum are usually normal, unless the pancreatic duct is obstructed by the cyst [28, 41]. The fluid of congenital pancreatic cyst can contain extremely high levels of pancreatic enzymes [2, 20], as in the present case no. 1, although it was not communicating with the pancreatic duct. Congenital pancreatic cyst containing normal levels of amylase and lipase was also reported [15].

Surgical treatment of pancreatic cyst is usually favored, although there is no consensus regarding the timing of treatment. In the first case, we chose to wait the age of 1 year, as the infant was asymptomatic and to facilitate the laparoscopic approach. The progressive increase of the cyst diameter suggests that congenital pancreatic cyst should be treated before obstruction of the biliary and pancreatic ducts. Surgical removal is also the standard treatment of dermoid cysts. Different treatment strategies were reported. Ultrasound-guided puncture of the cyst has been tempted, even prenatally, but was not sufficient [20, 36]. Complete resection or enucleation should be performed whenever possible, with maximum preservation of the pancreatic tissue [2, 4, 12, 19, 22, 25, 27, 32, 33, 35–37, 42–45]. Distal pancreatectomy is possible when the cyst is located into the pancreatic tail [13, 15, 17, 21, 28, 31, 40, 48]. When the cyst is in the pancreatic head, complete removal is not always possible. The less invasive option in this situation is to perform a gastrocystostomy or duodenocystostomy [2, 7, 24–26]. Alternatively, other surgeons choose to perform a cysto-jejunostomy by the Roux-en-Y technique with good results [2, 3, 14, 16, 20, 30, 33]. The present cases show that laparoscopic removal is a viable treatment option for congenital pancreatic cysts and pancreatic teratomas in infants, with minimal post-operative pain, minimal scaring, and short hospital stay [4, 35].

The final diagnosis can only be confirmed by histopathological examination. True pancreatic cysts contain epithelial tissue not seen in pseudo-pancreatic cyst developing after trauma or infection [27]. These cysts are lined by a single layer of cuboidal epithelium with a deeper layer of acinar tissue [3]. Cystic teratomas are lined by stratified squamous epithelium and connective tissues composed of collagen fibers, and containing multiple tissues from more than one of the three primitive germ cell layers. They usually contain a combination of cystic and solid elements, including teeth, hair, cartilage and dermal appendages such as hair follicles, sweat glands, and abundant sebaceous material. One case of immature dermoid cyst of the pancreas was reported, containing microfoci of immature tissue [44].

We recommend long-term clinical and radiological follow-up, with dosage of α-fetoprotein levels in case of pancreatic teratoma.

Conclusion

Congenital pancreatic cysts and pancreatic teratomas are rare lesions in children. They can be diagnosed by prenatal ultrasonography. Although pancreatic teratomas often contain calcifications seen on imaging studies, they can be difficult to differentiate from congenital cysts. Clinical presentation is not specific and these lesions are often asymptomatic. Preoperative evaluation and detailed imaging are the corner stone for surgical planning. Available treatments include ultrasound-guided puncture, enucleation, distal pancreatectomy, cystogastrostomy, and cysto-jejunostomy by the Roux-en-Y technique. Laparoscopic excision can be performed shortly after birth and is the best option in our opinion.

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Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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