Srp Arh Celok Lek. 2013 May-Jun;141(5-6):384-386
ПРИКАЗ БОЛЕСНИКА / CASE REPORT
DOI: 10.2298/SARH1306384M
UDC: 616.37-006

Solid and Cystic Pseudopapillary Tumor of the Pancreas: A Case Report

Bojan Z. Milošević[1,2], Rade M. Marković[1], Aleksandar M. Cvetković[1,2]

[1]Department of Gastrointestinal Surgery, Clinic for Surgery, Clinical Center Kragujevac, Kragujevac, Serbia;
[2]Surgical Department, Faculty of Medical Sciences, University of Kragujevac, Kragujevac, Serbia

SUMMARY

Introduction Solid and cystic pseudopapillary tumor of the pancreas is a rare tumor of the pancreas, for the first time described by Frantz et al. in 1959. The majority of patients are young females and most of them are asymptomatic.

Case Outline We report a case of 25-year old woman who was admitted to our institution with abdominal pain and a palpable mass in the left hypochondrial area. US and CT scan revealed a solid and cystic pseudopapillary tumor in the head of the pancreas. The patient was treated by Whipple procedure, modification Longmire-Traverso. There was no metastatic disease either in the liver or peritoneum. Histologically the tumor was diagnosed as a solid and cystic pseudopapillary tumor of the pancreas.

Conclusion The unclear pre-operative diagnoses, together with incidence of potential malignancy as well as good outcome with resection, suggest that all suspected cystic tumors of the pancreas should be resected. The exact diagnosis is based on histological findings.

Keywords: solid; cystic; pseudopapillary; tumor; pancreas

INTRODUCTION

Solid and cystic pseudopapillary tumor of the pancreas is a very rare clinical pathologic entity. It is a rare tumor of the pancreas for the first time described by Frantz et al. in 1959 [1]. The tumor is nine times more frequent in the body and tail of the pancreas. The majority of patients are young females and most of them are asymptomatic [2]. Cases in men are also reported [3]. Its origins remain unclear. The difficulties with pre-operative diagnosis, together with high incidence of tumors with potential malignancy, and good outcome with resection, suggest that all suspected cystic tumors of the pancreas should be resected [4, 5].

CASE REPORT

A 25-year-old woman was admitted to surgical department of the Clinical Center in Kragujevac due to epigastric pain and palpable mass. The first complaints appeared 3 months before admission. Ultrasound and CT scan showed the presence of solid and cystic tumor localized within the head of the pancreas.

On examination, the patient's blood pressure was 120/70 mmHg, pulse 72 beats/min, body temperature was 36.8°C, and her skin and sclera were not jaundiced. There was tenderness in the epigastric region and left hypochondrial area where the tumor itself was palpable.

Laboratory tests revealed 10.4×10^9/L white blood cells, 3.33×10^{12}/L red blood cells, hemoglobin 120 g/L, platelets 386×10^9/L, 52 IU/L blood amylase, 12.1 μmol/L total bilirubin and 5.3 μmol/L direct bilirubin, 66 IU/L alkaline phosphatase, AST 28 IU/L, ALT 34 IU/L, and total protein 70 g/L.

The patient underwent the Whipple's procedure, modified according to Longmire-Traverso (Figure 1). There was no metastatic disease either in the liver or peritoneum. Histologically tumor was diagnosed as a pancreatic solid and cystic pseudopapillary tumor (Figures 2 and 3). Histochemistry of the tumor showed CKAE 1/3, CK7, CK8, vimentin, NSE, chromogranin, synaptophtysin, CD 56, cyklin D1, and progesterone receptor positivity.

Recovery was uneventful and the patient was discharged from the Clinic after two weeks and has remained disease-free at 24 months up to now.

DISCUSSION

Solid and cystic pseudopapillary tumor of the pancreas is very rare. We report this case because tumor was localized in the head of the pancreas, and it is well known that the tumor is more frequent in the body and tail of the pancreas [6, 7]. We performed a radical surgical intervention involving Whipple's pylorus preserving cephalic duodenopancreatectomy, modification Longmire-Traverso and complete surgical removal of the tumor, which is the recommended treatment. Cystic tumors of the pancreas are often misdiagnosed as pseudocysts and are inappropriately managed [8]. MRI is highly specific and sensitive diagnostic method for these tumors [9]. The exact diagnosis is based on histological findings [10]. After surgical removal prognosis is excellent because malignant potential of solid-cystic-

Correspondence to:
Aleksandar M. CVETKOVIĆ
Surgical Department
Faculty of Medical Sciences
University of Kragujevac
Svetozara Markovića 69
34000 Kragujevac
Serbia
draleksandarcvetkovic@gmail.com

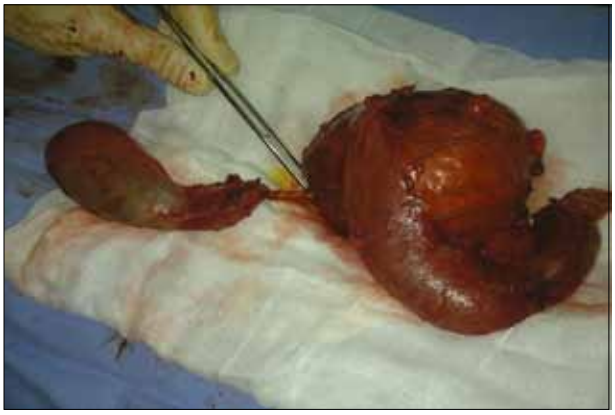

Figure 1. Macroscopic specimen achieved by cephalic duodenopancreatectomy, containing duodenum, the head of the pancreas with tumor, the part of common bile duct (marked by instrument) and gallbladder

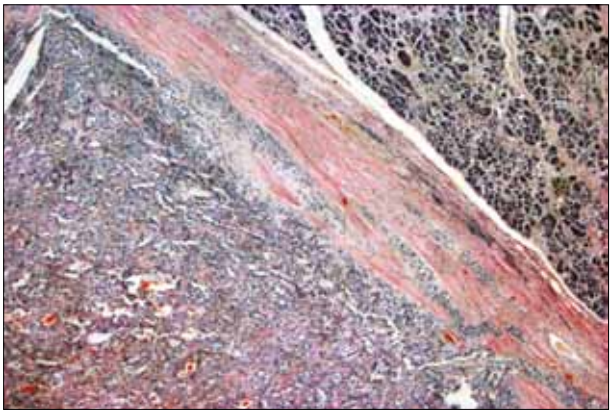

Figure 2. Histology showing polygonal tumor cells that are bright or highly eosinophilic, with oncocyti cytoplasm and irregular nuclei with finely dispersed chromatin

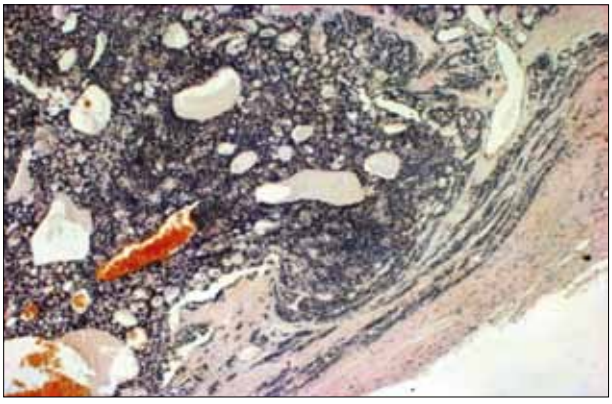

Figure 3. Histology showing tumor tissue built from solid, partly cystic pseudopapillary and zones in which protrude pseudopapillary proliferation. There are foci of cystic degeneration and hemorrhage hyalinization. Tumor tissue is separated from the pancreatic connective tissue that permeates in some places.

pseudopapillary tumor of the pancreas is low. Metastases of the tumor and local recurrence are rare [6]. Metastatic disease can occur, usually in the liver, and its management is not well defined [11]. Complications such as rupture, bleeding or secondary infections are also rare [7]. Solid and cystic pseudopapillary tumor of the pancreas should be considered in the differential diagnosis in all patients with a history of unclear epigastric pain and abdominal mass localized in the retro-peritoneum, especially in younger female patients. Its nature differs from the more common pancreatic adenocarcinoma in that it has a young female predilection; it is often asymptomatic and carries a better prognosis [8]. Delay in the diagnosis increases the frequency of associated metastatic disease and suboptimal surgical therapy. The unclear pre-operative diagnosis, the high incidence of malignant tumors and the good outcome after surgical intervention, suggest that all suspected cystic tumors of the pancreas should be resected. Thus, radical surgical intervention with complete removal of tumor is always justified, despite the price of mutilating surgery. Often, intraoperative finding of solid and cystic pseudopapillary tumor of the pancreas is the first contact with this rear entity, so awareness of possible presence of this uncommon disease is very important for the surgeon.

ACKNOWLEDGEMENTS

A part of this research is supported by the Ministry of Education, Science and Technological Development of Serbia, Grants III41007 and III41010.

REFERENCES

1. Pailoor K, Kini H, Rau AR, Kumar Y. Cytological diagnosis of a rare case of solid pseudopapillary neoplasm of the pancreas. J Cytol. 2010; 27(1):32-4.
2. Danzi M, Grimaldi L, Reggio S, Giordano M, Caterino M, Danzi R. Solid pseudo papillary tumor of the pancreas: case report and literature review. G Chir. 2008; 29(6-7):271-5.
3. Mehta N, Modi L, Patel T, Shah M. Study of cytomorphology of solid pseudopapillary tumor of pancreas and its differential diagnosis. J Cytol. 2010; 27(4):118-22.
4. Rebhandl W, Felberbauer FX, Puig S, Paya K, Hochschorner S, Barlan M, et al. Solid-pseudopapillary tumor of the pancreas (Frantz tumor) in children: report of four cases and review of the literature. J Surg Oncol. 2001; 76 (4):289-96.
5. Seo HE, Lee MK, Lee YD, Jeon SW, Cho CM, Tak WY, et al. Solid-pseudopapillary tumor of the pancreas. J Clin Gastroenterol. 2006; 40(10):919-22.
6. Čolović R, Micev M, Zogović S, Čolović N, Stojković M, Grubor N. Solid and cystic-papillary tumor of the pancreas. Srp Arh Celok Lek. 2000; 128(11-12):393-6.
7. Karamarković AR, Micev M, Ćulafić D, Popović N, Djukić V. Solid cystic pseudopapillary tumor of the pancreas. Srp Arh Celok Lek. 2004; 132(11-12):431-4.
8. Paik KY, Choi SH, Heo JS, Choi DW. Solid tumors of the pancreas can put on a mask through cystic change. World J Surg Oncol. 2011; 9:79.
9. Chang H, Gong Y, Xu J, Su Z, Qin C, Zhang Z. Clinical strategy for the management of solid pseudopapillary tumor of the pancreas: Aggressive or less? Int J Med Sci. 2010; 7(5):309-13.
10. Ooi LL, Ho GH, Chew SP, Low CH, Soo KC. Cystic tumors of the pancreas: a diagnostic dilemma. Aust NZJ Surg. 1998; 68 (12):844-6.
11. Vollmer C, Dixon E, Grant D. Management of a solid pseudopapillary tumor of the pancreas with liver metastases. HPB (Oxford). 2003; 5(4):264-7.

Солидни и цистични псеудопапиларни тумор панкреаса – приказ болесника

Бојан З. Милошевић[1,2], Раде М. Марковић[1], Александар М. Цветковић[1,2]

[1]Одељење гастроинтестиналне хирургије, Клиника за хирургију, Клинички центар Крагујевац, Крагујевац, Србија;
[2]Хируршка катедра, Факултет медицинских наука, Универзитет у Крагујевцу, Крагујевац, Србија

КРАТАК САДРЖАЈ

Увод Солидни и цистични псеудопапиларни тумор панкреаса је редак тумор панкреаса, а први пут су га описали Франц (*Frantz*) и сарадници 1959. године. Већина болесника су младе жене и углавном су без симптома обољења.

Приказ болесника Двадесетпетогодишња жена је примљена у нашу установу због бола у трбуху и палпабилне масе у левој подребарној регији. Налази ултразвука и компјутеризоване томографије су указали на солидни и цистични псеудопапиларни тумор у глави панкреаса. Болесница је лечена модификованом Випловом (*Whipple*) процедуром (*Longmire-Traverso*). Није било метастатских депозита у јетри и по перитонеуму. Тумор је хистолошки дијагностикован као солидни и цистични псеудопапиларни тумор панкреаса.

Закључак Нејасна дијагноза пре хируршког лечења, могућност да је тумор малигне природе и добар исход операције показују да се сви сумњиви цистични тумори панкреаса морају ресецирати. Тачна дијагноза се заснива на хистолошком налазу.

Кључне речи: солидни; цистични; псеудопапиларни; тумор; панкреас

Примљен • Received: 30/08/2011 **Ревизија • Revision:** 15/03/2013 **Прихваћен • Accepted:** 21/03/2013

doi: 10.2298/SARH1306384M