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PRACA KAZUISTYCZNA
CASE STUDIES

Frantz tumor – a rare cause of abdominal pain in children – case report

Guz Frantz rzadką przyczyną bólu brzucha u dzieci – opis przypadku

Małgorzata Piechota-Mikuła[1], Lidia Strzelczuk-Judka[1], Przemysław Mańkowski[2], Katarzyna Jończyk-Potoczna[1]

[1] Zakład Radiologii Pediatrycznej Katedry Radiologii Ogólnej i Zabiegowej Szpitala Klinicznego im. Karola Jonschera Uniwersytetu Medycznego im. Karola Marcinkowskiego w Poznaniu

[2] Katedra i Klinika Chirurgii, Traumatologii i Urologii Dziecięcej Szpitala Klinicznego im. Karola Jonschera Uniwersytetu Medycznego im. Karola Marcinkowskiego w Poznaniu

ABSTRACT

The Frantz tumor (solid pseudopapillary neoplasm – SPN) is a rare, usually benign tumor of unclear etiology having a predilection for young women.
This usually asymptomatic tumor may, however, be a cause of abdominal pain or discomfort.
In our case, the mentioned tumor was a cause of abdominal pain in a 14-year-old, a so far healthy patient. Abdominal imaging – USG and computed tomography scan (CT) were performed showing a large mass in the left epi- and mesogastrium with primary location probably in the pancreas. A laparoscopic biopsy was performed and confirmed the initial diagnosis. A tumor along with the pancreatic tail were removed by laparotomy without complications.
The case we described shows that a solid pseudopapillary neoplasm can be a rare cause of abdominal pain in pediatric patients.

KEY WORDS
Frantz tumor, pancreas tumor, abdominal pain

STRESZCZENIE

Guz Frantz (lity pseudobrodawkowaty nowotwór trzustki, guz Hamoudiego) jest rzadkim, zwykle łagodnym guzem o nieznanej etiologii, występującym głównie u młodych kobiet. Guz ten jest zazwyczaj bezobjawowy, może być jednak przyczyną bólu lub dyskomfortu ze strony jamy brzusznej.

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Address for correspondence: Lek. med. Lidia Strzelczuk-Judka, Zakład Radiologii Pediatrycznej Szpitala Klinicznego im. Karola Jonschera Uniwersytetu Medycznego im. Karola Marcinkowskiego w Poznaniu, ul. Szpitalna 27/33, 60-572 Poznań, tel./fax (61) 8491-597, e-mail: lidka_strzelczuk@wp.pl

W opisywanym przez nas przypadku guz był przyczyną bólu brzucha u 14-letniej, zdrowej dotychczas pacjentki. W wykonanych badaniach obrazowych – ultrasonografii oraz tomografii komputerowej – uwidoczniono dużą masę guza w nadbrzuszu i śródbrzuszu lewym, którego punktem wyjścia była najprawdopodobniej trzustka. Wykonano biopsję metodą laparoskopową, potwierdzającą wstępne rozpoznanie.
Guz wraz z ogonem trzustki usunięto metodą laparotomii bez powikłań.
Opisany przez nas przypadek pokazuje, iż lity pseudobrodawkowaty guz trzustki może być rzadką przyczyną bólu brzucha u pacjentów pediatrycznych.

SŁOWA KLUCZOWE
bóle brzucha, guz Frantz, guz trzustki

INTRODUCTION

The Frantz tumor (synonyms: solid pseudopapillary neoplasm, SPN, solid pseudopapillary tumor) is an uncommon pancreatic neoplasm with low-metastatic potential, which occurs mainly in young women. The tumor is characterized by good prognosis, slow growth and usually asymptomatic progress for many years [1,2].
SPN should be considered in differential diagnosis for each patient with solid or solid – cystic pancreatic tumors, especially in women younger that 35, including children [3,4].

CASE REPORT

A 14–year–old girl complaining of abdominal pain for about 3 days was admitted to our emergency department. The pain was not associated with defecation or meals, and was specified by the patient as "different" comparing to that usually occurring during menstruation. A physical examination found a palpable mass in the upper left abdomen without other accompanying symptoms.
Ultrasound revealed a large, well-defined, heterogeneous, solid–cystic mass in the upper abdomen and left mesogastrium. Diagnostic imaging of the abdomen was extended to a computed tomography (CT) scan with intravenous contrast administration.
CT showed a large, well demarcated mass lesion measuring 9.0 x 11.5 x 12.3 cm in the left epi- and mesogastrium (Th12–L2 level) containing fluid areas and a single small calcification. After intravenous iodine contrast agent administration, the lesion showed heterogeneous contrast enhancement. The tumor was compressing and dislocating the pancreatic body and tail, contacting and thinning the pancreatic body. The mass lesion was also dislocating the splenic artery and vein, left kidney and left renal vessels, and moving the superior mesenteric artery slightly to the right. No evidence of local invasion or intra-abdominal metastases were found. The CT scan suggested the pancreas as the primary location, and the suspected diagnosis of a solid pseudopapillary neoplasm was given (Fig. 1, 2, 3).

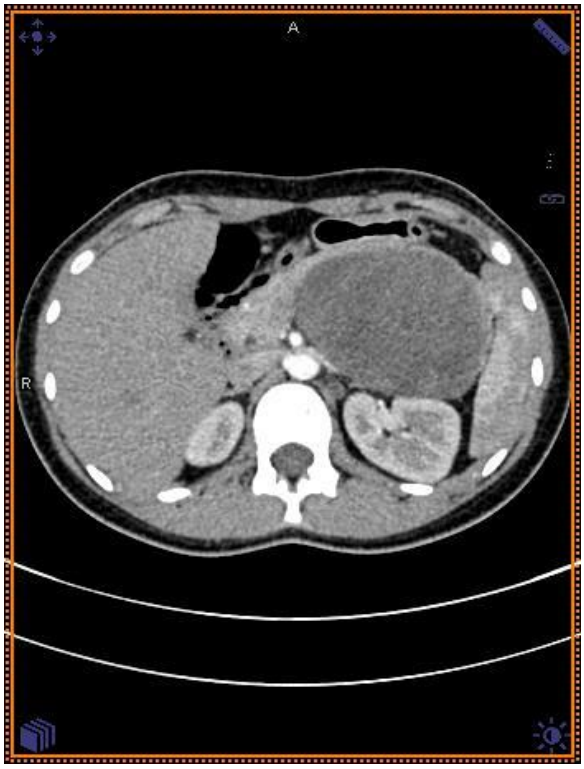

Fig. 1. Abdominal CT with contrast administration.
Ryc. 1. TK jamy brzusznej z kontrastem.

The patient underwent a laparoscopic biopsy, which confirmed the preliminary diagnosis. A few days later, the patient underwent resection of the mass together with the pancreatic tail.
The patient developed well in the postoperative period, with normal ultrasound examinations (1, 2, 4, 6, 8 and 10 months). Follow-up CT scans (4, 5 and 9 months) showed gradually decreasing thrombus in the inferior mesenteric vein, and no evidence of residual disease. The patient is in contact with the Oncology Outpatient Clinic. Informed consent was given by the patient's parents for using clinical data.

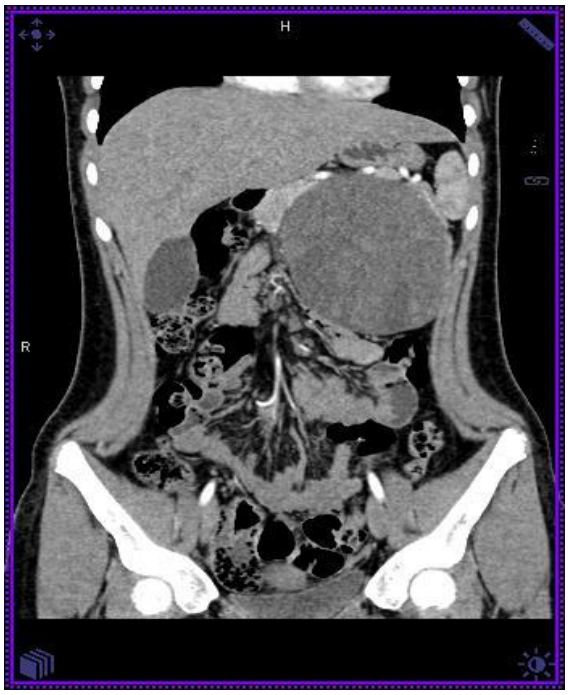

Fig. 2. Preoperative CT, arterial phase, anterio-posterior AP reconstruction: a large mass shaping and moving the body and tail of the pancreas, causing its thinning.
Ryc. 2. Faza tętnicza – widoczna duża masa guza modelująca i przemieszczająca trzon i ogon trzustki powodując jego ścieńczenie.

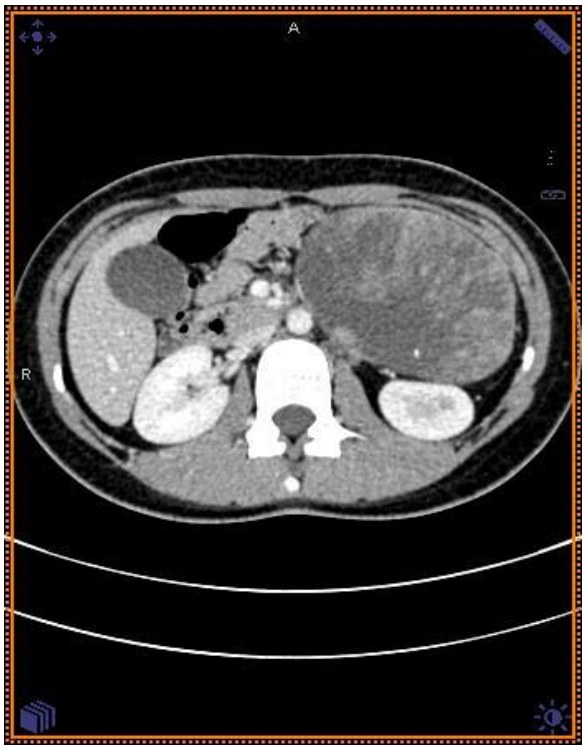

Fig. 3. Preoperative CT, venous phase: heterogeneously enhancing tumor with single calcification.
Ryc. 3. Faza żylna – widoczne niejednorodne wzmocnienie guza oraz pojedyncze ognisko zwapnienia.

DISCUSSION

Pancreatic tumors are rarely considered in the differential diagnosis of solid and cystic abdominal lesions in pediatric patients. The most common abdominal masses diagnosed in the pediatric population include benign lesions and malignancies: neuroblastoma, Wilms' tumor, hepatoblastoma, lymphoma, and germ cell tumors [5]. A solid pseudopapillary neoplasm is a rare tumor of unclear etiology and of 1–2% of exocrine pancreatic neoplasms. It was first described in 1959 by the pathologist Virginia Kneeland Frantz and is often called "Frantz tumor" [1,2,6,7]. Sometimes SPN is also called Hamoudi tumor in honor of the pathologist who in 1970 described the histology of the mentioned tumor.

This rare, commonly benign tumor usually occurs in young women in the 2nd–3rd decade of life, 85% of patients are under 30-years old [8,9,10], although very rare cases in children and men have been reported [4,11]. SPN is generally benign, slow-growing and less aggressive than that of many other pancreatic tumors, and its prognosis is better [12], although very rare forms of malignancy have been reported [13,14]. Then these tumors were more frequently observed in men [6].

Differential diagnosis includes a variety of pancreatic tumors including the non-functioning islet tumour, pancreatoblastoma, acinar cell cancer, mucinous cystic neoplasm, serous cystadenomas, lymphoma pseudocyst, and non-pancreatic tumor, such as pelvic origin lesions extending into the abdomen, including ovarian cysts and teratoma. The only way to confirm diagnosis is biopsy or excision [7,15].

SPN may occur anywhere in the pancreas, but most often involves the head or tail of the pancreas [3]. The tumor mass is usually large, measuring 2–17 cm in diameter (average 9.3 cm), well-demarcated, appears macroscopically as a round lesion with necrotic regions, bleeding and cystic areas [13,16,17,18].

Clinical presentation of SPN is noncharacteristic: patients usually are asymptomatic, although they may complain of abdominal pain, discomfort or nausea. In cases of large-sized tumors, symptoms caused by compression of the adjacent organs may be presented, e.g feeling of fullness, dyspepsia or, less frequently, vomiting [2,3].

Physical examination is usually without abnormalities, although abdominal mass may be palpable in cases of large tumor sizes. Laboratory tests usually shows no abnormalities.

Diagnosis is mainly established incidentally, for example when performing diagnostic imaging for another reason – usually an ultrasound or CT scan.

There has been controversy regarding the optimal surgical procedure for SPN. Although a tissue-sparing

minimal resection such as enucleation would be enough to prevent postpancreatectomy diabetes mellitus, however, considering the malignant potential of the tumors, enucleation cannot ensure oncological safety [14]. Complete resection is usually curative and so radical surgery is the treatment of choice. No adjuvant therapy is recommended [17].

Prognosis is favourable, even in the presence of distant metastasis, and survival rates of more than 10 years have been described [19]. There are almost 100 cases of SPN of the pancreas reported in children among about 700 cases described [20].

The tumor we diagnosed had most of the characteristic features: the pancreas as the primary locaation and macroscopic features of a large, heterogeneous, well demarcated mass with calcification and fluid areas. The diagnosis was established on histopathological examination. A one year follow-up period showed no evidence of residual disease. Unfortunately, there are no conclusive radiological features that allow one to distinguish SPN from other pancreatic tumors. Biopsy and histopathology are necessary to make a definitive diagnosis [13,16,21].

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