Solid pseudo papillary tumor of pancreas: Presenting as acute abdomen in a female child

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ABSTRACT

Solid pseudo papillary tumor (SPT) or Frantz's tumor is a slow-growing low-grade malignant tumor, commonly seen in young patients with a female predominance, which is commonly located in the body and tail of the pancreas. We report a case of SPT arising from the body of the pancreas in a 12-year-old girl who presented with acute abdomen and was treated successfully by local excision of the tumor with preservation of head of pancreas and spleen.

Key words: *Frantz's tumor, pancreatic neoplasm, solid pseudo papillary tumor, splenectomy*

INTRODUCTION

Frantz described first time in 1959 about a rare and characteristic tumor arising from body and tail of the pancreas known as solid pseudo papillary tumor (SPT) of pancreas.[1] The incidence of SPT is 0.13-2.7% of all pancreatic tumors.[2] It usually affects young women, with a 10:1 predominance over men, at an average age of 24 years.[3] SPT usually present with an abdominal mass, abdominal pain or discomfort, but sometimes, it is an incidental finding on imaging studies for other reasons. Many of the times, general surgeons are unaware of the SPT until the diagnosis is made by computed tomography (CT) scan of the abdomen, especially when it is presenting as acute abdomen. We report a case where a female child with SPT presented as acute abdomen with hemoperitoneum. The hemoperitoneum was due to spontaneous rupture of SPT. In few case reports, the causes of rupture being blunt trauma to the abdomen,[4-6] and while spontaneous rupture is quite uncommon.

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CASE REPORT

A female child aged 12 years, initially presented to a surgeon elsewhere, with a history of sudden onset of severe pain in the upper abdomen and no history of trauma to the abdomen. There was tachycardia with normal blood pressure (BP). The surgeon on abdominal examination found to have a vague epigastric mass associated with tenderness. Then the patient underwent ultrasonography of the abdomen and suspected to have the possibility of internal bleeding within pseudocyst of the pancreas, occupying whole of the lesser sac. Then patient was referred to us for further management.

There was severe pain in the abdomen with tachycardia and BP was 100/60 mm of Hg. As other routine blood investigations were within normal limits, patient was subjected to CT scan of the abdomen, which revealed a well-demarcated mass of size 10 cm × 13 cm arising from the body of pancreas, which was composed of a solid-cystic portion with enhancement of contrast in solid portions and occupying the whole of the lesser sac, posteriorly abutting to the splenic vessels with a breach in the anterolateral part of the capsule (rupture) of tumor with minimal hemoperitoneum. The distal tail of pancreas could not be made out. The impression was SPT arising from the body of pancreas without any metastasis to the liver.

Then it was planned for surgical excision of the mass along with possible splenectomy. An informed consent was taken for the same. The roof top incision was taken. On the exploration, there was a significantly large tumor mass arising from the body of the pancreas, extending to the left subhepatic region, displacing the gastrohepatic ligament anteriorly. The tumor was abutting the transverse mesocolon. There were no metastatic lesions on the surface of the liver. There was minimal hemoperitoneum, which may be due to rupture of the tumor. With meticulous dissection whole mass was removed along with a tail of pancreas. The dissection to separate splenic vessels from tumor and tail of pancreas was possible; in view of low malignant potential of tumor and without any metastasis, it was decided to preserve the spleen. The head of the pancreas was preserved by dividing the head from the body by using staplers.

The excised tumor was of size 10 cm × 13 cm. The histopathological examination (HPE) of specimen revealed an encapsulated tumor with hemorrhagic and necrotic areas admixed with solid areas. The surgical margins were negative. Microscopic examination revealed features of SPT. Immunohistochemistry was reactive for CD56 and alpha-1-antitrypsin and negative for chromogranin.

DISCUSSION

This case of SPT demonstrates how rupture of tumor capsule and hemorrhage within the tumor as reported in HPE, can cause hemoperitoneum and severe abdominal pain, respectively, presenting as acute abdomen. The incidence of the rupture of SPT was reported in 2.7% of 292 cases.[3] The causes of rupture are, either trauma to abdomen[4-6] or spontaneous, due to hemorrhage within tumor.[3,7] SPT of pancreas usually not diagnosed on clinical examination, unless a CT abdomen is done to reveal it. Even radiologically the differential diagnoses for SPT are, solid and cystic lesions of pancreas such as serous microcystic adenoma, cystadenocarcinoma, mucinous cystic neoplasms, other congenital and acquired postinflammatory and infectious cysts.[8]

The SPT is slow-growing and less aggressive than that of many other pancreatic tumors, and its prognosis is better. Surgical removal of the tumor will result in almost total survival (>95%) for those patients with tumors confined to the pancreas and no metastasis. It has a low potential for local infiltration thus recurrence is rare following complete excision. The metastasis occurs only in up to 15% of cases, usually synchronous and confined to the liver or peritoneum. Lymphatic involvement is not a feature.[9]

Local resection or enucleation can be performed for tumors confined to the pancreas. Distal pancreatectomy combined with or without splenectomy can be performed for pancreatic body and/or tail tumor, and pancreatoduodenectomy for pancreatic head tumor. Patients may survive a long time after radial resection of the tumor. The prognosis of SPT patients even with unrespectable metastasis is good. The role of chemotherapy and radiotherapy remains to be studied.[10] At present, the surgical procedure of choice for SPT still remains controversial. If possible the surgical procedures are preserving pancreatic parenchyma and function should be selected, considering the nature of SPT. Similarly in our case, local excision of the tumor with preservation of head of pancreas and spleen has been done.

In recent years, laparoscopic procedures of enucleation and distal pancreatectomy have already been done and have shown that this approach can be considered for the low-grade malignant SPT.[11-13] It was mentioned that there were no statistically significant differences in the postoperative complications and the prognosis between laparoscopic surgery and open surgery for SPT.[14] In acute cases, like our case with rupture of SPT, open surgery is always better. Recently, the robotic distal pancreatectomy is becoming as a possible minimally invasive technique for patients with SPT. Nevertheless, its oncological indications are yet to be defined.[15]

CONCLUSION

A relatively uncommon case of hemorrhage within tumor and spontaneous rupture of SPT of the pancreas, presenting as acute abdomen in a female child has been described. We were able to locally resect SPT completely with preservation of head of pancreas and spleen. SPT is considered to be having a good prognosis after the surgery.

REFERENCES

1. Frantz VK. Tumors of the pancreas, Atlas of tumor pathology, VII. In: Fascicles 27 and 28. Washington: Armed Forces Institute of Pathology; 1959. p. 32-3.
2. Crawford BE 2nd. Solid and papillary epithelial neoplasm of the pancreas, diagnosis by cytology. South Med J 1998;91:973-7.
3. Mao C, Guvendi M, Domenico DR, Kim K, Thomford NR, Howard JM. Papillary cystic and solid tumors of the pancreas: A pancreatic embryonic tumor? Studies of three cases and cumulative review of the world's literature. Surgery 1995;118:821-8.
4. Potrc S, Kavalar R, Horvat M, Gadzijev EM. Urgent Whipple resection for solid pseudopapillary tumor of the pancreas. J Hepatobiliary Pancreat Surg 2003;10:386-9.
5. Sanchez JA, Newman KD, Eichelberger MR, Nauta RJ. The papillary-cystic neoplasm of the pancreas. An increasingly recognized clinicopathologic entity. Arch Surg 1990;125:1502-5.

6. Huang HL, Shih SC, Chang WH, Wang TE, Chen MJ, Chan YJ. Solid-pseudopapillary tumor of the pancreas: Clinical experience and literature review. World J Gastroenterol 2005;11:1403-9.
7. Panieri E, Krige JE, Bornman PC, Graham SM, Terblanche J, Cruse JP. Operative management of papillary cystic neoplasms of the pancreas. J Am Coll Surg 1998;186:319-24.
8. Warshaw AL, Compton CC, Lewandrowski K, Cardenosa G, Mueller PR. Cystic tumors of the pancreas. New clinical, radiologic, and pathologic observations in 67 patients. Ann Surg 1990;212:432-43.
9. Vollmer CM Jr, Dixon E, Grant DR. Management of a solid pseudopapillary tumor of the pancreas with liver metastases. HPB (Oxford) 2003;5:264-7.
10. Yu PF, Hu ZH, Wang XB, Guo JM, Cheng XD, Zhang YL, *et al.* Solid pseudopapillary tumor of the pancreas: A review of 553 cases in Chinese literature. World J Gastroenterol 2010;16:1209-14.
11. Dedieu A, Rault A, Collet D, Masson B, Sa Cunha A. Laparoscopic enucleation of pancreatic neoplasm. Surg Endosc 2011;25:572-6.
12. Zhang T, Du X, Zhao Y. Laparoscopic surgery for pancreatic lesions: Current status and future. Front Med 2011;5:277-82.
13. Takamatsu S, Nagano H, Ohtsukasa S, Kawachi Y, Maruyama H. A case of spontaneous ruptured solid pseudopapillary tumor of pancreas resected by laparoscopic surgery. Case Rep Med 2013;2013:953240.
14. Cai H, Zhou M, Hu Y, He H, Chen J, Tian W, *et al.* Solid-pseudopapillary neoplasms of the pancreas: Clinical and pathological features of 33 cases. Surg Today 2013;43:148-54.
15. Ntourakis D, Marzano E, De Blasi V, Oussoultzoglou E, Jaeck D, Pessaux P. Robotic left pancreatectomy for pancreatic solid pseudopapillary tumor. Ann Surg Oncol 2011;18:642-3.

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