

Pediatric pancreatic teratoma

A case report and literature review

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Abstract

Rationale: Pediatric pancreatic teratoma (PPT) is a rare tumor with unclear clinicopathologic features and treatment strategy.

Patient concerns: A 13-month-old boy was admitted to the hospital with a complaint of a palpable epigastric mass.

Diagnoses: The lesion was diagnosed as benign mature cyst teratoma via postoperative pathological examination.

Interventions: Pylorus-preserving pancreaticoduodenectomy (PPPD) was performed on the patient. The entire mass was resected from the head of the pancreas and sent to the laboratory for frozen section evaluation.

Outcomes: The patient was followed up for 15 months. He did not undergo recurrence or PPPD-related complications.

Lessons: The differential diagnosis of retroperitoneal occupying lesions among children must consider mature cystic teratomas. Complete surgical resection combined with subsequent postoperative outpatient follow-up remains the primary choice for the management of PPTs.

PPTs are extremely rare tumors with unclear clinicopathologic features and treatment strategy. This study aims to explore the clinical characteristics of and treatment strategy for these tumors. We reported a 13-month-old patient with pancreatic teratoma who underwent pylorus-preserving PPPD. The operation lasted approximately 6 hours. The mass was completely removed, and the patient recovered uneventfully. Complete surgical resection combined with outpatient follow-up is the primary choice for the management of PPTs.

Abbreviations: AFP = alpha fetoprotein, CT = computerized tomography, EUS-FNA = endosonography-guided fine needle aspiration biopsy, MR = magnetic resonance, NSE = enolase, PICU = pediatric intensive care unit, PPPD = pylorus-preserving pancreaticoduodenectomy, QOL = quality of life.

Keywords: pancreaticoduodenectomy, pediatric pancreatic teratoma, surgical resection

1. Introduction

Teratomas are common germ cell tumors among the pediatric population and are composed of tissues originating from all 3 dermal layers: endoderm, mesoderm, and ectoderm.^[1] The most

common sites of these tumors are the sacrococcygeal region, ovaries, and mediastinum. However, teratomas derived from the pancreas are exceedingly rare among the pediatric population, resulting in difficulty in their preoperative diagnosis and treatment.^[2] Herein, we present the case of a pediatric patient with mature teratoma of the pancreatic head and who underwent pancreaticoduodenectomy (PD). We explore the safety and efficacy of PD for pediatric patients with teratomas in the pancreas to further understand this rare tumor.

This study was conducted in accordance with the principles outlined in the Declaration of Helsinki and authorized by the Ethics Committee of the hospital. Written consent was obtained from the parents before operation.

2. Case presentation

A 13-month-old boy was admitted to the institute due to a palpable abdominal mass that was incidentally noted by his mother during child health service. His physical examination was unremarkable, with only a round, hard, elastic, and rigid mass in his upper abdomen without tenderness. The parents did not complain of any discomfort or special family history. A solid cystic mass was revealed by abdominal contrast-enhanced computed tomography (CT) and magnetic resonance (MR) scans at the right side of the mid-upper retroperitoneum (Figs. 1 and 2). The tumor was approximately 9 cm × 6 cm × 5 cm in size and closely related to the pancreatic head. Laboratory data were

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JW and YY have contributed equally to this work.

Institutional review board of West China Hospital approved this case report. A written informed consent was obtained from the patient for publication of this case report and any accompanying images.

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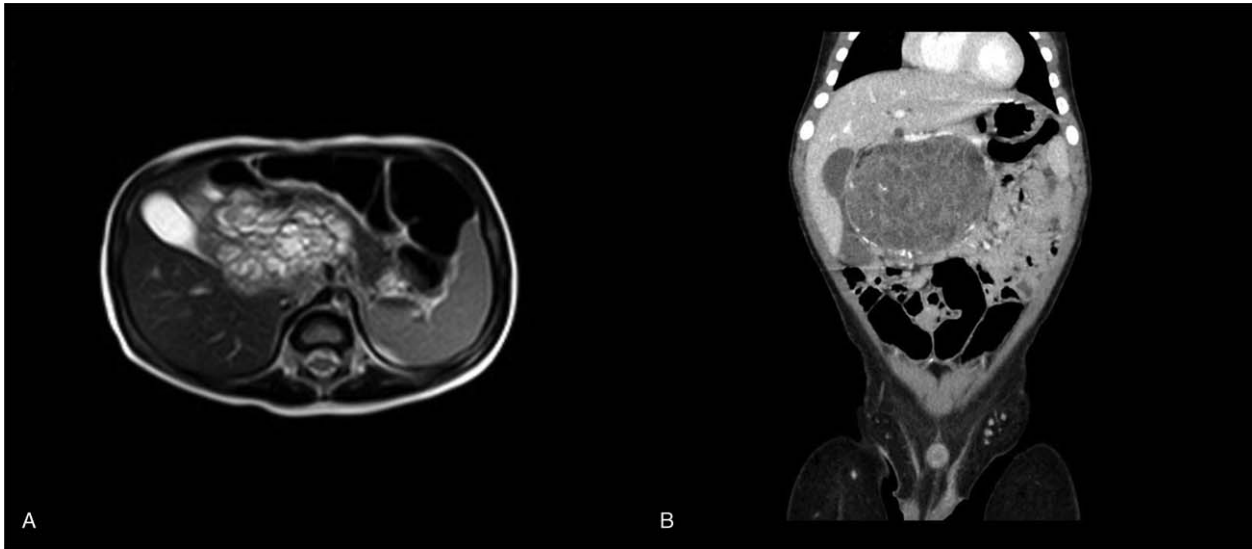


Figure 1. (A, B) Preoperative contrast-enhanced CT scan of the pediatric pancreatic teratoma (A B). The cystic solid mass in the right retroperitoneal region of the mid-upper abdomen is mainly cystic, with honeycomb septation and patchy shadow. The mass is closely related to the head of the pancreas. CT=computed tomography.

as follows: alpha fetoprotein (AFP): 704.60 ng/mL (reference range: 0–8 ng/mL), enolase (NSE): 43.29 ng/mL (reference range: 0–3.4 ng/mL), serum carbohydrate antigen (CA)-199: 86.70 U/mL (reference range: 0–22 U/mL), and normal liver and kidney function tests.

An exploratory laparotomy was performed after preparation prior to surgery. During operation, intraoperative findings displayed a huge mass originating from the pancreatic head with high surface tension and a complete capsule. The duodenum and superior mesenteric vessels were involved, making the separation of the lesion from the surrounding involved tissue difficult. A subsequent pylorus-preserving pancreaticoduodenec-

tomy (PPPD) procedure was ultimately performed on the patient. The entire mass was resected from the head of the pancreas and sent to the laboratory for frozen section evaluation.

In our case, gross pathological evaluation presented a mass that was 12 cm × 9 cm × 8 cm in size. Histologically, the mass contained abundant keratinous debris and components of adipose tissue, cartilage, and hair follicle. The morphological features were consistent with a mature cystic teratoma of the pancreas, a type of benign tumor (Fig. 3). Teratomas are classified in accordance with the classification of the World Health Organization as mature and immature on the basis of morphological characteristics and differentiation degree.

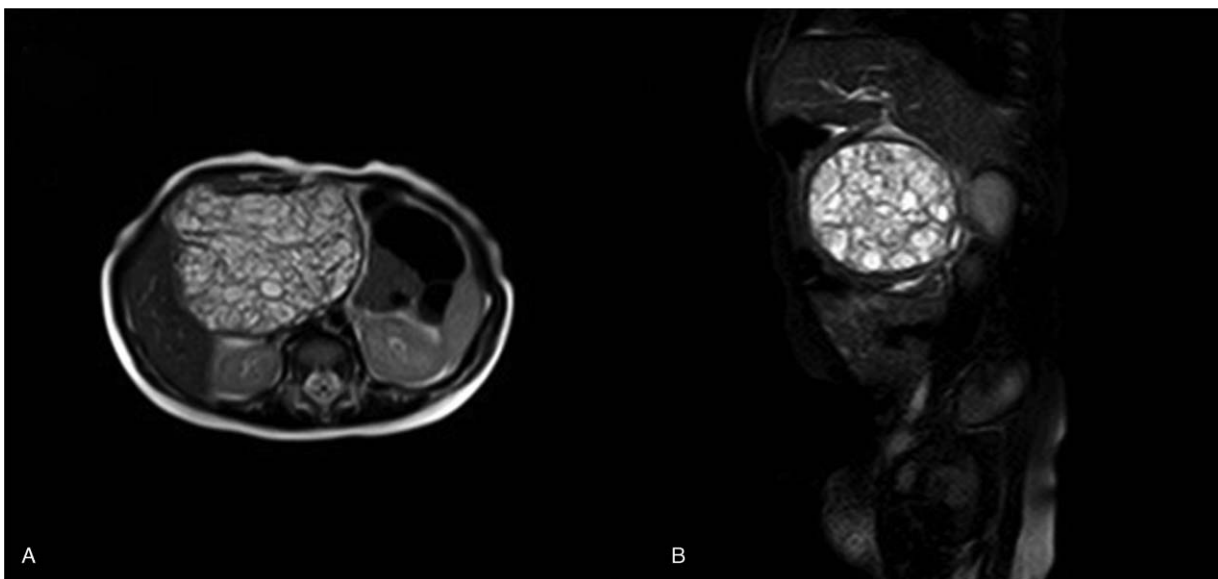


Figure 2. (A, B). Preoperative MRI scan of the pediatric pancreatic teratoma. Tumors contain a large amount of fat and sebum, and the outer membrane is intact. MRI=magnetic resonance imaging.

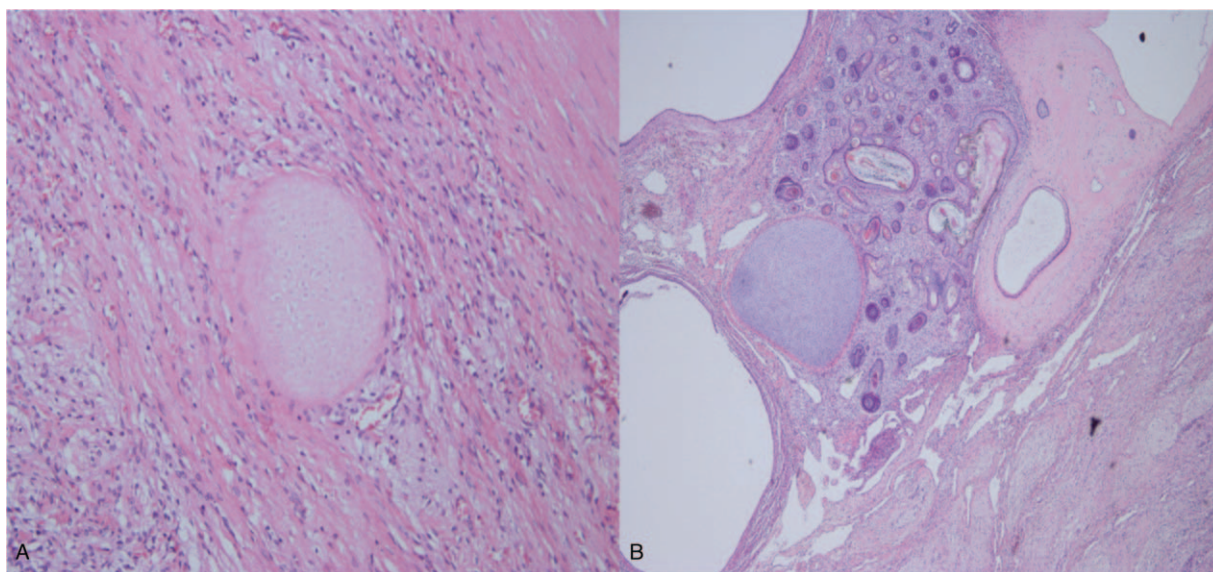


Figure 3. (A, B). Pathological findings of the cystic lesion. A. Hair follicle tissue and cartilage tissue in this pathological section (H&E stain). B. The keratinized squamous epithelium formed the wall of the tumor cyst (H&E stain).

Postoperatively, the patient developed fever and pulmonary infections that required antibiotic treatment of bacteremia but did not experience PPPD-associated mortalities, such as pancreatic leak, delayed gastric emptying, and fat malabsorption. The boy was discharged on postoperative day 22. At 3 months postoperative, the AFP level decreased to normal range, and the patient did not complain of any discomfort.

3. Discussion

Kerr^[3] first described pancreatic teratoma in 1918. To date, 9 pediatric patients with pancreatic teratomas have been reported in the English-language literature, as summarized in Table 1. Clinically, pediatric pancreatic teratoma (PPT) presents with

nonspecific symptoms, and most of these cases manifest as a palpable abdominal mass and/or tenderness.^[11]

The preoperative diagnosis of PPT is difficult. In several cases, the characteristic of fat and calcium combination in the radiologic appearance of magnetic resonance/computerized tomography (MR/CT) is considered highly predictive of mature teratomas. In our case, however, MR and CT only revealed the position and shape of the tumor without this characteristic. Similarly, tumor markers, such as enolase (NSE), carcinoembryonic antigen, and CA-199, lack specificity and sensitivity, except for serum AFP, the level of which is suggestive of disease activity. Endosonography-guided fine needle aspiration biopsy is proposed to help in the preoperative diagnosis of pancreatic tumors; however, this technique is limited by lack of equipment and expertise.^[12]

Table 1
Reported cases on pediatric pancreatic teratomas in the English literature.

Case	Age/Gender	Presentation	Tumor location	Tumor size, cm	Management	Fellow-up/duration
1 ^[4]	11 mos/F	Epigastric painless mass, anorexia, vomiting	Body/Tail	18 × 15 × 10, 16 × 12 × 11 (△)	Cystectomy+Roux-en-Y cystojejunostomy, distal pancreatectomy (△)	Uneventful/2 y
2 ^[5]	12 y/F	Epigastric painless mass	Head (uncus)	7 × 6.5	Enucleation	Reoperation for postoperative SMAS/—
3 (▽)	13 mos/M	Epigastric painless mass	Head	9.2 × 8.5 × 4	PPPD	Uneventful/15 mos
4 ^[6]	4 mos/F	Epigastric painless mass	Body/Tail	9.5 × 8.5 × 8	Resection	Uneventful/—
5 ^[7]	2 y/M	Epigastric painless mass	Head/body	12 × 12 × 12	Resection	NM/—
6 ^[8]	11 y/F	Epigastric painless mass	Body	9 × 8 × 6	Resection	Uneventful/—
7 ^[1]	16 y/M	Progressive jaundice, weight loss weight loss	Head	NM	Resection, choledochoduodenostomy	NM/—
8 ^[9]	2 y/F	Vomiting, epigastric mass	Body	NM	Resection and drainage	Uneventful/—
9 ^[10]	8 y/M	Abdominal pain, vomiting, epigastric mass	Body	NM	External drainage	Uneventful/8 mos
10 ^[11]	5 mos/M	Asymptomatic mass	Head	8 × 7	Deroofing/simple cystectomy	Lost to follow-up/—

— = unavailable; △ = the tumor relapsed at the 18th month of follow-up; ▽ = we reported; F = female; M = male; NM = not mentioned; PPPD = pylorus-preserving pancreaticoduodenectomy; SMAS = superior mesenteric artery syndrome.

Therefore, postoperative pathological evaluation remains the primary diagnostic approach.

With regard to treatment, surgical resection is the mainstay of therapy for PPTs, but detailed surgical guidelines are unavailable. In general, complete surgical resection without damaging vital structures is recommended, while incomplete resection is associated with poor oncological outcome. The operation procedure is determined on the basis of tumor location.^[13] PD is a common procedure in the treatment of numerous benign and malignant pancreatic or periampullary diseases. To our knowledge, PD is a rare procedure among the pediatric population because of the low rate of periampullary diseases in this population and the possibility of growth retardation. Notably, PD may lead to impaired exocrine and pancreatic endocrine functions and gastrointestinal problems. Thus, the postoperative endocrine and exocrine functions of the pancreas and the digestive function must be evaluated; moreover, serum AFP level and quality of life must be closely monitored during follow-up.^[14–16]

In conclusion, PPTs are exceedingly rare tumors. Complete surgical resection combined with follow-up remains the primary treatment. PD is a safe and efficient method for the management of these tumors but must be performed by a surgeon with sophisticated expertise.

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