Cystic teratoma of the pancreas

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Abstract
Cystic teratoma also called dermoid cyst, are congenital benign germinal tumors most commonly found in the ovaries and testes. Pancreatic localization is the least common site of presentation, which make its preoperative diagnosis even more challenging. Herein, we report the case of an 8-year-old girl who presented with abdominal pain localized in the epigastrum. The CT scan showed a well-defined heterogenous cystic mass in the head of the pancreas. At surgery, the entire mass was excised. The diagnosis of a dermoid cyst was then confirmed macroscopically and by histopathology. The main goal of our case report is to show the rarity of this entity and to make a quick review of the literature.

Key words: Dermoid; Pancreatic Cyst; Teratoma; CT.

1. Introduction
Mature cystic teratomas are congenital benign germinal tumors that may arise from any of the three germinal layers. They are heterogenous tumors with ectodermal components (hair follicles, sebaceous glands, skin) and can arise anywhere along the path of germ cell migration during embryogenesis. The most frequent location are the ovaries and testes, the pancreas remains a rare site of development. Thus, the preoperative diagnosis is challenging due to its rarity and the inadequate diagnostic tests.

We report a case of 8-year-old girl, representing the 52nd case reported in the literature (8).

2. Case report
An 8-year-old girl, with no personal nor family history, was admitted for vague abdominal pain 20 days prior to evaluation. There were no other symptoms associated. The physical examination, showed discrete epigastric tenderness however no abdominal masses were palpable. At first blood tests were done, including a complete blood count (CBC), liver function tests, turned out to be within normal limits while the lipase level was 10 times more than the normal limit. At first the diagnosis of pancreatitis was suspected; hence an abdominal CT scan was done showing a heterogenous tumor in the head of the pancreas with 2 components cystic and fat, it was well circumscribed with wall calcification. Due to its volume and anatomical location, it exerted a mass effect on the portal vein causing a development of multiple venous collaterals (Figure 1). However, the tumor did not invade any of the adjacent structures.

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Due to the resectability of the mass, the decision of a surgical excision was made after the consent of the girl’s parents. The surgeons proceeded with a midline laparotomy that showed no abnormalities in the abdominal cavity. Then lesser sac was dissected, the stomach retracted superiorly, displaying the head of the pancreas, with its palpable white-tan mass. (figure 2) After the tumor removal there were tufts of hair in the bottom of the cystic mass (figure 3). There were no complications during the operation.
However, the clinical follow up of the patient were marked by a cholestatic jaundice with a disturbance in the hepatic tests. Then a CT scan showed a dilation of the intrahepatic and common bile duct with no further stenosis.

![CT scan](image1)

**Figure 4** Abdominal CT scan after contrast injection, showing: The dilation of the intrahepatic bile duct (star (A)) as well as the common bile duct (arrow (B)).

Hence, a Magnetic resonance cholangiopancreatography (MRCP) was performed showing an extensive stenosis of the lower bile duct, with no fluid collection causing a dilatation of the bile duct upstream (Figure 5).

![MRCP](image2)

**Figure 5** Abdominal MRI showing an extensive stenosis (star) of the lower bile duct, causing a dilatation of the bile duct upstream (arrow).

Afterwards, an endoscopic retrograde cholangiopancreatography (ERCP) confirmed the distal stenosis of 10 mm. Thus, the girl was treated by a placement of an endoprosthesis in the external sphincter after a sphincterectomy.

We then made sure of the right positioning of the endoprosthesis by an abdominal ultrasound examination (figure 6).
The post-operative follow up was favorable with a complete disappearance of the jaundice; the girl went home with the decision of a control CT scan one month after the procedure, showing the regression of the dilation after the introduction of the prosthesis, with no tumor recurrence (figure 7).

Figure 7 Abdominal CT scan showing a dilation regression of the bile duct (star) after the introduction of the prosthesis (arrow)

3. Discussion

Teratomas are tumors that are composed of tissue derived from all three germ layers. They are divided into three categories: mature (benign), immature (malignant), and monodermal or highly specialized (5). Most mature teratomas are cystic and mostly known as dermoid cyst. The pancreatic site is the rarest site of presentation, it was first described in 1918 by Kerr [2] and assigned to cystic pancreatic lesions by Primrose in 1922 [6].

Mature cystic teratomas (MCTs) usually occur in a younger age group, with a mean age at diagnosis of 37.7 years (range: 4 months to 74 years old) [9], as found in a study led by Li Zhe of 50 patients. There is a slight male predominance (54% of men vs 46% of women) [9].

The clinical presentation is nonspecific, and can be diagnosed incidentally. Patients might present symptoms like abdominal pain, back pain, anorexia, fatigue, nausea and vomiting [1].

The laboratory tests are unremarkable, they can present with hyperbilirubinemia or an increase of the lipase level explained by the mass effect exerted by the tumor on the biliary tract especially if located in the head of the pancreas. Moreover, the CEA and CA19-9, used for the evaluation of pancreatic cysts are significantly lower in dermoid cysts [1,10].
Given the non-specificity of the clinical or biological findings, the use of imaging techniques can help in the diagnosis. In fact, the presence of fat/fluid or hair/fluid levels is considered pathognomonic of dermoid cysts, and can be seen in CT scans as well as in sonograms.

Ultrasound findings are usually a cystic mass, with distinct margins and without septations. The fatty component appears hypechoic with focal areas of acoustic shadowing, secondary to the presence of calcified tissues. CT on the other hand, will confirm these areas of calcifications and fat and characterize the fluid as sebum, serous or complex. Magnetic resonance imaging (MRI) can also be performed for further characterization, showing low signal intensity on T1-weighted images, areas of fat-fluid level, if present, and distinct margins.

Though, its pathognomonic appearance, its presence in the pancreas remains minor (4), hence its differentiation with other diagnosis becomes even harder. In fact, there are many pancreatic cystic lesions that has each a specific imaging finding distinguishing it from the others including pseudocyst, mucinous and serous cystadenoma, lymphoepithelial cyst, intraductal papillary mucinous neo-plasm (IPMN), and solid pseudopapillary tumor. Lymphoepithelial cysts, most commonly located in the pancreas is the diagnosis resembling most the dermoid cysts, however, the absence of epidermal appendages such as hair follicles or sebaceous glands, makes its difference [11].

Another distinguishing feature is that suppurative infections occur more frequently in dermoid cysts than in other "squamous lined" pancreatic cysts [11].

Due to the ambiguous pre-operative diagnosis, and despite their benign nature, surgical removal remains the gold standard treatment of dermoid cysts of the pancreas in order to exclude malignant or borderline malignant neoplasms. In fact, it was established that more than 70% of excised pancreatic cystic lesions reported were either malignant or premalignant, and based on these findings it is recommended that surgical excision be undertaken for any symptomatic cystic lesion as well as any lesion larger than 2-3 cm in size, particularly in elderly patients (11). Moreover, external drainage and marsupialization have been long criticized due to the risk of the development of chronic fistulas.

4. Conclusion

In conclusion, dermoid cyst of the pancreas is a rare germinal tumor, mostly benign. Its clinical and biological aspects are nonspecific. Imaging can help distinguish cystic teratomas from other neoplasms if the findings are characteristic, if not, the preoperative diagnosis remains challenging and ambiguous. Although the surgical resection is the standard treatment with total surgical excision.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

References


