THE EUROPEAN SOCIETY OF MEDICINE
Medical Research Archives, Volume 12 Issue 8

CASE REPORT

Recurrent Solid Pseudopapillary Tumor of the Pancreas with Direct Extension into Gastric Mucosa: A Case Report with Consideration for Patient Management

Vinaya Rajahraman BS[1], James Lindberg MD[2,3], R. Leslie Shelton MD[3], Tasneem Ali MD[4,5], Neil Marya MD[4,5], Lacey McIntosh DO[6,7], Elisa Franquet-Elia MD[6,7], Larry Zheng MD[6,7], Tasneem Lalani MD[6,7], Alan Goldstein MD[6,7], Brian Midkiff MD[6,7], Andrew Fischer MD[8,9], Salwa Khedr MD[8,9], Vijay Vanguri MD[8,9], Yansong Geng MS[10], Linda Ding PhD[1,10], Maryann Bishop-Jodoin MEd[1], TJ FitzGerald MD[1,10*]

[1] Department of Radiation Oncology, University of Massachusetts Chan Medical School, Worcester, MA 01655
[2] Department of Surgery, University of Massachusetts Chan Medical School, Worcester, MA 01655
[3] Department of Surgery, UMass Memorial Health, Worcester, MA. 01655
[4] Department of Medicine, University of Massachusetts Chan Medical School, Worcester, MA 01655
[5] Department of Medicine, UMass Memorial Health, Worcester, MA. 01655
[6] Department of Radiology, University of Massachusetts Chan Medical School, Worcester, MA 01655
[7] Department of Radiology, UMass Memorial Health, Worcester, MA. 01655
[8] Department of Pathology, University of Massachusetts Chan Medical School, Worcester, MA
[9] Department of Pathology, UMass Memorial Health, Worcester, MA. 01655
[10] Department of Radiation Oncology, UMass Memorial Health, Worcester, MA 01655

OPEN ACCESS

PUBLISHED
31 August 2024

CITATION
Rajahraman, V., Lindberg, J., et al., 2024. Recurrent Solid Pseudopapillary Tumor of the Pancreas with Direct Extension into Gastric Mucosa: A Case Report with Consideration for Patient Management. Medical Research Archives, [online] 12(8).
https://doi.org/10.18103/mra.v12i8.5515

DOI
https://doi.org/10.18103/mra.v12i8.5515

ISSN
2375-1924

ABSTRACT

A 77-year-old woman with a history of a pancreatic mass underwent distal pancreatectomy with splenectomy in 2007 in Brazil. After presentation in September 2021 with melena and coffee-ground emesis, she received a computed tomography scan and esophagogastroduodenoscopy revealing a prominent mass lesion including the pancreas at the level of the resection margin with extension into the gastric fundus. Subsequent endoscopic ultrasound and fine needle aspiration biopsy confirmed the lesions as recurrent solid pseudopapillary tumor with direct extension to the gastric fundus. The treatment strategy included chemotherapy, intensity-modulated radiation therapy, and surgery. In this case report, we discuss strategic applications of tri-modality therapy, including surgical removal of residual gastric disease.

Introduction

Solid pseudopapillary tumors (SPTs), first described by Frantz in 1959, are extremely rare tumors of the pancreas comprising just 0.3-2.7% of all pancreatic tumors.[1–3] These tumors possess a female predominance, with a bimodal age distribution at 28 and 64 years of life.[2,4,5] Though most SPTs are benign, 10-15% of SPTs possess malignant features such as local and distance metastasis, most commonly to the liver.[6–8] Prognosis is typically highly favorable with surgical resection, though may result in rare instances of recurrence.[8,9] Multidisciplinary treatment for these rare recurrences is being explored.[6] This case report presents the clinical considerations in the tri-modality management of recurrent SPT of the pancreas with direct extension to the gastric fundus.

Case Presentation

We present the case of a 77-year-old woman with a history of a pancreatic mass who underwent distal pancreatectomy with splenectomy in 2007 in Brazil. She was told the mass was benign and would require no further treatment or surveillance. In September of 2021, she presented to our institution with three days of new-onset melena and coffee-ground emesis. Hemoglobin was 10.1 grams per deciliter (g/dL), a 1.5 g/dL drop in her baseline. She underwent an esophagogastroduodenoscopy (EGD) which showed a 16 x 17 millimeter (mm), medium-sized, ulcerated mass in the gastric fundus (Figure 1).

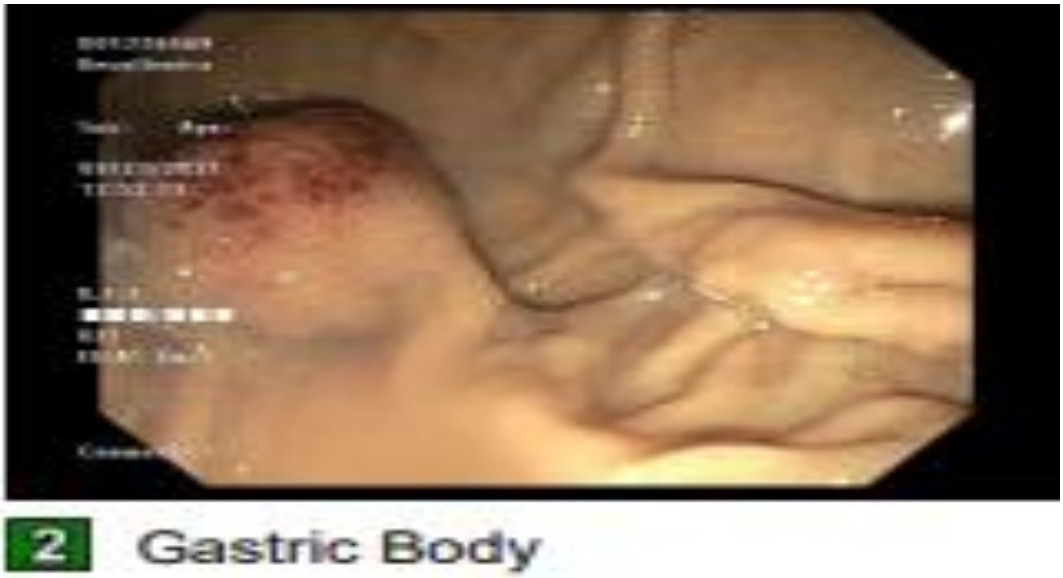

Figure 1. Mass Extending Into Gastro Fundus

Computed tomography (CT) scan of the abdomen showed a well-circumscribed, 5.8 x 4.9 centimeter (cm) heterogeneous mass within the pancreatectomy bed and a 3.8 cm filling defect in the gastric fundus (Figure 2). Endoscopic ultrasound (EUS) with fine needle aspiration of both lesions was performed to retrieve material for analysis. The tumor cells were positive for beta-catenin and synaptophysin and were negative for CK-AE1/3, CD117 and chromogranin A; the immunohistochemical studies were consistent with solid pseudopapillary tumor.

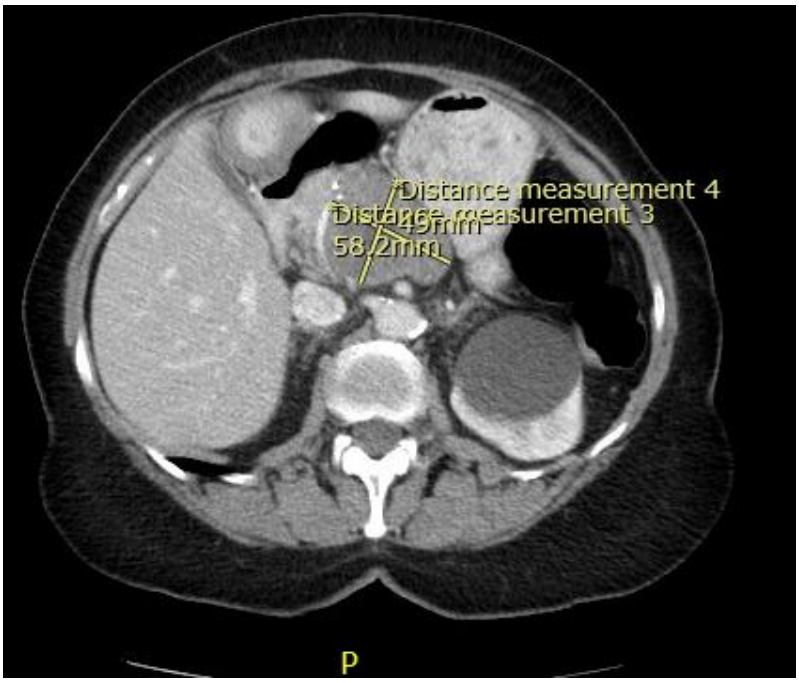

Figure 2. CT Pancreas Recurrence Extending Into the Fundus

The patient was evaluated by providers from all oncology services. Surgical colleagues recognized the locally advanced disease process with the epicenter in the pancreas involving the gastric fundus by direct extension. Surgical resection would include completion pancreatectomy with resection of the portal vein with certain positive margins at the superior mesenteric artery. Surgery would include gastrectomy; therefore, it was felt by all providers that surgical intervention was not initially feasible given the extent of disease and likely extended recovery. Therefore, the decision was made to move forward with systemic management.

In November 2021, neoadjuvant chemotherapy with gemcitabine and nab-paclitaxel was initiated with the plan of administering four cycles to shrink the tumor prior to re-visiting local regional management of the disease. Repeat CT scan of the abdomen in July 2022 showed a stable pancreatic lesion and decrease in size of the gastric lesion to 10.8 x 13 mm. Considering the improvement in size of the gastric lesion and regional involvement of the superior mesenteric and portal veins and close abutment of the superior mesenteric artery and other local vasculature, surgical oncology did not believe surgical resection with negative margins would be possible. Another repeat CT scan of the abdomen in June 2023 showed decrease in size of the pancreatic lesion to 4 x 3.5 x 4.4 cm and an unchanged hypodensity within the gastric fundus. The patient continued chemotherapy until July 2023 and was referred for radiation therapy following discussion at our Gastrointestinal Oncology Tumor Board Conference.

At the time of presentation for radiation therapy, the disease in the pancreas and former pancreatic bed remained unresectable from a vascular perspective and there was persistent disease extension into the gastric fundus. Because of disease location in the gastric fundus, response to radiation therapy could foreshadow a precarious situation for both generation of a fistula with perforation, especially if the entire target was treated to high dose. Therefore, the entire target volume was treated in an initial phase of management to all visible disease identified on anatomic volumetric imaging. After the initial phase of management, the volumes would be re-evaluated to determine if supplemental therapy could be delivered to the unresectable component. Resection of the gastric component would be considered if a tissue plane could be defined between the pancreatic disease and the gastric component. Radiation therapy was administered from July 27, 2023 to September 1, 2023 given the partial response to chemotherapy treatment and preference to avoid surgical resection. Intensity-modulated radiation therapy (IMRT) with image guidance was used for treatment. The high-risk clinical target volume (CTV) consisted of the area of recurrence in the pancreatic bed and received 5400 cGy/200 cGy fraction. The intermediate-risk CTV consisted of the area of extension of disease into the gastric fundus and peripheral tissues and received risk reduced 4860 cGy at 180 cGy/fraction with single plan dose painting (Figure 3). Following initial radiation therapy, a cleavage plane could be seen between the pancreatic component of disease and the gastric fundus (Figure 4). Supplemental IMRT directed to the site of the recurrent disease in the pancreatic bed was administered from September 19, 2023 until September 26, 2023. An additional 1800 cGy in 6 fractions of 300 cGy were delivered to bring the total dose to the recurrent disease in the pancreas to 7200 cGy (Figure 5). She received concurrent capecitabine chemotherapy.

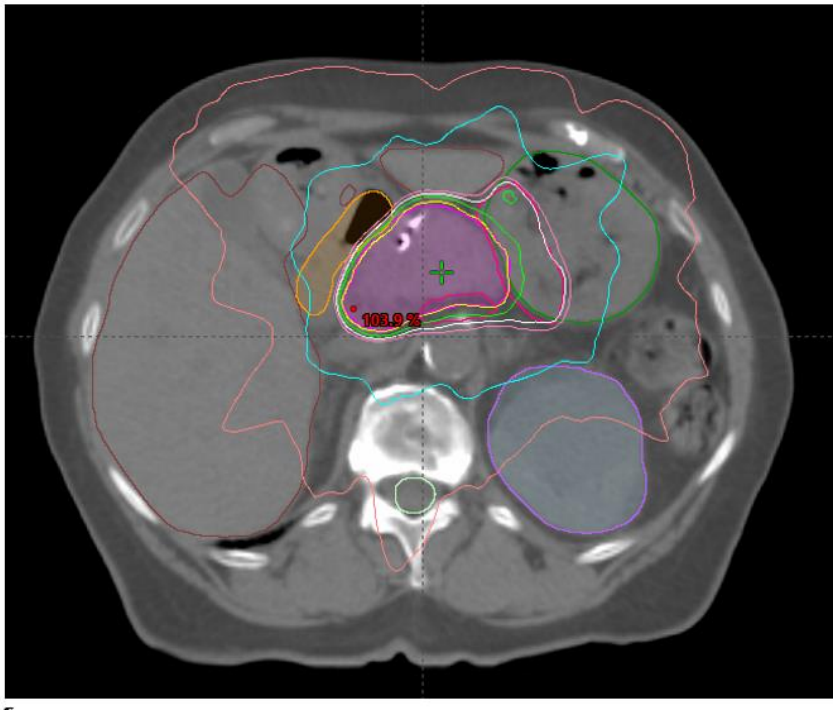

Figure 3. Initial Radiation Therapy Plan with Coverage of the Recurrence and Extension into the Fundus. Dose Painting Was Used to Include the Gastric Component of the Recurrence

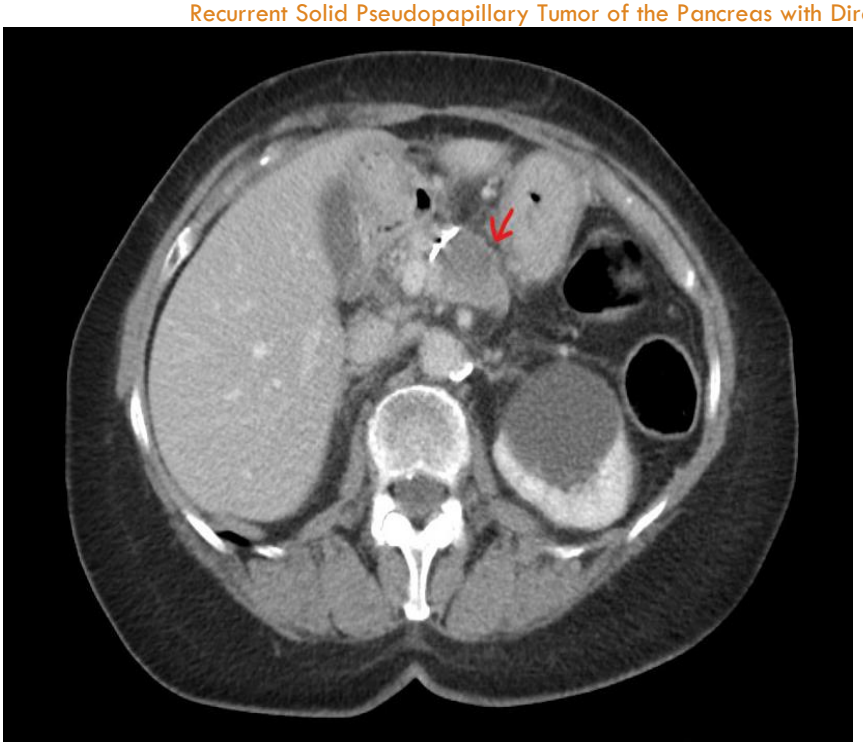

Figure 4. CT After Initial Radiation Therapy Demonstrating a Plane between the Pancreatic Component of the Recurrence and the Fundus (Red Arrow)

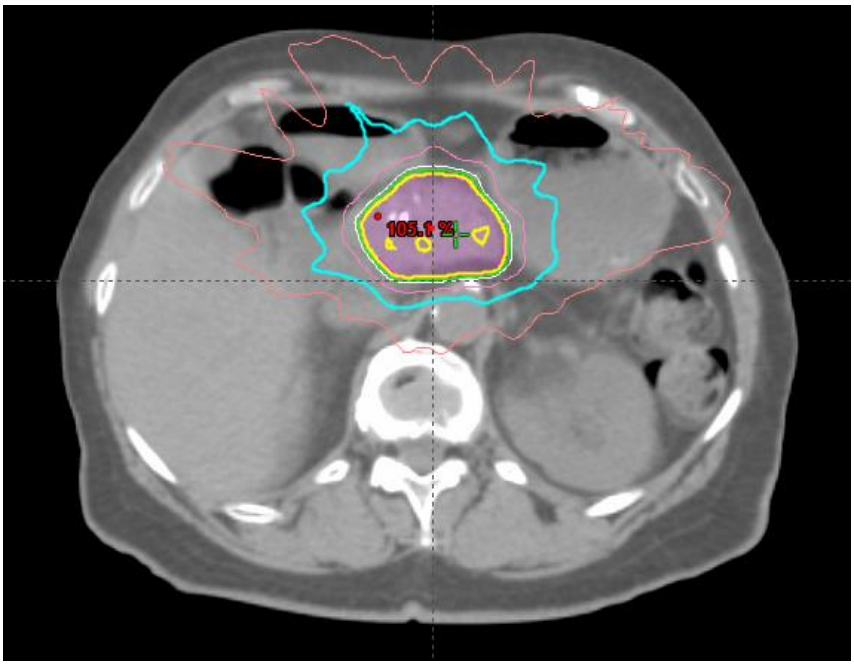

Figure 5. Boost to the Pancreatic Component of the Recurrence

Given the presence of a tissue plane between the SPT recurrence and the area of involvement in the gastric fundus on serial imaging, high-dose IMRT to the gastric fundus could result in perforation of the tissue. Therefore, surgical resection of the gastric lesion following completion of radiation therapy for the pancreatic recurrence was decided as the optimal course of action. She underwent a partial gastrectomy involving tumor resection and primary repair of the gastric fundus in January 2024 with good recovery. A 1.5 cm of residual disease was identified in the surgical resection specimen extending to, but not through, the gastric serosa (Figure 6).

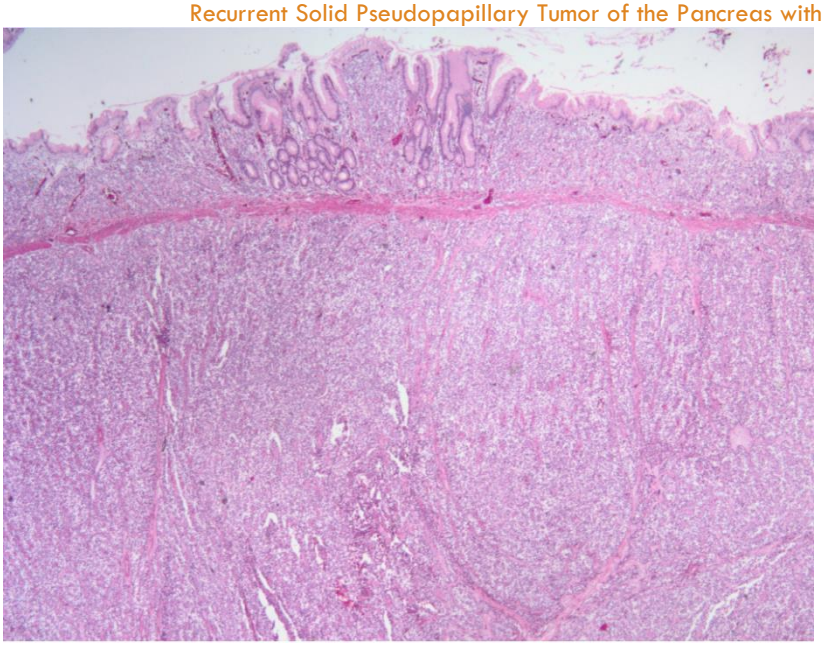

Figure 6. Residual Disease at the Serosa

Discussion

The presented case underscores important clinical considerations in the management of recurrent SPT of the pancreas with direct extension to the gastric fundus. Although SPTs typically exhibit low malignant potential, there remains a possibility of relapse following resection, with recurrence rates estimated to range between 3% and 9%.[10,11] The case of this 77-year-old woman highlights the need for continued surveillance even after apparently successful surgical resection of the primary tumor, as recurrence can occur years later, evidenced by her presentation in 2021, fourteen years after initial surgery. At the time of identification of recurrent disease, the disease was locally advanced and precluded opportunity for surgery, however likewise created challenges for definitive radiation therapy. Definitive radiation therapy is difficult to perform when disease involves the gastric region as the risk of perforation is significant when disease extends through the serosa into the mucosa as response can expose fistula formation with perforation.

Typically, pancreatic cancer metastasizes or recurs in sites such as the liver, lymph nodes, peritoneum, and lungs.[12–14] Although instances of direct invasion by pancreatic cancer into the stomach have been sporadically reported, gastric extension from a primary or recurrent pancreatic cancer remains an uncommon occurrence.[15,16] Of 209 autopsies of pancreatic cancer patients, Oda et al. found only 2 instances of gastric metastasis (0.96%).[15,17] Gastric involvement from pancreatic cancer may be due to hematogenous or lymphatic metastasis, intraluminal or intramural dissemination, intraoperative seeding, or direct invasion.[13,17,18] In this case, recurrent disease was an extension of the recurrence as seen on diagnostic imaging.

The treatment strategy for locally advanced recurrent SPT with direct extension in the gastric wall warrants careful consideration, weighing the benefits and risks of various therapeutic modalities. The recurrence and extension into gastric mucosa created a vulnerable situation where both the disease and response to therapy could create a clinically precarious situation, potentially generating the need for urgent intervention. In this case, neoadjuvant chemotherapy was initiated with gemcitabine and nab-paclitaxel to reduce tumor burden prior to considering re-visiting surgical resection, recognizing that the relationship to vascular structures including the portal vein and superior mesenteric artery would make resection unlikely. Though complete resection of the recurrent tumor is usually the preferred treatment of choice for primary and recurrent SPT when feasible, surgical involvement was deferred due to concerns about vascular/retroperitoneal involvement and the risk of surgical complications in this patient given her age and medical status.[19,20] As an alternative, the patient underwent radiation therapy targeting the entire pancreatic recurrence including the gastric component with margin as well as additional tissues considered at risk for disease. Multiple case reports have documented the benefits of using radiotherapy to reduce the size of these lesions.[6,21] Zauls et al. reported a case of a 33-year-old woman with an unresectable SPT who underwent IMRT alone resulting in decreased tumor volume and symptom control.[22] On serial imaging after the initial phase of radiation management in this patient, there appeared to be a cleavage plane between the gastric component and recurrent pancreatic tumor providing an opportunity to approach the situation in a more creative manner than anticipated. If the gastric component was irradiated at the same dosage as the pancreas, there would be a high likelihood of perforation. The literature examining dose restraints for the stomach are heterogenous, however there is a consensus across studies, acknowledging the significance of this concern.[23-25] In a case study by Furman et al., a patient treated with stereotactic body radiation therapy for liver metastasis received 50 Gy in 10 fractions, resulting in gastric perforation, presumably due to radiation dose exposure to the expanded planning target volume within the radiation field, which included the portion of the gastric wall, which was adherent to the resected portion of the liver.[26,27] Therefore, in this patient, following radiation therapy, which included an additional boost to the region of the pancreatic recurrence that could not be resected, surgical resection of the gastric

component of disease was performed to achieve tumor control and mitigate the risk of perforation.

Conclusion

In the management of recurrent disease, often the clinical team requires a unique approach to address potential risks of management in a controlled manner. In this circumstance, surgery played a crucial role in mitigating potential damage often associated with radiation therapy. The pace of the disease permitted a step-by-step approach to the care of the patient, optimizing her care with the strengths of each discipline involved.

The patient remains well and to date has the benefits of management with no visible sequelae noted. The case represents a unique multidisciplinary approach to problem solving in a clinically challenging situation.

Conflicts of Interest Statement

The authors have no conflicts of interest to declare.

References

1. Reddy S, Cameron JL, Scudiere J, 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(5):950-957. doi:10.1016/j.jamcollsurg.2009.01.044
2. Younan G. Pancreas solid tumors. *Surg Clin North Am*. 2020;100(3): 565-580. doi: 10.1016/j.suc.2020.02.008
3. Karakas S, Dirican A, Soyer V, Koç S, Ersan V, Ates M. A pancreatic pseudopapillary tumor enucleated curatively. *Int J Surg Case Rep*. 2015;10:118-120. doi:10.1016/j.ijscr.2015.03.040
4. Lubezky N, Papoulas M, Lessing Y, et al. Solid pseudopapillary neoplasm of the pancreas: management and long-term outcome. *Eur J Surg Oncol*. 2017;43(6): 1056-1060. doi:10.1016/j.ejso.2017.02.001
5. Flores RL, Rossi R, Castiblanco A, Gallardo A, Schiappacasse G. Solid bifocal pseudopapillary neoplasm of the pancreas: a case report. *Int J Surg Case Rep*. 2021;84: 106131. doi:10.1016/j.ijscr.2021.106131
6. Tanoue K, Mataki Y, Kurahara H, et al. Multidisciplinary treatment of advanced or recurrent solid pseudopapillary neoplasm of the pancreas: three case reports. *Surg Case Rep*. 2022;8(1):7. doi:10.1186/s40792-022-01358-0
7. Kang CM, Kim KS, Choi JS, Kim H, Lee WJ, Kim BR. Solid pseudopapillary tumor of the pancreas suggesting malignant potential. *Pancreas*. 2006;32(3): 276-280. doi:10.1097/01.mpa.0000202956.41106.8a
8. Eder F, Schulz HU, Röcken C, Lippert H. Solid-pseudopapillary tumor of the pancreatic tail. *World J Gastroenterol*. 2005;11(26):4117-4119. doi:10.3748/wjg.v11.i26.4117
9. Stefanova N, Kalinov T, Kolev N, Kalchev E. Frantz tumor: a case report of solid pseudopapillary tumor of pancreas. *Cureus*. 2023;15(7):e41698. doi:10.7759/cureus.41698
10. Law JK, Ahmed A, Singh VK, et al. A systematic review of solid-pseudopapillary neoplasms: are these rare lesions? *Pancreas*. 2014;43(3):331-337. doi:10.1097/MPA.0000000000000061
11. Perez A, Arcilla C, Fontanilla MRK, Berberabe AE. Resection of a recurrent solid pseudopapillary neoplasm of the pancreas after duodenal sparing pancreaticoduodenectomy: a case report. *Int J Surg Case Rep*. 2021;88: 106526. doi:10.1016/j.ijscr.2021.106526
12. Blastik M, Plavecz E, Zalatnai A. Pancreatic carcinomas in a 60-year, institute-based autopsy material with special emphasis of metastatic pattern. *Pancreas*. 2011;40(3): 478-480. doi:10.1097/MPA.0b013e318205e332
13. Umezaki N, Hashimoto D, Nakagawa S, et al. Cystic gastric metastasis from pancreatic cancer. *Surg Case Rep*. 2018;4(1):31. doi:10.1186/s40792-018-0443-2
14. Yamada Y, Sasaki T, Takeda T, et al. Multiple gastric metastases after distal pancreatectomy for pancreatic cancer. *Intern Med*. 2022;61(18):2741-2746. doi:10.2169/internalmedicine.8848-21
15. Oda, Kondo H, Yamao T, et al. Metastatic tumors to the stomach: analysis of 54 patients diagnosed at endoscopy and 347 autopsy cases. *Endoscopy*. 2001;33(6):507-510. doi:10.1055/s-2001-14960
16. Yang J, Yuan Y, Zhang S, Lv Y. Gastric metastasis from pancreatic cancer characterized by mucosal erosion: a case report and literature review. *Journal of International Medical Research*. 2021;49(4). doi:10.1177/03000605211003759
17. Sasajima J, Okamoto K, Taniguchi M. Hematogenous gastric metastasis of pancreatic cancer. *Case Rep Gastroenterol*. 2016;10(1):75-80. doi:10.1159/000444249
18. Feczko PJ, Collins DD, Mezwa DG. Metastatic disease involving the gastrointestinal tract. *Radiol Clin North Am*. 1993;31(6):1359-1373. doi:10.1016/S0033-8389(22)00325-6
19. Gao H, Gao Y, Yin L, et al. Risk factors of the recurrences of pancreatic solid pseudopapillary tumors: a systematic review and meta-analysis. *J Cancer*. 2018;9(11):1905-1914. doi:10.7150/jca.24491
20. Strauss JF, Hirsch VJ, Rubey CN, Pollock M. Resection of a solid and papillary epithelial neoplasm of the pancreas following treatment with cis-platinum and 5-fluorouracil: a case report. *Med Pediatr Oncol*. 1993;21(5):365-367. doi:10.1002/mpo.2950210511
21. Sibio S, Di Carlo S. Current highlights on solid pseudopapillary neoplasm of the pancreas. *World J Hepatol*. 2022;14(1):300-303. doi:10.4254/wjh.v14.i1.300
22. Zauls JA, Dragun AE, Sharma AK. Intensity-modulated radiation therapy for unresectable solid pseudopapillary tumor of the pancreas. *Am J Clin Oncol*. 2006;29(6):639-640. doi:10.1097/01.coc.0000190457.43060.fd
23. Zheng Y, Gao W, Spratt DE, Sun Y, Xing L. Management of gastrointestinal perforation related to radiation. *Int J Clin Oncol*. 2020;25(6):1010-1015. doi:10.1007/s10147-020-01662-5
24. Meyer JE, Kharofa J. The role of dose escalation in pancreatic cancer: go big or go home? *Int J Radiat Oncol Biol Phys*. 2023;115(2):395-397. doi: 10.1016/j.ijrobp.2022.09.050
25. Lee D, Komatsu S, Terashima K, et al. Surgical spacer placement for proton radiotherapy in locally advanced pancreatic body and tail cancers: initial clinical results. *Radiat Oncol*. 2021;16(1):3. doi: 10.1186/s13014-020-01731-z
26. Thomas TO. Hasan S, Small W Jr, et al. The tolerance of gastrointestinal organs to stereotactic body radiation therapy: what do we know so far? *J Gastrointest Oncol*. 2014;5(3):236-246. doi:10.3978/j.issn.2078-6891.2014.024
27. Furman MJ, Whalen GF, Shah SA, Kadish SP. Gastric perforation following stereotactic body radiation therapy of hepatic metastasis from colon cancer. *Pract Radiat Oncol*. 2013;3(1):40-44. doi:10.1016/j.prro.2012.03.005