Association between Solid Pseudopapillary Pancreatic Tumor (Frantz's Tumor) and Left Portal Hypertension in Pediatrics: Case Report

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Associação entre Tumor Sólido Pseudopapilar Pancreático (Tumor de Frantz) e Hipertensão Portal Esquerda em Pediatria: Relato de Caso

Asociación entre Tumor Sólido Pancreático Pseudopapilar (Tumor de Frantz) e Hipertensión Portal Izquierda en Pediatría: Informe de Caso

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ABSTRACT

Introduction: The solid pseudopapillary tumor of the pancreas is a rare pathology, accounting for less than 3% of all pancreatic exocrine tumors. This tumor occurs mainly in young women, between 20 and 30 years old, has low malignant potential, causing symptoms only when they are large. **Case report:** Female patient, 14 years of age, presented hematemesis and varicose veins in the gastric fundus. After submitting to computed tomography, a hypodense mass of hypovascular behavior was shown in the pancreatic tail, raising the suspicion of primary pancreatic neoplasia promoting compression of the splenic vein. The patient underwent body-caudal pancreatectomy associated with splenectomy in February 2021. The biopsy of the hepatic nodule, evidenced on abdominal ultrasound, confirmed metastasis and the patient underwent subsequent hepatic segmentectomy in July 2021. **Conclusion:** The case reported is extremely rare because it is a Frantz tumor in childhood associated with left portal hypertension due to tumor compression of the splenic vein, which consequently presented upper digestive hemorrhage as an initial clinical manifestation; similar cases are not easily found in the literature in the pediatric age range.
Key words: pancreas; pediatrics; hypertension, portal.

RESUMO

Introdução: O tumor sólido pseudopapilar do pâncreas é uma patologia rara responsável por menos de 3% de todos os tumores exócrinos pancreáticos. Tal tumor ocorre principalmente em mulheres jovens, entre 20 e 30 anos, e geralmente possui baixo potencial de malignidade e causa sintomas apenas quando são de grandes volumes. **Relato do caso:** Paciente feminina, 14 anos, iniciou quadro de hematêmese, sendo identificadas varizes de fundo gástrico. Após realizar tomografia computadorizada, evidenciou-se massa hipodensa de comportamento hipovascular na cauda pancreática, levantando-se a suspeita de neoplasia pancreática primária, promovendo compressão da veia esplênica. A paciente foi submetida à pancreatectomia corpo-caudal associada à esplenectomia em fevereiro de 2021. A biópsia do nódulo hepático, evidenciado em ultrassonografia abdominal, confirmou metástase, e a paciente foi submetida à posterior segmentectomia hepática em julho de 2021. **Conclusão:** O caso relatado é extremamente raro por tratar-se de um tumor de Frantz na infância associado à hipertensão portal esquerda por compressão tumoral da veia esplênica e que, por consequência, apresentou hemorragia digestiva alta como manifestação clínica inicial, não sendo encontrados facilmente na literatura casos similares na faixa pediátrica.
Palavras-chave: pâncreas; pediatria; hipertensão portal.

RESUMEN

Introducción: El tumor sólido pseudopapilar de páncreas es una patología rara, representando menos del 3% de todos los tumores pancreáticos exocrinos. Este tumor se presenta principalmente en mujeres jóvenes, entre 20 y 30 años, generalmente tiene un bajo potencial maligno y causa síntomas sólo cuando son de gran tamaño. **Informe del caso:** Paciente femenino de 14 años de edad, inició con hematemesis y se identificaron varices en fondo gástrico. Tras realizar tomografía computarizada se evidenció una masa hipodensa de comportamiento hipovascular en la cola pancreática, lo que hizo sospechar de neoplasia pancreática primaria promoviendo la compresión de la vena esplénica. La paciente fue sometida a pancreatectomía cuerpo-caudal asociada a esplenectomía en febrero de 2021. La biopsia del nódulo hepático, evidenciada en ecografía abdominal, confirmó metástasis y la paciente fue sometida a posterior segmentectomía hepática en julio de 2021. **Conclusión:** El caso reportado es extremadamente raro por tratarse de un tumor de Frantz en la infancia asociado a hipertensión portal izquierda por compresión tumoral de la vena esplénica y que, en consecuencia, presentó como manifestación clínica inicial hemorragia digestiva alta, no encontrándose fácilmente en la literatura casos similares en el rango de edad pediátrica.
Palabras clave: páncreas; pediatría; hipertensión portal.

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INTRODUCTION

Solid pseudo-papillary tumor of the pancreas known as Frantz's tumor is a rare pathology accounting for less than 3% of all exocrine pancreatic tumors[1,2]. These tumors are of low malignant potential causing symptoms when large, the most common manifestations are pain or abdominal discomfort, jaundice, compressive symptoms and palpable masses[2-4].

Still more rare is the association with sinistral portal hypertension, a disease caused by the intrinsic or extrinsic obstruction of the splenic vein, leading to the development of gastric varices[4].

The objective of this article is to report a rare case of Frantz's Tumor associated with left-sided portal hypertension of an adolescent referred to a reference hospital in Santa Catarina, Brazil. In addition, it attempts to demonstrate the importance of a thorough study of the clinical status of the patient due to the rarity of the presentation of portal hypertension and disclose to health professionals the pathology of this case.

The Institutional Review Board of the institution approved the study, report number 4872561 (CAAE (submission for ethical review): 48498721.1.0000.5361) in compliance with ethical guidelines for studies with human beings and securing the anonymity of the information according to Directive 466/12[5] of the National Health Council.

CASE REPORT

Female adolescent, 14 years of age, sought assistance at the outpatient pediatric gastroenterology due to hematemesis in August 2020.

The upper digestive endoscopy was performed promptly revealing varices of the gastric fundus associated with erythematous flat rashes. Hematemesis stopped after she initiated treatment with pantoprazole and propranolol. Her legal representative reported abdominal pain due to difficulty to evacuate, no weight loss or melena. At the admission physical exam, the patient was in good clinical condition, rosy cheeks without weight loss. Abdomen was flaccid, painless, without visceromegaly or palpable masses.

Abdominal ultrasound was performed in November 2020, revealing homogeneous splenomegaly (13.3 cm) with hilar collateral circulation in addition to hepatic node in transition of segments VII and VIII measuring 3.2 x 3.1 x 2.9 cm. Computed tomography (CT) on January 2021 showed hypodense mass of hypovascular behavior at the pancreatic tail, raising the suspicion of primary pancreatic neoplasm, compressing the splenic vein and consequently, splenomegaly and gastric varices in the esophagogastric transition also identified by the exam. New upper gastric endoscopy performed in January 2021 revealed gastric fundus varices once again and initiated follow-up with the oncology pediatric team in the same month.

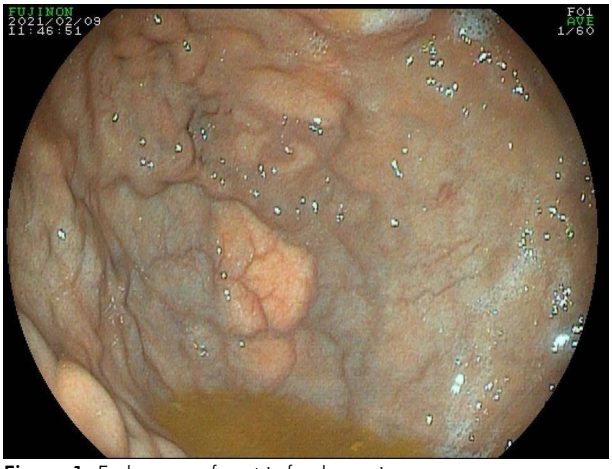

Figure 1. Endoscopy of gastric fundus varices

The patient was submitted to corpora-caudal pancreatectomy with splenectomy in February 2021. Intraoperatory findings showed tumor at the pancreas tail responsible for the enlargement of splenic vein, splenic hilar vessels and short gastric vessels.

The anatomopathology of the surgical piece confirmed solid pseudopapillary neoplasm with metastasis to two of the 15 lymph nodes submitted to biopsy. Immunohistochemistry presented expression of beta-catenin, CD56, cyclin D1, focal synaptophysin, SOX-11 and LEF1, confirming pancreatic pseudopapillary tumor. The patient remained hospitalized for 11 days and was discharged nine days post-surgery without complications. The biopsy of hepatic node confirmed metastasis and the patient was submitted to liver posterior segmentectomy in July 2021.

DISCUSSION

Solid pseudopapillary tumor described for the first time by V.K. Frantz in 1959[1] is rare disease responsible for less than 3% of pancreas neoplasms having occurred in only 700 cases in the world, of which 25% in pediatric patients[1,3].

It has a strong predilection for women (10:1) with less than 30 years of age, the mean pediatric age is 9.5 years[2,3]. This pattern for adult females is similar to pediatric patients, but gender ratio is lower (2:1)[2].

Quite often, signs and symptoms are unspecific as abdominal pain, nausea, vomits, jaundice and palpable masses at physical examination[2].

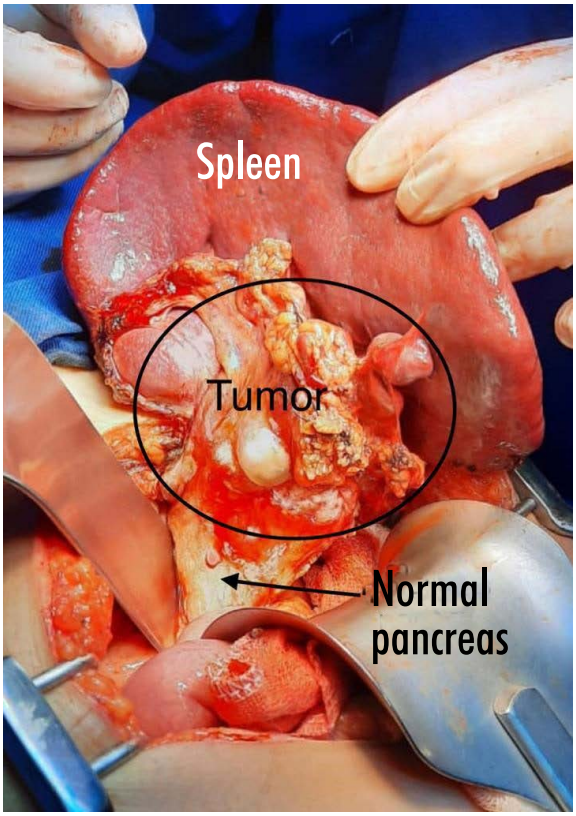

Figure 2.Surgery showing the position of the tumor and relation with adjacent structures

Related publications are scarce in Brazil and most of the available information consists in case reports, with poor epidemiological information of the tumor. This scenario can be attributed to the rarity of these tumors in the country, clearly the main cause for paucity of comprehensive epidemiological analyzes.

Initially, the patient presented upper gastric hemorrhage and similar cases in pediatric patients involving pancreatic neoplasm and upper hemorrhage are rarely found in the literature. A similar condition was reported for an adult patient with compromise of the splenic vein and appearance of gastric fundus varices but without hematemesis as clinical manifestation[6].

The tumor in question compressed the splenic vein causing blood reflux to the splenic hilum and short gastric veins as alternative venous drainage route of the portal system to the azygous system, causing similar symptoms of portal hypertension, including gastric fundus varices and hematemesis. The main cause of this phenomenon would be the intrinsic (thrombosis) or extrinsic obstruction of the vein (pancreatic mass)[7].

Isolated gastric fundus varices associated with splenomegaly and normal liver function should raise the hypothesis of sinistral or left-sided portal hypertension, a rare cause of upper gastric hemorrhage (<1%)[7-9].

Computed tomography (CT) is essential to identify the location of the mass, presence of portal hypertension, vascular involvement and relation with adjacent structures[4]. Diagnostic confirmation of the tumor depends on histopathology and immunohistochemistry[2]. The most common histochemistry findings are positive chromogranin and synaptophysin. The patient presented a positive immunohistochemical pattern for beta-catenin, CD56, cyclin D1, focal synaptophysin , SOX-11 and LEF1.

This disease has good prognosis, except in males, patients with expression of nuclear protein Ki-67 and tumor larger than 5 cm[2]. Metastases occur in 10-15% of advanced cases, the main sites are liver, peritoneum, omentum and regional lymph nodes[2,3].

Surgery is the curative treatment in more than 95% of the cases[2], the most common is pancreatectomy as described for the case. Lymphadenectomy is indicated for tumors larger than 5 cm due to malignancy risk[2].

The presence of portal hypertension can be aggravating at surgery, however with early preoperative vascular reconstruction and intraoperatory caution, resection can cause minimal blood loss[6]. In cases of sinistral portal hypertension with gastric bleeding, the treatment recommended is to associate splenectomy to decompress the portal venous system. In less severe cases, embolization of the splenic artery is effective[7,9].

CONCLUSION

The case report of a Frantz's tumor in a pediatric patient stands out due to its clinical rare presentation. The left portal hypertension triggered by tumor compression of the splenic vein associated with upper gastric hemorrhage as initial clinical manifestation was treated with successful pancreatectomy as occurs in most of the cases. Share and disseminate the knowledge acquired with the case are actions that help other medical professionals and enrich the understanding of this specific condition.

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CONTRIBUTIONS

All the authors contributed equally to all the stages of the article and approved the final version to be published.

DECLARATION OF CONFLICT OF INTERESTS

There is no conflict of interests to declare.

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None.

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