Case Report

Solid pseudopapillary tumor of the pancreas in a child: A case report and review of the literature

P. C. Das
Department of Pediatric Surgery, Fr Muller Medical College and Hospital, Mangalore - 575 002, India

Correspondence: Dr. P. C. Das, Department of Pediatric Surgery, Fr Muller Medical College and Hospital, Mangalore - 575 002, India. E-mail: drpcdas@yahoo.com

ABSTRACT

Solid pseudopapillary tumor of the pancreas (SPT) is an extremely rare pancreatic tumor, which has a low malignant potential and occurs mainly in young women. We report a case of SPT arising from the head of the pancreas in a 13 year-old girl who presented with pain in the epigastrium and was treated successfully by local excision of the tumor with preservation of duodenum. A short review of the literature is also included.

KEY WORDS: Benign neoplasm of the pancreas, Frantz's tumor, pancreatic neoplasm, solid pseudopapillary tumor of the pancreas

INTRODUCTION

Solid pseudopapillary tumors (SPTs) of the pancreas are rare and most commonly occur in young women in their second to fourth decade of life. Its incidence in children is still rarer. The pathogenesis of these tumors is controversial. Some researchers assume that the tumors have an endocrine origin while others postulate its exocrine origin. SPTs are regarded as benign or low-grade malignant tumors.[1-3] We report a case of SPT of the pancreas in a pubertal girl.

CASE REPORT

A 13–year-old girl was admitted with epigastric pain of three days' duration and nonbilious vomiting of two days' duration. Examination revealed a stable child with no fever, pallor, icterus or lymphadenopathy. Abdomen was not distended but tenderness with guarding was present in the epigastrium without any mass being felt. Her hemogram and liver function tests were normal, serum amylase was 61 units/L.Ultrasonography of the abdomen showed a complex echogenic, well-defined mass of 10 cm x 10 cm size situated in the groove between the 2nd part of duodenum and the head of the pancreas. Computed tomography (CT) of the abdomen revealed a well-encapsulated, heterogenous mass of 10 cm x 10 cm arising from the head of the pancreas, very close to the duodenum. Common bile duct, pancreatic duct and liver were normal. Gastroduodenoscopy examination was normal. On laparotomy, there was a well-encapsulated, soft, solid, round mass of 8 cm diameter arising from the head of the pancreas near the duodeno-pancreatic groove. The tumor was mostly exophytic with its attachment to the anterior surface of the pancreatic head and anteromedial aspect of the 2nd part of the duodenum. The duodenum could not be separated from the mass. Hence, exision of the mass along with the attached portion of the anteromedial aspect of 2nd part of the duodenum was carried out. The ampulla of Vater was far away from the excised margin. The duodenal defect was repaired by a jejunal patch. The child had an uneventful postoperative recovery. There was no leak from the duodenal repair site, which was confirmed by postoperative Barium meal X-ray [Figure 1]. The child is doing well and symptom-free on follow-up one year after surgery.

Histopathological examination of the mass shows monotonous cells arranged in trabecular, pseudopapillary and diffuse pattern. Stroma showed myxoid area, haemorrhage, necrosis, fibroblastic proliferation with normal pancreatic tissue. Resected margins were free of tumor. Sections from the resected duodenum were unremarkable.

DISCUSSION

Pancreatic tumors have been categorized as exocrine and endocrine tumors and have been subdivided into solid

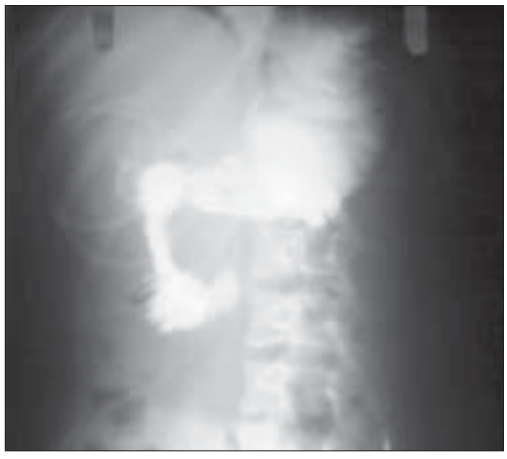

Figure 1: Postoperative barium meal X-ray showing free flow of contrast in the duodenum without any evidence of leak

and cystic epithelial tumors or also as functional and non-functional endocrine tumors. Cystic tumors include serous cystic, intraductal papillary mucinous and solid pseudopapillary tumors as per the reclassification of pancreatic tumors by the World Health Organization in1996.[2] SPT was first reported by Frantz in 1959, hence it is also called Frantz tumor.[4] SPT has other names such as papillary epithelial neoplasm, papillary cystic carcinoma, solid and cystic tumor of the pancreas and low-grade papillary tumor.[1] It is regarded as a benign or low-grade malignant neoplasm. Seven hundred and eighteen cases of SPTs have been reported from 1933 to December 2003.[5] The incidence of SPT is 0.2-2.7% in adult pancreatic patients and its location is most commonly in the head of the pancreas.[1] But its incidence in children is about 8-16.6%.[6,7] SPTs of the pancreas are common in young women in the age group of 20-40 years and rarely in men and the elderly. Children accounted for approximately 20%.[1,5]

The presenting features of the SPTs are nonspecific. Abdominal pain is the most common symptom followed by a slowly enlarging mass in the upper abdomen. Some patients are completely asymptomatic and the mass is detected either during routine examination or after injury.[5] Patients presenting with icterus due to bile duct obstruction are uncommon and symptoms of intestinal obstruction are very rare.[1] SPT is easily detected as a heterogenous mass by image examinations such as ultrasonography, CT or magnetic resonance imaging (MRI) but the findings are not specific.[1,5] In addition, clinicians lack knowledge of this rare disease, so misdiagnosis and inappropriate therapy are hard to avoid. Due to these factors, preoperative and operative diagnoses are often inaccurate. The rate of misdiognosis is 45-50%.[6] Sometimes, the origin of the tumor can not be clearly confirmed by an imaging study because it is larger, it envelops the surrounding tissues or it arises at the pancreatico-duodenal groove as in our case.[1] Histologically, SPT is usually encapsulated and is composed of a mixture of cystic, solid and hemorrhagic components. Both a capsule and intramural hemorrhage are important clues to its diagnosis because they are rarely found in other pancreatic neoplasms.[1,2,5] The fact that SPTs occur predominantly in young women led to the study of gender hormonal receptors by several authors without any evidence of estrogen receptors in the pathogenesis of the tumor.[5]

As SPT is a low-grade malignant tumor, surgical excision remains the mainstay of treatment. The choice of local tumor resection, resection of part of pancreas or radical resection depends on the judgment of the tumor's boundary. In Japan, 35% of the SPTs originating in the head of the pancreas have been treated with enucleation.[8] Recurrence of SPTs after complete resection has not been reported. About 15% of patients are reported to have liver metastasis, which can be removed operatively without endangering the life of the patients.[1] Radiotherapy has been suggested in cases of unresectable SPTs because they are radiosensitive.[5] In conclusion, SPT should be suspected in young women and girls presenting with epigastric pain. More than 95% of patients with SPT limited to the pancreas are cured by complete surgical excision. SPTs arising from the head of the pancreas can be excised completely without doing pancreaticoduodenectomy as was done in our case and also by others.[8] Operations must be (with particular respect to radical oncology) as conservative as possible because of the relatively low malignancy and the encapsulated form of the neoplasm.[9]

ACKNOWLEDGEMENT

We are thankful to the Dean, Fr Muller Medical College and Hospital for allowing us to publish this paper.

REFERENCES

1. Chen X, Zhou GW, Zhou HJ, Peng CH, Li HW. Diagnosis and treatment of pseudopapillary tumors of the pancreas. Hepatobiliary Pancreat Dis Int 2005;4:456-9.
2. Kloppel G, Solcia E, Longnecker DS, *et al*. Histological typing of tumors of the exocrine pancreas. 2nd ed. (WHO international histological classification of tumors). Springer: Berlin, Heidelberg, New York; 1996.
3. Solcia E, Capella C, Kloppel G. Tumors of the pancreas. *In*: Atlas of the tumor pathology, fascicle 2. Armed Forces Institute of Pathology: Bethesda, Maryland; p. 29-30.
4. Frantz VK. Tumors of the pancreas. *In*: Atlas of tumor pathology. Armed Forces Institute of Pathology: Washington DC; 1959. p. 32-3.
5. Papavramidis T, Papavramidis S. Solid pseudopapillary tumors of the pancreas: review of 718 patients reported in English literature. J Am Coll Surg 2005;2:965-72.
6. Jaksic T, Yaman M, Thorner P, Wesson DK, Filler RM, Shandling

B. A 20-year review of pediatric pancreatic tumors. J Pediatr Surg 1992;27:1315-7.
7. Grosfeld JL, Vane DW, Rescorla FJ, McGuire W, West KW. Pancreatic tumors in childhood: Analysis of 13 cases. J Pediatr Surg 1990;25:1057-62.
8. Akiyama H, Ono K, Takano M, Sumida K, Ikuta K, Miyamoto O. Solid-pseudopapillary tumor of the pancreatic head causing marked distal atrophy: A tumor originated posterior to the main pancreatic duct. Int J Gastrointest Cancer 2002;32:47-52.
9. Nadler EP, Novikov A, Landzberg BR, Pochapin MB, Centeno B, Fahey TJ, *et al*. The use of endoscopic ultrasound in the diagnosis of solid pseudopapillary tumors of the pancreas in children. J Pediatr Surg 2002;37:1370-3.

Source of Support: Nil, **Conflict of Interest:** None declared.