J Med Sci 2020;40(1):34-37
DOI:10.4103/jmedsci.jmedsci_46_19

CASE REPORT

Solid Pseudopapillary Neoplasm of Pancreas in a 12-Year-Old Girl

Prosanta Kumar Bhattacharjee[1], Tanvi Goel[2], Oindrila Biswas[1]

[1]Department of Surgery, I.P.G.M.E and R/S.S.K.M Hospital, Kolkata, West Bengal, [2]Department of Pediatric Surgery, All India Institute of Medical Sciences, New Delhi, India

A-12-year old girl presented with acute exacerbation of mild upper abdominal pain and nausea associated with weight loss. Subsequent work up revealed raised serum lipase and a bulky (7.8 cm x 4.9 cm x 2 cm) mass involving the head of the pancreas with areas of necrosis and cystic degeneration and peripheral rim enhancement, without any calcification. MPD was dilated (6mm). The cyst fluid had normal amylase, CA 19-9 level, CEA levels, and cytology revealed atypical epithelial cells. Patient underwent an uneventful pancreaticoduodenectomy. Subsequent histopathology revealed solid pseudopapillary tumor of the pancreas of low-grade malignancy.

Key words: Pancreatic cyst, solid pseudopapillary neoplasm, diagnosis, management

INTRODUCTION

Solid pseudopapillary neoplasm (SPN) is a rare heterogeneous solid and cystic lesion of the pancreas seen mostly in women in their 20s and 30s.[1] They usually present with abdominal pain and less frequently with jaundice and pancreatitis. These lesions are potentially malignant, and hence, surgical resection is warranted. We present a successfully managed rare case of malignant SPN of the head of pancreatic in a 12-year-old girl who presented with mild acute pancreatitis.

CASE REPORT

A 12-year-old girl was referred to the emergency department of our hospital with acute and severe pain localized to the mid-upper abdomen associated with nausea and anorexia. The pain was nonradiating in nature. On inquiry, she said that she was experiencing intermittent episodes of pain of lesser intensity over the past 6 months and it got exacerbated 5 days ago. She lost around 2-kg weight over the past 6 months. There was no history suggestive of gallstone disease or history of abdominal trauma. There was no history of fever, jaundice, hematemesis, any relevant past medical history or any family history of pancreatic or other cancers. Her menstrual history was normal.

On examination, the child was of average built with a body mass index of 18.67. Her general physical and systemic examinations were essentially normal. The patient had reported with some investigation reports done from another hospital where she reported initially. The routine hematological and biochemical examinations including liver and renal function tests were all within normal limits, except for a borderline increase in serum amylase (110 U/L; Reference value =30.00–96.00) and significantly raised serum lipase (222 U/L; Reference value = 5.00–60.00), suggesting acute pancreatitis. The abdominal ultrasound scan report revealed a 7.45 cm × 4.89 cm solid retroperitoneal space-occupying lesion of heterogeneous echotexture probably arising from the head of the pancreas.

We performed an abdominal contrast-enhanced computed tomography (CECT) scan for better delineation of the lesion and to plan our treatment. It showed a single, well-delineated, bulky (7.8 cm × 4.9 cm × 2 cm) mass of heterogeneous density with peripheral rim enhancement involving the head of the pancreas, abutting and pushing the superior mesenteric vessels medially, without any encasement. The tumor showed areas

Received: March 09, 2019; Revised: May 30, 2019; Accepted: June 17, 2019; Published: September 04, 2019
Corresponding Author: Dr. Prosanta Kumar Bhattacharjee, Flat No 5, 4th Floor, "Suryatoran Apartment," 114/A, Barasat Road, Kolkata - 700 110, West Bengal, India. Tel: +918820025064; Fax: +9103325650403. E-mail: prosantabh@rediffmail.com

How to cite this article: Bhattacharjee PK, Goel T, Biswas O. Solid pseudopapillary neoplasm of pancreas in a 12-year-old girl. J Med Sci 2020;40:34-7.

of necrosis and cystic degeneration [Figure 1]. No significant retroperitoneal lymphadenopathy was evident. The main pancreatic duct (MPD) was dilated and measured 6 mm.

Investigative findings of a large pancreatic head lesion with worrisome recognized risk factors such as its symptomatic nature, size >3 cm, dilatation of the MPD prompted us to advice endoscopic ultrasound (EUS) and EUS-guided aspiration cytology and fluid analysis. The EUS characterization of the tumor was in sync with CECT findings; moreover, it also ruled out any communication with MPD. The cyst fluid had normal amylase, carbohydrate antigen 19-9 level, and carcinoembryonic antigen levels. Cytological study revealed atypical epithelial cells.

Although we could not arrive at a definite preoperative diagnosis, the symptomatic nature of the tumor with red flags detected on workup suggesting a risk of malignancy warranted surgical intervention. The patient underwent pancreaticoduodenectomy after detailed discussion regarding the risks and benefit of surgery.

Macroscopically, the tumor measured 8 cm × 5 cm × 2 cm and the cut open specimen showed solid and cystic areas [Figure 2]. She had an uneventful postoperative recovery.

The histopathological picture of the resected specimen showed polygonal monomorphic cells with eosinophilic cytoplasm arranged in a pseudopapillary pattern around delicate vessels with areas of hemorrhage and necrosis consistent with the diagnosis of a solid pseudopapillary tumor of the pancreas of low-grade malignancy [Figure 3]. Resection margins were free of tumor.

She was referred to our medical oncologist who advised no adjuvant therapy in view of the localized, low-grade nature of the lesion and negative resection margins. She has been on regular follow-up for over 3 years and has shown no evidence of either local recurrence or distant metastasis. She has gained weight and has been pursuing her normal academic activities, but complains of occasional abdominal colics.

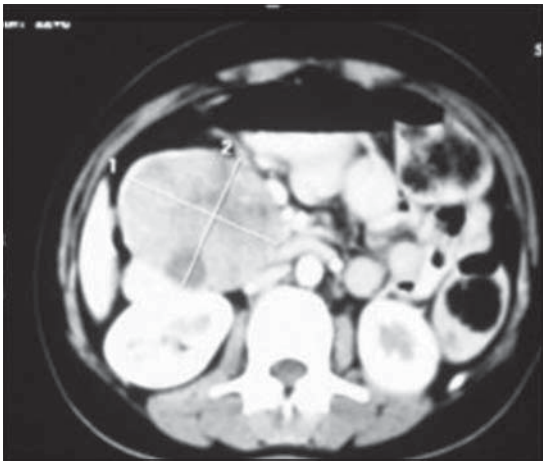

Figure 1: Contrast-enhanced computed tomography abdomen shows a single well-delineated, 78 mm × 49 mm × 20 mm, pancreatic head mass of heterogeneous density and peripheral rim enhancement, abutting and pushing the superior mesenteric vessels medially, with areas of necrosis and cystic degeneration

DISCUSSION

Pancreatic cystic neoplasms are being diagnosed with increasing frequency because of the widespread use of ultrasonography and other newer generation cross-sectional imaging as tools for evaluation of patients with abdominal symptoms. Often these tumors are diagnosed following imaging done for some other reasons.[2]

SPNs of the pancreas are rare, large, solitary, well-circumscribed solid tumors of the pancreas, most commonly involving the pancreatic tail, with areas of cystic degeneration and a thick fibrous wall without much desmoplasia.[3] Due to these macroscopic solid and cystic components, they are also referred to as solid and cystic tumors of the pancreas.[4]

Frantz first described four cases of SPN as papillary tumors of the pancreas in 1959.[5] Hamoudi *et al.* further characterized them in 1970.[6] They are thus often referred to as Frantz or Hamoudi tumors.[7] They constitute around 1%–2% of all exocrine pancreatic neoplasms, but 52%–71% of all pancreatic tumors in children and adolescents, and seen mostly in young women.[3,8-10] In India, the median age at presentation is 24 years, females are six times more commonly affected, and the most common mode of presentation is abdominal pain.[11] These neoplasms carry a low malignant risk of around 10%–20%.[1,3,10]

The etiology and the natural history of this neoplasm are unknown, its relationship to sex hormones has been suspected

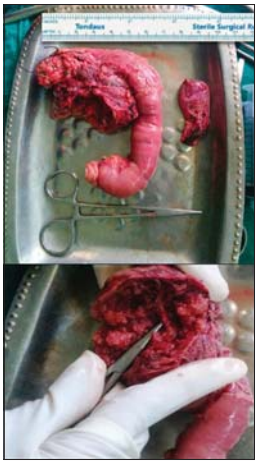

Figure 2: Cut open specimen of pancreatic head mass. Hemostat pointing toward a cystic area within

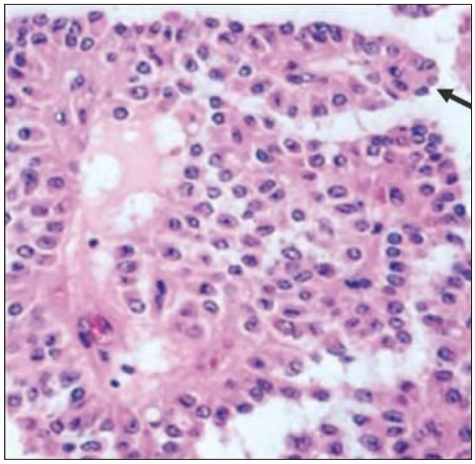

Figure 3: Polygonal, eosinophilic, loosely arranged cells surrounding delicate blood vessels forming pseudopapillary structures (shown by the arrow) with hyaline stromal bands in between (H and E, ×100)

but not confirmed, but activating β-catenin gene mutations have been noted.[7,12] There are no specific clinical presentations of this neoplasm, and they are usually detected, as in our case, during cross-sectional imaging done for nonspecific chronic, recurrent abdominal pain.[13]

Characteristically, solid pseudopapillary tumors appear, on CECT, as large well-delineated and encapsulated mass that contains both solid and cystic components without dilatation of MPD. The solid-enhancing components with calcifications are seen at the periphery of the lesion, whereas the cystic areas are more central.[6,9] One should consider pancreatic duplication cyst and other cystic neoplasms of the pancreas as important differential diagnoses.[14]

Our findings were mostly consistent with other reports of this rare disease except for an earlier presentation, the atypical CECT findings of dilated MPD, and absence of calcifications.

The malignant potential of different cystic neoplasms varies, and an accurate diagnosis is essential for proper management.[2] Primary solid pseudopapillary tumors have a low malignant potential, and its prognosis is much better than other pancreatic neoplasms, but it may occasionally present with local invasion and multiple distant metastases mostly in the liver and regional lymph nodes.[3]

Complete surgical resection is the treatment of choice. Recurrence is uncommon, even without adjuvant therapy, provided the surgeon achieves R0 resection.[10] The role of neoadjuvant and adjuvant therapies are limited.[11] This makes meticulous surgery even more important. Our patient underwent pancreaticoduodenectomy as the tumor was located in the head of the pancreas. All our resection margins were free of tumor. She received no adjuvant therapy and is recurrence free till date.

CONCLUSION

SPN is overall a rare pancreatic neoplasm. In the pediatric population, SPN is more common than other pancreatic tumors. Preoperative diagnosis is difficult and requires a strong suspicion based on cross-sectional imaging studies. Patients may have an overall 5-year survival of over 95%, provided the diagnosis was early and the surgeon achieves complete extirpation of the tumor.[11]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given her consent for her images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Kim TS, Fernandez-del Castillo C. Diagnosis and management of pancreatic cystic neoplasms. Hematol Oncol Clin North Am 2015;29:655-74.
2. Spinelli KS, Fromwiller TE, Daniel RA, Kiely JM, Nakeeb A, Komorowski RA, *et al.* Cystic pancreatic neoplasms: Observe or operate. Ann Surg 2004;239:651-7.
3. 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.
4. Klöppel G, Solcia E, Longnecker DS, Capella C, Sobin Leslie H. Histological Typing of Tumors of Exocrine Pancreas. 2nd ed. Berlin; Springer-Verlag; 1996. Available from: http://www.who.int/iris/handle/10665/37470. [Last accessed on 2019 Feb 02].
5. Frantz VK. Papillary tumors of the pancreas: Benign or malignant? Tumors of the pancreas. In: Atlas of tumor Pathology. 1st ed. Washington DC: Armed Forces Institute of Pathology; 1959. p. 32-3.
6. Hamoudi AB, Misugi K, Grosfeld JL, Reiner CB. Papillary epithelial neoplasm of pancreas in a child. Report of a case with electron microscopy. Cancer 1970;26:1126-34.

7. Reddy S, Cameron JL, Scudiere J, Hruban RH, Fishman EK, Ahuja N, *et al.* Surgical management of solid-pseudopapillary neoplasms of the pancreas (Franz or Hamoudi tumors): A large single-institutional series. J Am Coll Surg 2009;208:950-7.
8. Yagcı A, Yakan S, Coskun A, Erkan N, Yıldırım M, Yalcın E, *et al.* Diagnosis and treatment of solid pseudopapillary tumor of the pancreas: Experience of one single institution from Turkey. World J Surg Oncol 2013;11:308.
9. El Nakeeb A, Abdel Wahab M, Elkashef WF, Azer M, Kandil T. Solid pseudopapillary tumour of the pancreas: Incidence, prognosis and outcome of surgery (single center experience). Int J Surg 2013;11:447-57.
10. Mahida JB, Thakkar RK, Walker J, Shen R, Kenney BD, Prasad V, *et al*. Solid pseudopapillary neoplasm of the pancreas in pediatric patients: A case report and institutional case series. J Ped Surg Case Rep 2015;3(4):149-53.
11. Kumar NA, Bhandare MS, Chaudhari V, Sasi SP, Shrikhande SV. Analysis of 50 cases of solid pseudopapillary tumor of pancreas: Aggressive surgical resection provides excellent outcomes. Eur J Surg Oncol 2019;45:187-91.
12. Cienfuegos JA, Lozano MD, Rotellar F, Martí P, Pedano N, Arredondo J, *et al.* Solid pseudopapillary tumor of the pancreas (SPPT). Still an unsolved enigma. Rev Esp Enferm Dig 2010;102:722-8.
13. Brugge WR, Lauwers GY, Sahani D, Fernandez-del Castillo C, Warshaw AL. Cystic neoplasms of the pancreas. N Engl J Med 2004;351:1218-26.
14. Chakhachiro ZI, Zaatari G. Solid-pseudopapillary neoplasm: A pancreatic enigma. Arch Pathol Lab Med 2009;133:1989-93.