SOLID PSEUDOPAPILLARY TUMOUR OF THE PANCREAS:
TWO CASE REPORTS AND LITERATURE REVIEW

SOLIDINIS PSEUDOPAPILINIS KASOS NAVIKAS:
DU KLINIKINIAI ATVEJAI IR LITERATŪROS APŻVALGA

Gintarė Narušaitė1, Kęstutis Trainavičius2

1 Vilniaus universiteto Medicinos fakultetas
2 Vilniaus universiteto ligoninės Santariskių klinikų Pediatrijos centras

SANTRAUKA
Solidinis pseudopapilinis kasos navikas – ypač reta vaikų liga. Šis navikas retai metastazuoja, todėl jo prognozė po radikalaus gydymo yra gera. Straipsnyje aprašomi du šio naviko klinikiniai atvejai ir pateikiama trumpa literatūros apžvalga.

ABSTRACT
Key words: solid pseudopapillary tumour, pancreas, pancreatic tumor of child, surgical treatment.
Pancreatic cancer is a very unusual neoplasm of childhood. Most pancreatic tumours do not secrete hormones and there are no signs or symptoms of the disease. Solid pseudopapillary tumour is a rare exocrine neoplasm of uncertain pathogenesis, which rarely metastasize and usually occurs in girls and young women, and extremely rarely in boys. Surgery is the treatment of choice. It depends on the pancreatic tumour localization. The prognosis and five-year survival rate is good. We report two solid pseudopapillary tumour of the pancreas cases, treatment solutions and a short literature review.

INTRODUCTION
Pancreatic cancer is a malignant neoplasm originating from atypical cells arising in tissues forming the pancreas. It is the fourth most common cause of cancer-related deaths in the United States and the twelfth worldwide. Talking about children, pancreatic tumours, like other very rare paediatric neoplasms (gastrointestinal carcinoma, melanoma, pleuropulmonary blastoma, and others), are considered as orphan diseases and represent a very rare entity. Pancreatic cancer can be classified into two main types – tumours of pancreas and periampullary tumours of pancreas. Classification based on the function divides pancreatic tumors to epithelial (exocrine, endocrine), mesenchymal and other group of tumours.

Solid pseudopapillary tumour (SPT) of the pancreas is an uncommon, but distinct pancreatic exocrine neoplasm with low malignant potential that makes up between 0,2 % and 3 % of all pancreatic neoplasms [1, 2]. This tumour was first described in 1959 by Virginia Franz and was known by many different names up until 1996 when the World Health Organisation introduced the term of solid pseudopapillary tumour [3]. In this article, we report two SPT of the pancreas cases during 2012 and our experience with the surgical treatment at Vilnius University Children’s Hospital and literature review.

1. Case
A 7-year-old boy presented to an outside hospital with a 2-month history of epigastric discomfort and abdominal pain. The transabdominal ultrasound examination revealed a 20 mm homogenic, round parenchymal mass in the body of pancreas. The patient was referred to our hospital for further treatment. He was reassessed and physical
examination revealed no further abnormalities. All laboratory parameters such as general blood, total and direct bilirubin, alpha amylase, alanine transaminase, asparagine aminotransferase, glucose were normal. Contrast enhanced computed tomography (CT) showed a tumour 19.7 mm in diameter at the pancreatic body. (Fig. 1) The main pancreatic duct was without dilatation. After three weeks, laparotomy partial resection of the pancreas and biopsy of tumour was performed with „Roux-en-Y” anastomosis of small intestine and pancreatic tail. The tumor was well-circumscribed, solid, sized 20 x 30 mm (Fig. 2,3). Biopsy results confirmed radical solid pseudopapillary tumour of the pancreas elimination.

Laboratory results after the surgery were normal, the same as postoperative course, except alpha amylase parameters. They were increased for five days after the operation and became normal during the sixth postoperative day. The patient was discharged on the fourteenth postoperative day. The patient showed no evidence of disease for a follow-up period of 2 years.

2. Case

A 15-year-old, previously healthy girl presented to our hospital complaining of a palpable left hypochondrial region of the abdominal wall mass. Transabdominal ultrasound examination showed heteroeoic mass sized 85 x 75 mm with poor vascular circulation in the left superior retroperitoneal space. Blood chemistry was within normal ranges. Abdominal computed tomography (CT) revealed a 80 x 65 x 88 mm well-circumscribed heterogeneous mass in the tail of the pancreas. Other organs were normal without lymphadenopathy.

During the second transabdominal ultrasound examination, biopsy of the mass was performed. Biopsy results were described as pseudopapillary tumour of the pancreas.

Laparotomy resection of pancreatic tail with the pseudopapillary tumour extirpation and splenectomy was performed. Despite the fact that transabdominal ultrasound and CT were performed before the surgery and did not show the tumour overgrowth of spleen vessels, after opening up the abdominal cavity, the overgrowth was found and the splenectomy was chosen. Operative time was 195 minutes.

Laboratory investigations were normal except for increased leucocytes and alpha amylase after the operation. The postoperative transabdominal ultrasound showed postoperative parapancreatic cyst. The „wait and see” treatment was chosen and the cyst dissapeared on its own. The increased leucocytes and alpha amylase concentrations also became normal. Other laboratory results were normal.

![Fig. 1. Computed tomography (CT) of the abdomen. Location of the tumor](image1)

![Fig. 2. Solid, well-circumscribed tumor of the pancreas](image2)

![Fig. 3. Solid, well-circumscribed tumor, with the size 20 x 30 mm](image3)
After discharge from the hospital, the patient showed an uneventful postoperative course and no evidence of disease for a 2-year follow-up period.

DISCUSSION

Solid pseudopapillary tumour (SPT) of the pancreas is more common in children than in adults and comprises about 30 % of pediatric pancreatic tumors. SPT occurs mostly in young females, who are less than 30 years old. About 90 % of female patients are with a mean age of 22 years. It is extremely rare in boys and only few male SPT patients have been reported so far. The female to male ratio is 9 to 1 [8]. There is a strong predilection for Asians, being less common among Europeans [5].

There are no typical clinical signs or symptoms of this tumor. SPT may present as an abdominal mass with discomfort, distention or pain, or it may be an incidental finding after performing transabdominal ultrasound. Some authors mention the cases when the main symptom was vomiting, diarrhea, weight loss, back pain and even pruritus [4–7]. Jaundice also may present as a symptom, if the tumor localisation is in the pancreatic head and the biliary tree is obstructed [8]. Laboratory abnormalities are rare and nonspecific. As in our cases, blood chemistry is usually within normal ranges.

SPT is usually diagnosed using transabdominal ultrasound (US), CT scan or magnetic resonance imaging (MRI). On US, SPT presents as well-circumscribed heterogeneous mass surrounded by a pseudocapsule of compressed pancreatic tissues [13]. On CT, tumor presents as solid, encapsulated masses with areas of necrosis, hemorrhage and cystic degeneration [12]. MRI has the advantage of avoiding radiation exposure and of a higher sensitivity for pancreatic masses. However, these imaging studies are not specific [12]. Two studies report the advantages of using endoscopic ultrasound-guided fine needle aspiration (EUS FNA) in diagnosing SPT [3, 12]. This diagnostic procedure should be chosen after using US, CT or MRI if the diagnosis is still in doubt. Otherwise, EUS FNA must be performed by experienced gastroenterologist and that is the main reason why this procedure is not available at all centers.

Solid pseudopapillary tumor of the pancreas is an epithelial tumor with solid and cystic macroscopic features [4]. The tumour can occur anywhere in the pancreas, but is slightly more frequent in the body and tail [3]. About 40 % of all SPT cases mass locations are the pancreatic head and 60 % – the body and tail of the pancreas [8]. Rarely, SPT can be found in extrapancreatic locations, such as adrenals, mesocolon, liver or greater omentum [9].

Surgery remains the keystone of treatment for pancreatic tumors in pediatric age as in adults. Complete surgical excision is curative for more than 95 % of patients with SPT limited to the pancreas. Pancreatic tissue should always be preserved as much as possible, as a result, types of surgical procedures depends on tumor location [7, 11]. If the tumor is located in the body or tail of the pancreas, an open distal pancreatectomy (with preservation of the spleen if possible) is an option of treatment. The other option is laparoscopic distal pancreatectomy, which was first performed in 1996. These two procedures were compared by M. van den Akker et al. As they presented, the laparoscopic distal pancreatectomy (LDP) had more advantages than the open distal pancreatectomy (ODP). The duration of LDP surgery was shorter than ODP, the median intraoperative blood loss was less, the median duration of hospitalization was shorter in the LDP group than in ODP group. However, they suggest that LDP is an appropriate choice for pancreatic tumors up to a maximal length of 10 cm [2, 4, 6, 14].

Some authors report cases, when enucleation of the tumor has been performed. They explain that tissue-sparing surgery is possible and justified, because SPT is mostly surrounded by a dense fibrous capsule and has favourable biological characteristics [1].

When the SPT is located in the head of the pancreas, pancreaticoduodenectomy is usually performed. It is also called Whipple operation, which was first described in the 1930’s by Allan Whipple. The most common technique of a pancreaticoduodenectomy consists of the en bloc removal of the distal segment (antrum) of the stomach, the first and second portions of the duodenum, the head of the pancreas, the common bile duct, and the gallbladder.

In some cases, pylorus-preserving pancreaticoduodenectomy, also known as Transverso-Longmire procedure, is also performed if the tumor is in the pancreatic head. The main advantage of this technique is that the pylorus, and thus normal gastric emptying, is preserved [6].

There are articles reporting about metastasing of SPT. Usually metastases occur in the liver, regional lymph nodes, but multiple metastases are rare. The morphology of them is mostly the same or similar to that of primary tumor. Surgical debulking of metastases is reported to be a good opportunity of reducing tumor’s spread, so the resection of metastatic lesions should be chosen whenever possible [1] The long-term prognosis is excellent. The overall 5-year survival after surgery is reported to be 95 % and the estimated 10-year survival rate is 93 % [3].

In conclusion, SPT is a rare differential diagnosis of a pancreatic mass in children. It might have no typical signs and may be incidentally found during transabdominal ultrasound examination. Surgery is the first treatment of choice and types of surgical procedures depends on tumour location. The prognosis of SPT is favourable after curative resection.
REFERENTES

54 medicinos teorijos ir praktikos 2015 - T. 21 (Nr. 1)