ACTA MEDICA SALINIANA

CASE REPORT

Frantz Tumor - Pseudopapillary Cystic Neoplasm of the Pancreas in Children: Report of Two Patients

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Case report. Pseudopapillary cystic neoplasm of the pancreas (Frantz tumor) is rare slow growing tumor with low malignant potential usually affecting young women. PCN have good prognosis in case of radical removal. Tumor was described for the first time by Frantz in 1959. A retrospective clinical analyses was made of two patients with PCN admitted in our hospital in the last 10 years. A 16 year old girl admitted in our hospital with intermittent abdominal pain lasting for 3 years. Abdominal ultrasound and CT-scan showed the presence of the tumor in a head of pancreas. Excision was performed. A 13 year boy came to routine US because frequent urological infection. Abdominal US and after CT are showed the presence of the tumor in a neck of pancreas. Excision was performed. In both of patients metastases were not noticed. The follow up period was 9 years in female patient and 2 years in male patient, Surgical resection of PCN affords an excellent prognosis without other ways of treatments.

Keywords. *Solid pseudopapillary tumor of the pancreas, Frantz tumor, children*

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Competing interests
The authors declare no competing interests.

INTRODUCTION

Solid pseudopapillary tumor of the pancreas (Frantz tumor) is rare slow growing tumor predominantly found in young women. This tumours are very important because even benign tumors have a malignant potential [1]. Only and correct treatment were done with total surgical resection [2]. Tumor was described for the first time by Frantz in 1959 [3]. Clinical presentation sometimes included a palpable abdominal mass with pain and vague abdominal discomfort. Sometimes the tumor found during routine examination of the abdomen without other symptoms [4].

We would like to present two children, female and male, with different clinical signs but with the same surgical treatment.

CASE REPORT

Patient 1

A 13 years old boy came to the hospital to do an ultrasound to frequent inflammation of the urinary tract. Examination of the abdomen found in the pancreatic neck well limited formation 4 cm in diameter. The border was well defined and the content displayed a markedly heterogeneous echo structure with scattered hypoechoic regions. Ductus pancreaticus was slightly expanded but routine laboratory examinations, amylase and lipase levels were within the limits of normal values. We have indicated surgery on suspicion of the PCN. At laparotomy we found a well-limited tumor in diameter 4 cm, which was attended from neck of the pancreas. We did the resection of pancreatic neck with tumor only. Head of the pancreas had drainage through ductus pancreaticus. The body and tail draining pancreatic enzymes in a separate bowel loop in a Roux-en-Y fashion [Figure 1, Figure 2] [5].

Patient 2

A 16 year old girl was admitted to our hospital because of intermittent abdominal pain lasting for 3 years. Physical examination revealed tenderness in epigastric region with palpable abdominal mass without any other suggestive physical findings. Routine laboratory examination (peripheral blood smear, urinalysis and serum electrolytes) was normal. Abdominal ultrasound showed the presence of a lesion in a head of pancreas 6, 5 cm in diameter. The border was well defined and the content displayed a markedly heterogeneous echo structure with scattered hypoechoic regions. CT showed round lesion 6 cm in diameter with low density structure and with high density ring located in the head of the pancreas [Figure 3.]. There was no evidence of metastatic disease to the liver or in the other part of abdomen. At laparotomy a well encapsulated tumor growing from the head of the pancreas was found. Excision was performed and all excision margins were free of the tumor without significant damage to normal pancreatic tissue.

Postoperative recoveries in both patients were uneventful with amylase and lipase levels normalizing within 7 days. The first patient was discharged on the 12 postoperative day and second patient on 14 post-

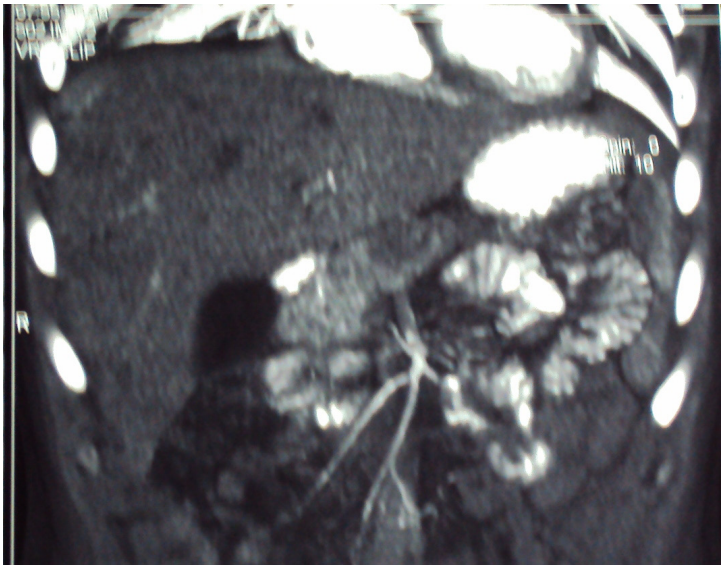

Figure 1. Frantz tumor- pancreatic neck with tumor

operative days. Histology revealed a tumoral lesions formed by cells arranged in pseudopapilary pattern with cystic spaces. The cells were cuboidal or cylindrical, small to medium, polygonal in shape with ovoid nuclei with very rare mitotic figures.

The follow-up period was in female patient 7 years and in male patient 1 year. Both of patients are alive in good condition with no recurrences, without local invasion or metastasis.

DISCUSSION

Solid pseudopapillary tumor of the pancreas or Frantz tumor are very uncommon entities. It is uncommon in children under 18 year old [6]. PCN occurs predominantly in young women in their second and third decade. The most frequent symptoms of this tumor are upper intermittent abdominal pain, abdominal tenderness, sometimes vomiting, abdominal discomfort and sometimes a palpable abdominal mass. Finding tumors in the small number of patients were found incidentally on abdominal sonographic examination for other reasons [7]. In most of patients complete blood test count, liver, pancreas and renal function test were normal. The PCN diagnosis always starts with abdominal ultrasound. The majorities of the tumors are located in the pancreatic body and tail [8].

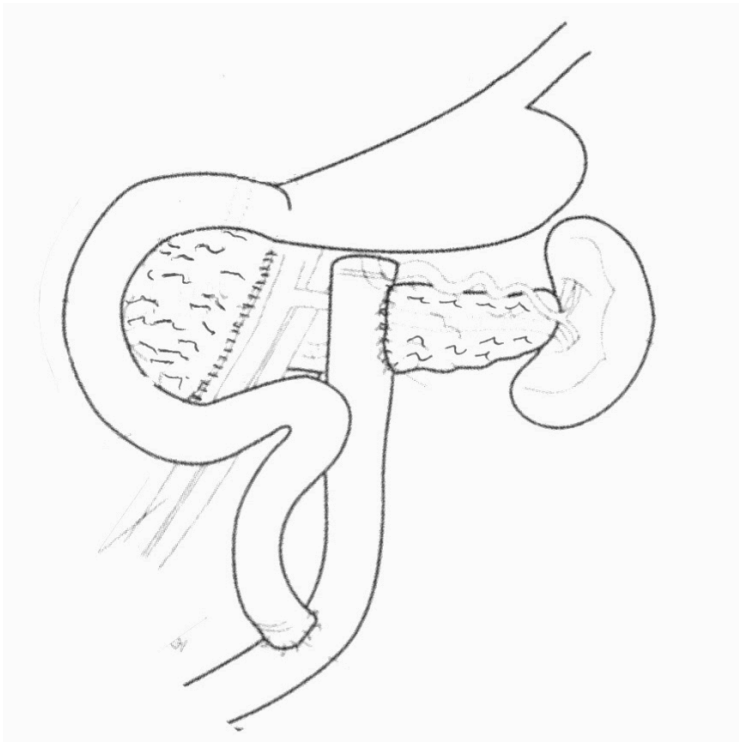

Figure 2. Operative details- The body and tail draining pancreatic enzymes in a separate bowel loop in a Roux-en-Y fashion

When you find a tumor of the pancreas with ultrasound it is necessary to make CT scan (computerized tomography) and MRI (magnetic resonance imaging) searching. Ultrasonography demonstrates solid mass, well demarcated and containing cystic areas of variable number and size. CT and MRI always revealed mass in the pancreas (head, neck or tail) with no evidence of invasion into surrounding tissues [9]. Regardless of the tumor is not very malignant it is necessary establish the existence of metastasis, invasion into surrounding tissues or existence of pathological lymph nodes. Once the diagnosis was established, the patient was submitted to laparotomy. Tumor must be completely removed and there are several ways of surgical treatment. Surgical treatment included enucleation and excision of the tumor, distal pancretectomy, distal pancreatectomy with splenectomy, pancreaticoduodenectomy, Whipple procedure, a pylorus preserving Whipple procedure. Surgical resection should be carried out with the required radicality, but pancreas preservation is advised in order to provide a relatively good quality of life. [10-13]. In the last period current surgical treatment of the PCN is laparoscopic treatment. In world medical literature only few reports have been made. All operation treatment in pancreatic surgery today should be made by laparoscopic treatment [14,15]. In recent years pancreas preserving strategy has been introduced for PCN. Pancreas preserving approach is feasible and safe. Different techniques for reconstruction have been adopted, one of them is jejunal anastomosis with distal pancreatic remnant. With this kind of preserving surgical procedure we also made spleen preservation. The potential risk of endocrine and exocrine insufficiency following removal of more than 50% of pancreas are thus prevented. [16-18]

In one our patient excision of the tumor was performed and in second patient resection of pancreatic neck with tumor was done. Head of the pancreas had drainage through ductus pancreaticus and tail pancreatic enzymes had drainage through a small bowel because we were made Y- Roux drainage.

PCN, in differential diagnosis, any cystic and solid pancreatic tumors should be considered. Radical resection if feasible should be considered the therapy of choice. There is no role of nonoperative management [13, 19]. The localization and presence of metastasis affect the surgical management. Characteristic ultrasonography, computed axial tomography and magnetic resonance imaging scans combined with sex and age should be sufficient for decision to operate. Complete surgical

removal of the PCN offers excellent prognosis. In cases of PCN chemotherapy is not necessary. The neoplasm usually behave like a very low-grade malignancy, so that complete removal is the treatment of choice for the tumor arising anywhere in the pancreas

On histopathological examination, the tumor was very cellular and well-circumscribed without involvement of adjacent pancreas. Pathologic examination of biopsy material revealed solid islets of small to medium sized cells with uniform spherical nuclei and tumoral cells forming papillary structures. Mitotic figures were few and no vascular invasion was seen. With these classical histological features, final diagnosis of PCN of pancreas was made.

Even in cases with local metastases, chemotherapy is not required. Solid pseudopapillary tumors show 15% metastatic rate involving liver and peritoneum. Even in the presence of disseminated disease, the clinical course is usually protracted and overall 5-year survival rate is 97%. The treatment of PCN is surgical. Overall, prognosis is good after limited resections because of its indolent growth [19, 20]

SUMMARY

PCN of the pancreas, which has a benign clinical course, is rarely observed as a low grade malignant tumor. Preoperative diagnosis is very important for the surgical strategy. Because the radiologic findings are not specific, radiologists and surgeons should take this tumor into consideration in the differential diagnosis of pancreatic malignancies, especially in young female patients. For real and accurate diagnosis of tumors we have to do ultrasound and CT diagnosis with surgical biopsy of the tumor. Formal surgical resection may be performed safely and is associated with long term survival. We recommend pancreas parenchyma preserving procedure without postoperative morbidity or mortality. In all cases of PCN long-term follow up is advisable [1, 13, 18-20].

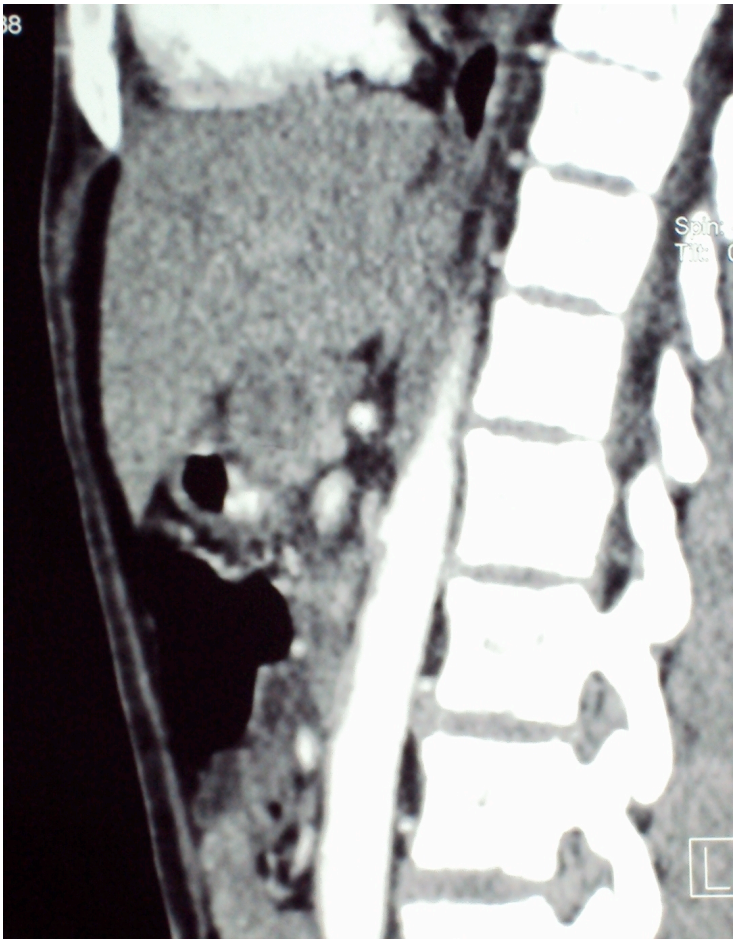

Figure 3. Frantz tumor- pancreatic head with tumor

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