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Case of Solid Pseudopapillary Neoplasm of Pancreas

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Aim: to discuss the clinical presentation, management, and outcome of the clinical case of solid pseudopapillary tumour of the pancreas (SPTP).
Key points. SPTPs are rare tumours known for their low malignant potential. They are predominantly found in younger females, typically occurring in the tail of the pancreas. Surgical resection of SPTP with negative margins is associated with improved survival rates and lower chances of recurrence. In this study, we discuss the case of a 69-year-old woman diagnosed with an SPTP originating from the tail of the pancreas, which had spread to the hilum of the spleen. A radical resection was performed, successfully obtaining negative surgical margins. However, the patient subsequently developed metastasis to the liver and lungs, prompting the initiation of systemic chemotherapy. The patient died seven months after the development of metastasis and 39 months following the primary diagnosis. This case underscores that, despite initial successful surgical outcomes, high-risk histopathological features can lead to recurrence and metastasis, necessitating reassessment of treatment strategies.
Conclusion. Negative surgical margins are critical for a favourable prognosis in the treatment of SPTPs; however, close monitoring for potential recurrence is essential. Our findings highlight that even after achieving negative margins, patients with high-risk features should undergo rigorous follow-up and consider postoperative systemic chemotherapy. This clinical case emphasizes the complexity of managing SPTP and the need for individualized treatment strategies, as early detection of recurrence can significantly influence survival outcomes.
Keywords: solid pseudopapillary tumor, pancreas, metastasis, surgery, resection
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Солидно-псевдопапиллярная опухоль поджелудочной железы (клиническое наблюдение)

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Цель: писать клиническую картину, лечение и исход случая солидной псевдопапиллярной опухоли (СППО) поджелудочной железы.
Основные положения. СППО поджелудочной железы — редкие опухоли с низким злокачественным потенциалом, которые чаще всего встречаются у молодых женщин и обычно располагаются в хвосте поджелудочной железы. Хирургическая резекция СППО с негативными хирургическими краями связана с улучшением выживаемости и снижением вероятности рецидива. В данном исследовании мы рассматриваем случай лечения 69-летней женщины с СППО, возникшей в хвосте поджелудочной железы и распространившейся на ворота селезенки. Пациентке была проведена радикальная резекция с получением негативных хирургических краев. Однако впоследствии у нее развились метастазы в печень и легкие, что побудило начать системную химиотерапию. Пациентка умерла через семь месяцев после развития метастазов и через 39 месяцев после постановки первичного диагноза. Этот случай подчеркивает, что, несмотря на успешное хирургическое вмешательство, наличие гистопатологических признаков высокого риска может привести к рецидиву и метастазам, что требует пересмотра подходов к лечению.
Заключение. Негативные хирургические края имеют ключевое значение для благоприятного прогноза при лечении СППО. Тем не менее необходим тщательный мониторинг на предмет возможного рецидива. Наши результаты показывают, что даже после резекции с негативными краями пациенты с высокими рисками должны находиться под строгим наблюдением и для них должна рассматриваться возможность послеоперационной системной химиотерапии. Этот клинический случай подчеркивает сложность лечения СППО

поджелудочной железы и необходимость индивидуализированных стратегий, поскольку раннее выявление рецидива может значительно повлиять на результаты выживания.
Ключевые слова: солидно-псевдопапиллярная опухоль, поджелудочная железа, метастазы, хирургия, резекция
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Introduction

Solid pseudopapillary tumors of the pancreas (SPTP) are rarely seen tumors that have low malignant potential and are usually located at the tail of the pancreas [1]. This kind of neoplasms generally affects young women, and even though there are significant developments in imaging technology, the gold standard methods of diagnosis are histopathological and immunohistochemical examinations [2]. Resection of SPTP with negative surgical margins provides longer survival and lower recurrence rates [3].

An SPTP was first described as the “tumor of Frantz” by V.K. Frantz in 1959, and there is limited information about these malignancies in the literature [4]. SPTPs are listed as “rarely seen low-grade malignant tumors” in the Classification of Tumors of Digestive System by the World Health Organization (WHO) [5]. Even though the prevalence of SPTPs has increased with the developments in radiological viewing, there is still limited data regarding the etiopathology and clinical course of the disease, which is open to research [1].

In this study, we aim to report a case of surgical treatment of a pancreas tumor diagnosed as SPTP, which metastasized and led to the death of the patient.

Case report

Clinical course

A 69-year-old woman was admitted to the general surgery clinic with abdominal pain and bloating. Both her medical history and physical examination were unremarkable. Approximately five months prior, the patient had received treatment in internal medicine department for anemia and gastritis, during which she did not complain of any abdominal pain.

Radiology

Abdomen and pelvic computed tomography (CT) with intravenous contrast revealed a solid tumor measuring 13.0 × 10.5 cm, originating from the tail of the pancreas and extending to the hilum of the spleen (Fig. 1a). The tumor described as displacing the left kidney both cranially and caudally, and it exhibited a heterogeneous internal structure with areas of focal necrosis. In the paraaortic and paracaval regions adjacent to the greater curvature of the stomach, some lymph nodes measuring up to 8 mm were noted at the level of the renal artery. The CT scan did not reveal any metastatic lesions in the thorax.

Surgery

The patient underwent surgery following laboratory findings and tumor marker levels falling within the normal range, leading to an early diagnosis of a distal pancreatic tumor. A conventional midline incision was performed. Intraoperatively, the tumor was determined to originate from the tail and body of the pancreas and was found to be invasive to the spleen and its hilum. It extended towards the left kidney and retroperitoneum, resulting in occlusion of the splenic vein, which contributed to left portal hypertension and the formation of multiple collateral veins. An en-bloc resection of the tumor, along with the spleen, was performed, ensuring a clear surgical margin of 2 cm in the pancreas.

Pathology

Macroscopic examination of the resected specimens revealed the spleen measured 24 × 17 × 6 cm and the pancreas measured 8 × 6 × 5 cm. Serial sections identified a tumor located in the tail of the pancreas, extending into the hilum of the spleen, with dimensions of 13 × 9 × 7 cm. A thick capsule formation was noted around the portion of the tumor associated with the pancreas. The tumor tissue displayed a heterogeneous composition, containing firm areas alongside soft, spongy, and necrotic regions.

Histological examination revealed that the tumor invaded the spleen, exhibiting heterogeneous structure characterised by solid and cystic areas as well as papillary components with hyalinized vascular pedicle. The tumor cells were monotonous and displayed medium-level eosinophilic cytoplasm, containing hyaline globules (PAS positive and d-PAS positive) and perinuclear vacuoles (Fig. 2a). Nuclear pleomorphism among the tumor cells was minimal. Additionally, signs of ischemia, hemorrhage, cholesterol clefts, and

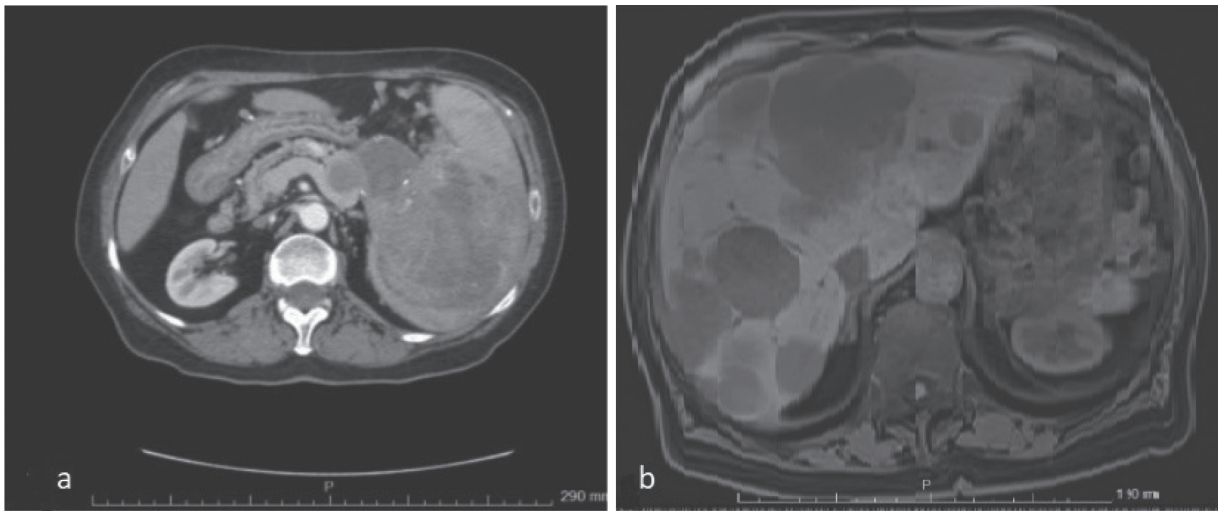

Figure 1. Preoperative CT imaging, revealing a solid pseudopapillary pancreatic tumor (a) and postoperative MRI, revealing metastatic liver lesions (b)

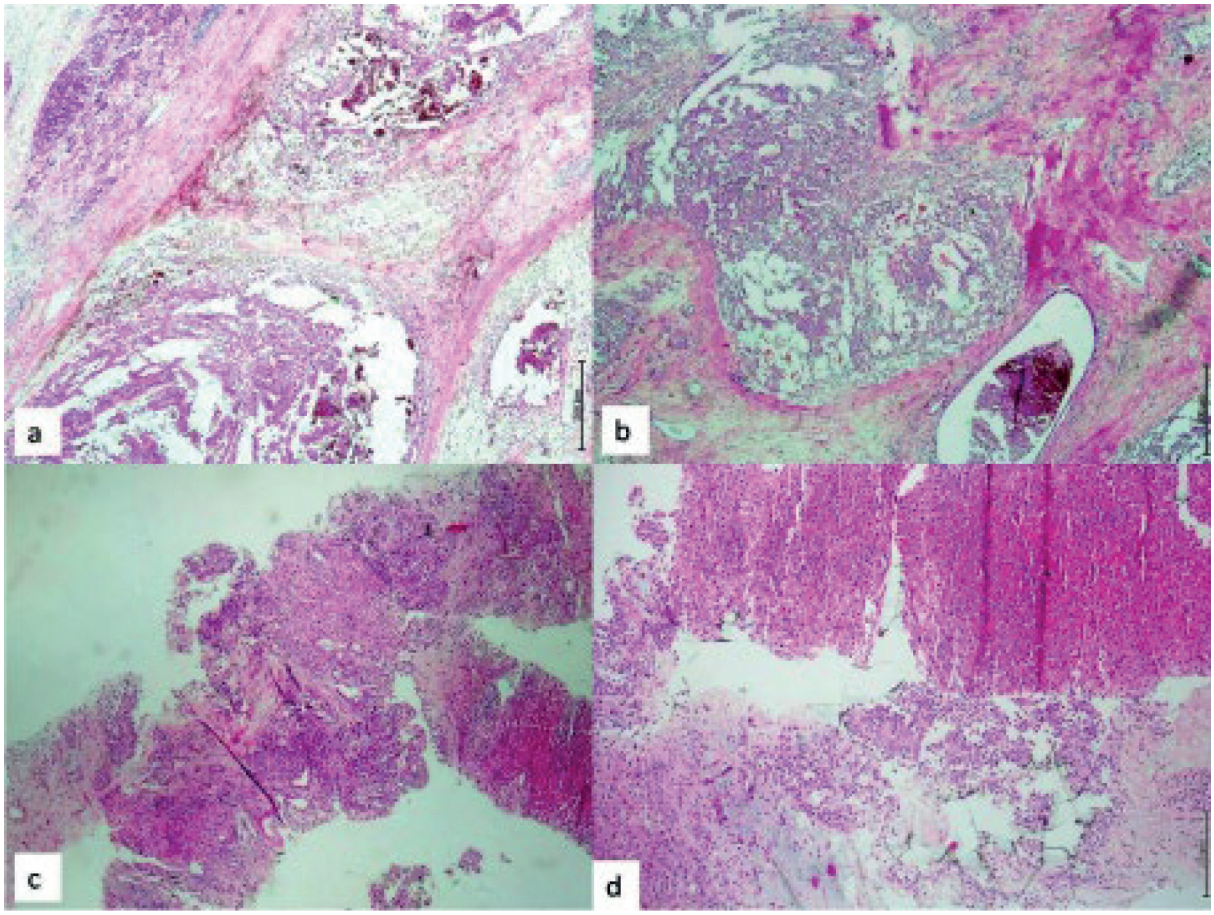

Figure 2. Histologic view of a pancreatic tumor showing papillary structures with solid and cystic areas (a–b); appearance of liver metastasis exhibiting morphology similar to that in the pancreas (c–d)

histiocytic aggregations were noted. The stroma exhibited degenerative changes, including hyalinization, hemorrhage, foamy macrophages, calcification, and cholesterol clefts (Fig. 2b). Two mitotic figures were identified in 10 high-power fields. There were no findings of perineural or lymphovascular invasion, and no tumor was detected at the surgical margins. During the surgery, three lymph nodes were excised, which exhibited reactive characteristics. Immunohistochemical examination revealed that tumor cells' cytoplasm was positive for synaptophysin, CD10, vimentin, and chromogranin-A, while both cytoplasm and nuclei showed positivity for β-catenin. The case was diagnosed as "solid pseudopapillary tumor of pancreas".

Follow-up

The patient was classified as high risk according to the WHO criteria, which identify poor prognostic factors such as a tumor size greater than 5 cm and invasion into adjacent organs. Additionally, the patient was enrolled in a close follow-up program based on the Fudan prognostic criteria, which also consider tumor size and the Ki67 index. This monitoring is essential for the prompt identification and management of any potential metastasis. Chemotherapy was initiated, with capecitabine administered at a dose of 2000 mg/m^2 every 21 days.

The patient was scheduled for follow-up every three months, during which abdominal and thoracic CT imaging was performed. At the check-up appointment after 23 months, no pathological findings were observed. However, abdominal magnetic resonance imaging (MRI) conducted 32 months posts-operation revealed multiple metastatic solid lesions, measuring up to 5 cm in size, located at the junction of liver segments 8 and 4A. These lesions appeared separate but tended to connect with each other and exhibited restricted diffusion (Fig. 1b). Tru-cut biopsy revealed a tumor that resembled the primary tumor in both immunohistochemical expression and morphology, leading to a diagnosis of "solid pseudopapillary tumor of the pancreas" (Fig. 2c, d).

The case was subsequently evaluated by the Council of Oncologic Tumors, which agreed on a chemotherapy regimen for the patient. She chose to continue chemotherapy treatment at another hospital, resulting in limited data regarding the chemotherapy regimen administered during that time. Despite ongoing chemotherapy, the liver lesions showed progression, and new metastases were identified in the lung and bone tissue. The patient died seven months after the development of metastasis and 39 months following the primary diagnosis.

Discussion

The limited information regarding this rare anatomic-clinical situation, coupled with the low number of documented cases, restricts our understanding of solid pseudopapillary tumors of the pancreas (SPTP). An increase in the reporting of SPTP cases, particularly the rare forms with poor prognosis, can enhance our knowledge from etiopathological, histopathological, and clinicopathological perspectives. This, in turn, may lead to updated treatment models and a better understanding of the disease. SPTPs are characterized by low malignant potential, and survival rates remain significantly high when surgical margins are negative. However, the diagnostic criteria, etiology, and factors contributing to malignancy in SPTP are still not well understood. Postoperative recurrence and metastasis are relatively uncommon [6, 7]. M. Del Chiaro et al. identified several poor prognostic factors, including tumor size greater than 5 cm, male sex, presence of necrosis, cellular atypia, vascular and perineural invasion, as well as invasion of adjacent tissues [8]. In our case, despite the negative surgical margin, splenic invasion necessitated close surveillance. Consequently, liver metastasis was identified in the short-term, aligning with findings in the literature.

SPTPs are typically found in the tail of the pancreas, although they can occur throughout the entire organ. There have been rare reports of SPTP cases in extra-pancreatic locations, including the ovary, mesentery, and adrenal gland [9]. In our case, the tumor was located in the tail of the pancreas, which is consistent with the findings documented in the literature.

The primary treatment for SPTP is surgical intervention. In case of irresectable or metastatic disease, radiotherapy or systemic treatment options can be considered; however, standardized models for systemic treatment are still lacking. Additionally, there is literature available on the use of mTOR inhibitors, such as everolimus, and multitarget tyrosine kinase inhibitors, including sunitinib, as potential method of treatment [9].

In our case, systemic treatment was not initially an option because radical surgery was performed with negative surgical margins. However, after the occurrence of metastasis, systemic treatment in the form of chemotherapy was administered. The chemotherapy regimen was adjusted due to disease progression. It has been reported that surgery following the onset of metastasis can improve survival rates. In cases of irresectable or isolated liver metastasis, liver transplantation may be considered as an alternative treatment option

[10]. In our situation, the patient was not a candidate for surgical excision due to generalized liver metastasis. A liver transplant was not pursued because of disease progression, the patient's age, and the uncertain outcomes associated with treatment models.

In a review of 1,384 SPTP cases, G. Mazzarella et al. reported that surgeries conducted with negative surgical margins are associated with high cure rates and lower recurrence rates [9]. However, in our situation, contrary these expectations, the patient's advanced age, presence of multicentric metastasis, limited survival duration, and lack of successful treatment despite negative surgical margins render this case particularly interesting and worthy of discussion.

Conclusion

A negative surgical margin is crucial in determining the prognosis for patients with pseudopapillary tumors of the pancreas. In patients exhibiting high-risk histopathological features, distant metastasis may still occur even after excision with a negative surgical margin. Therefore it is advisable to regard SPTP as a potentially poor prognostic factor and to inform patients about this possibility.

Patients at risk for clinical and histopathological recurrence and metastasis can be diagnosed at an early stage through close follow-up, which may prolong their lifespan with appropriate treatment [11]. Even when excision with a negative surgical margin has been achieved, postoperative systemic chemotherapy should be considered.

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