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Case Report

# Complicated Course of a Solid Pseudopapillary Pancreatic Tumour in Child

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## Abstract

A clinical case of the likelihood of a complicated, life-threatening course of a solid pseudopapillary pancreatic tumor in child due to peptic erosion of tissue and blood vessels with capsule rupture is described.

***Keywords:*** *Solid-Pseudopapillary Tumour; Pancreas; Rupture; Bleeding; Operation*

## Introduction

Solid pseudopapillary tumour (SSPT) of the pancreas (PZH) is an extremely rare neoplasm with unclear histogenesis [1-3]. For the first time such a tumour was diagnosed in 1927 in a 19-year-old patient, but it was described as a special nasology only in 1959 by V. Frantz in a two-year-old child who underwent pancreatoduodenal resection, therefore it was called Franz's tumour [3]. However, in the future, various terms were used to denote this pathology: solid-cystic tumour, papillary-cystic tumour, solid pseudopapillary tumour. In 1996, experts from the World Health Organization (WHO) adopted the name "solid-pseudopapillary tumour", reflecting two main microscopic signs - the presence of areas of solid structure and pseudopapillary formations.

According to the WHO definition, a pseudopapillary solid tumour is a rare, usually benign neoplasm that develops mainly in young women; morphologically represented by monomorphic cells with different expression of epithelial, mesenchymal and endocrine markers, which form solid and pseudopapillary structures with frequent development of cystic-haemorrhagic changes. In the international histogenetic classification, the tumour is characterized by slow growth, relatively favourable prognosis, rare malignancy, and extremely rare metastasis [4].

The analysis of domestic and world literature indicates that the incidence of SPP in the pancreas is 1 - 2% of the total number of tumours of the exocrine part and about 5% of cystic neoplasms of the pancreas [1,5]. Most often, the SPS of the pancreas is localized in different parts of the pancreas: in the tail - 41.7%, the head - 33.3%, and the body - 25% [1,2,5]. Among paediatric patients, girls are more likely to suffer, with an average age of 12 years [6-8]. By 2008, about 130 publications appeared, both describing individual clinical observations and analysing their own data [5]. Ethnically, SPP PZH in people of the Caucasian race practically does not occur, the highest incidence of the tumour was noted in Asians (Japan) [9].

Unfortunately, there is no evidence on the aetiology, histogenesis, and risk factors for the development of RVS. There are suggestions that it develops from derivatives of ductal [10,11], acinar [2,11] or colony-forming germ cells and from derivatives of the neural plate. In addition, in the literature there are descriptions of the extra organic development of RVSF from ectopic tissue of the gland - in the omentum, mesentery of the large intestine, liver [11]. Thus, there is no unanimous opinion on the etiopathogenesis of the RVF SPM to date.

Patients usually complain of nausea, vomiting, discomfort, and abdominal pain. The majority of children should be noted a sharp deficit in body weight, lack of appetite. It is extremely rare that a tumour manifests itself as acute pancreatitis or as post-traumatic pancreatic cysts [1,2,5].

Thanks to the analysis of the literature, it became obvious that a fundamental role in diagnostics is given to magnetic resonance imaging and computed tomography. Pancreatic tumour with ultrasound tomography is visualized as a heterogeneous solid formation with the presence of cystic zones [2,11].

The main method of treatment for PFD is radical surgery [6,12,13]. After radical surgical removal of the tumour, recovery occurs in more than 95% of cases. The forecast is generally favourable. If the tumour ruptures, local spread and dissemination in the abdominal cavity is possible.

As can be seen from the presented brief review of the literature, solid-pseudopapillary pancreatic tumour in childhood is extremely rare, therefore the purpose of this message is to attract the attention of clinicians, it is worth remembering the likelihood of this pathology in children. This article presents the result of successful surgical treatment of a patient with a complicated course of solid-pseudopapillary pancreatic tumour, admitted to the "CDNMP" in Almaty.

## Case Report and Discussion

In 2018, a 14-year-old child I.A. (No. i/b 1-7367) came to the emergency room at the Central Center for Lateral and Medical Treatment in Almaty with complaints of severe abdominal pain, nausea and vomiting.

From the anamnesis of the disease: According to the mother for the last few months, the girl periodically complained of abdominal pain, sometimes weakness, poor appetite. A week before admission, there were recurrent abdominal pain, nausea and 1-fold vomiting. The child was examined in a private clinic, where outpatient treatment was prescribed. On September 18, 2018, at about 22 hours, the pain in the abdomen increased sharply, there was 2-fold vomiting with gastric contents, in connection with which the ambulance team delivered the child to the emergency room of emergency surgery "CDNMP" accompanied by the mother.

**Anamnesis of life:** According to the mother, the child from 2 pregnancies, 2 births, full-term. The child grows and develops according to his age. Hereditary history is not burdened. Not registered on "D".

The child's condition at admission was assessed as severe due to persistent abdominal pain syndrome. On examination, lethargy and protruding pallor of the skin were noted. Signs of intoxication were moderate. Tongue dry, white coated. Pulse 108 beats per minute, satisfactory filling and tension. Heart sounds are muffled. There is no wheezing in the lungs. The liver and spleen at the edge of the costal arch are not enlarged. The abdomen is swollen, symmetrical, participates in the act of breathing. On palpation, painful in all parts, more pronounced in the epigastric region and in the lower floor of the abdominal cavity. At the same time, in all parts of the anterior abdominal wall, there was a passive muscular defence, as well as a pronounced positive symptom of Shchetkin - Blumberg. With digital rectal examination, the ampulla of the rectum is free, no overhang of the fornix and no pain was found.

**In the general blood test during hospitalization:** Haemoglobin 106 g/l, erythrocytes - 3.8 x $10^{12}$/l, haematocrit - 31.8%, CP - 0.83, platelets 396 x $10^{9}$/l, leukocytes - 24.2 x $10^{9}$/l, stab - 1%, segmented - 89%, eosinophils - 3%, monocytes - 4%, lymphocytes - 5%, ESR - 45 mm/h. General urine analysis without pathology.

**Biochemical blood test:** Glucose - 9.8 mmol/l, bilirubin - 10.27 mmol/l, AST - 21.2, ALT - 22.9, urea - 4.3 mmol/l, albumin - 35.3, total protein - 58.8 g/l, amylase - 38, cholesterol - 2, 6, creatinine - 54.6.

The child was examined by an anaesthesiologist. All anaesthetic risks are considered. According to the conclusion of the anaesthesiologist, an increased transfusion readiness was required, and 2 central venous access was also established (FFP (fresh frozen plasma), blood preparations were prepared in advance).

Considering that on the basis of the above clinical and laboratory data in the child it was not possible to exclude acute surgical pathology of the abdominal organs, after a short preoperative preparation, it was decided to conduct diagnostic laparoscopy.

During laparoscopy, about 250 ml of dark blood with clots was found in all parts of the abdominal cavity, including the small pelvis. With further revision in area r. mesogastrium revealed a swelling formation of dark purple color with dimensions of 12.0 x 10.0 cm. Given the impossibility of a full revision of the formation and installation of the source of bleeding, it was decided to switch to conversion.

An upper-midline laparotomy on the left flank of the abdominal cavity and along the edges of the greater omentum revealed a small amount of blood (150 ml) with clots of different sizes. The spleen and liver are intact. Opening the omental bursa in the projection of the body and tail of the pancreas between the xiphoid process and the umbilical ring revealed a rounded formation measuring about 12.0 x 10.0 cm, with a smooth capsule. On the anterior surface of the formation, there is a violation of the integrity of the capsule with rough edges about 3.0 cm long. From the place of rupture, there is intense bleeding. During further revision, blood and organized blood clots of about 200.0 - 250.0 ml were found in the formation cavity. In addition, sequestered nodular necrotic tissues of 1.5 - 2 cm in size and soft consistency took place in the formation cavity. Blood clots and necrotic tissues were removed, while the tissues of the inner wall of the formation were diffusely bleeding, and a source of intense bleeding from an arterial vessel was also installed at the bottom of the cavity, most likely one of the branches of the arch of the pancreatic-duodenal artery feeding this formation (arcus arteriosus pancreatico-duodenalis). The bleeding was stopped by the methods of vascular ligation, coagulation and using a tachocomb.

Due to the fact that it was not possible to peel the wall of the formation, a biopsy material was taken from the tissue for histological examination. Then the wall of the cyst was sutured with interrupted sutures, leaving drainage tube No. 14 in the cavity. No other pathology in the abdominal cavity was revealed. Sanitation of the abdominal cavity with aseptic solutions to clean waters. Drain tube # 14 is installed through a separate incision into the left lateral canal. The wound was sutured tightly in layers.

Anaesthetic treatment proceeded smoothly, with blood products being transfused. The child's condition in drug sleep throughout the operation was assessed as stable, did not worsen.

**Macro drug:** Areas of the cyst wall measuring 1.5 x 2.0 cm and dark purple necrotic tissue with blood clots.

In the postoperative period, the child was in the intensive care unit for 3 days, where intensive therapy was carried out. After stabilizing the condition, the child was transferred to the surgery department, where he continued to receive antibacterial, detoxification and resorption therapy. With dynamic ultrasound in the postoperative period, a heterogeneous, decreasing volumetric formation with clear and even contours were visualized. In general, the postoperative period was uneventful. Drainage tubes were removed on the 4th and 8th days after surgery.

Results of histological examination: solid pseudopapillary tumour of the pancreas with cystic-haemorrhagic changes, ICD-O code 8452/1.

On the 12$^{th}$ day, the child was discharged home in a satisfactory condition with recovery. Further observation by a paediatric surgeon, gastroenterologist and oncologist was recommended.

## Conclusion

The absence of a similar clinical observation in the available literature indicates that there is a likelihood of a complicated, life-threatening course of a solid pseudopapillary pancreatic tumour. The causes of acute internal bleeding, acute blood loss and haemorrhagic shock can be peptic erosion of tissue and vessels of a pseudopapillary pancreatic tumour, followed by rupture of the cyst capsule. The lack of expression of the clinical picture of acute bleeding in our case can be explained by the maximum mobilization of all types of compensatory capabilities of the organism, the initial stage of bleeding from the "reservoir of pseudopapillary tumour", i.e. from outside the bed of the general blood flow, and also probably by an early admission and timely surgical treatment. With a complicated course of a solid pseudopapillary tumour of the pancreas, the main method of treatment is surgical.

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