Abstract

In this article we describe uncommon case of solid-pseudopapillary tumor of the pancreas. Multi-modal method has been used: patient underwent CEUS and contrast-enhanced CT. In pre-operative period segmentation with 3D-modelling were made. It was shown that contrast enhancement is mandatory while pancreatic lesion is assumed.

Keywords: Solid-Pseudopapillary Tumor; Pancreas; CEUS; CT; Pre-Operative Modeling

Introduction

Solid-pseudopapillary tumor (SPEN) is a rather rare pancreatic epithelial tumor with a low malignancy potential. The tumor was first described by pathologist Virginia Franz in 1959, sometimes referred to as the "Franz tumor". It makes up about 1% of pancreatic tumors and about 3% of cystic structure tumors [1]. SPEN progresses rather slowly and has a favorable prognosis, but metastases are detected in 15% of cases [2]. Morphologically tumor is presented by monomorphic cells with different expression of epithelial, mesenchymal and endocrine markers with the formation of solid and pseudopapillary structures with frequent development of cystic hemorrhagic changes [3].

Most often, SPEN is diagnosed in young women of non-European races. The ratio of women to men is 9.78: 1, and the average age of patients is 21.97 years. Patients are usually asymptomatic, sometimes they feel an increasing tumor in the abdomen, or pain of varying severity. Most often, the diagnosis is made by chance during a study of the abdominal organs for other reasons [4]. Tumor is localized more often in the tail of the pancreas - up to 41.7% [5]. The size of the tumor may vary, but usually it is quite large, on average 6.08 cm [6].

Tumors of small size usually do not have cystic changes. Larger tumors have a fibrous pseudocapsule; hemorrhagic and cystic necrotic changes are present to varying degrees [7]. In cells, a positive reaction to vimentin, neuron-specific enolase and α1-antitrypsin is noted. Immunohistochemical analysis often reveals specific expression of markers CD10, CD56 [8].

Case Report

In this publication, we present the medical history of a patient with verified SPEN, which confirms the complexity of the diagnosis of this nosology. Patient B., female, born in 1986 first appealed for medical help in 2013 with complaints of a constant throbbing pain in the abdomen with irradiation to the left half of the body. Considers herself ill for 2 years, notes weight loss of 5 kg over the past year.
Results and Discussion

According to the patient information, an abdominal ultrasound was performed with the conclusion “Pseudotumorous pancreatitis”. In 2013 CT scan of the abdominal organs with bolus contrast enhancement was performed. While describing the pancreas: “The pancreas is of the usual location. Contours are uneven, clear. The structure of the parenchyma is not homogeneous - in the head of the pancreas, more in the area of the hook-shaped process, neoplasm with even clear contours of 46.5 x 41 mm in size is revealed. The structure of the formation is predominantly liquid, with high-density inclusions up to 31 x 27 mm in size, moderately enhanced with the introduction of contrast. Main duct slightly expanded distal to the tumor. Peripancreatic area is not infiltrated. Conclusion: pseudocyst of the pancreatic head with high-density contents” (Figure 1).

![Figure 1: CT scan with contrast from 10.07.13. Arterial phase. Tumor of the pancreatic head is indicated by the letter T (tumor).](image1)

9 of September 13 biopsy of the tumor with 22G needle was performed under ultrasound control. Result: Structureless substance, abundant mixed flora, elements of cellular decay. A diagnosis was made: Pseudocyst of the head of the pancreas. The patient was referred under the supervision of a surgeon in ambulatory clinic.

In 2016, the patient performed CT scan of the abdominal organs without contrast enhancement, the data of the previous CT were not presented. A description was made: “in the region of the head of the pancreas, a round-shaped lesion is detected with a size of up to 61 mm across, with a density of up to 20 HU. The remaining parts of the gland are without features. There is an expansion of the main pancreatic duct to 4 mm. Conclusion: CT signs of pseudocysts of the pancreatic head” (Figure 2).

![Figure 2: CT scan without contrast from 07/22/16. The formation of the pancreatic head is indicated by the letter T (tumor).](image2)
20 of July 2018, patient underwent transabdominal ultrasound with contrast media (CEUS) - the introduction of 5 ml of sulfur lyophl- licate hexafluoride (Sonovyu, Bracco). An ultrasound revealed: in the area of the pancreatic head, a tumor of 78 x 72 x 69 mm is deter- mined, which displaces the vessels of the abdominal cavity. The tumor of a rounded shape, hypoechoic, has a heterogeneous structure with a hyperechoic rim of uneven thickness, with single flow signals according to color Doppler mapping, randomly located on the pe- riphery of the formation. When contrasting, a homogeneous contrast enhancement of the tumor in the arterial phase (at 16 seconds) is determined with an increased contrast enhancement of walls (Figure 3). Conclusion: Pancreatic neoplasm, possible SPEN. The assump- tion about SPEN was made according to characteristics of the images given in work of Jiang L., et al. [9]: the presence of a hyperechoic rim during gray-scale ultrasound examination and amplification in the form of a rim along the periphery of the tumor in the arterial phase.

Figure 3: Transabdominal sonogram of the pancreas. Right: on the native gray-scale image, tumor of a rounded shape, hypoechoic, with a heterogeneous structure with a hyperechoic rim of uneven thickness is visualized. On the left is a sonogram with contrast enhancement. With contrast enhancement, a homogeneous contrast enhancement of tumor in the arterial phase with an increased contrast enhancement of the walls of the tumor is determined.

27 of July 2018 the patient was performed CT with bolus contrast enhancement. In the description: tumor of a predominantly solid character is determined in the head region, which is hypovascular in comparison with the parenchyma, with an inhomogeneous accumu- lation of contrast medium in the arterial and venous phases. Its dimensions are 80 x 68 x 85 mm. No invasion of the surrounding vessels is noted, deformation of the superior mesenteric and portal veins is determined. The body and tail are atrophied, thickness up to 15 mm. Main pancreatic duct expanded to 6 mm. The splenic vein is expanded to 8 mm. Conclusion: CT picture of pancreatic head neoplasm, probably SPEN (Figure 4).

Figure 4: CT scan. Venous phase. The tumor was shown to cause compression of the portal and superior mesenteric veins. The splenic vein is moderately expanded to 8 mm (early signs of subhepatic portal hypertension appeared).
According to the large size and complexity of the topography, the data was segmented for preoperative 3D modeling. For this, data in DICOM format was loaded into the Avtoplan program developed at Samara State Medical University [10]. In the semi-automatic mode, the liver and pancreas were segmented, the formation was isolated using the “incremental segmentation” plug-in, and vascular structures were automatically tracked. The resulting polygonal model is shown in figure 5.

![Figure 5: Polygonal model of a tumor, blood vessels and pancreas with an expanded Wirsung duct, shown in different colors.](image)

The patient was referred for consultation at the Samara Regional Clinical Oncology Center. Repeated biopsy, cytological conclusion: probably a neuroendocrine tumor. An immunohistochemical study revealed: expression of epithelial markers and CD 56 is observed in tumor cells of pancreatic formation.

On October 2, 2018, the patient underwent surgery: pancreatoduodenal resection. An autopsy study determined: a section of the duodenum 12 with an adjacent tumor with a diameter of 10 cm in the capsule, a crumbling tissue of gray-brown color in the section. Microdescription: A solid-pseudopapillary pancreatic tumor encapsulated. Without invasion of surrounding tissue, necrosis. With low methodic activity.

Conclusion

Transabdominal ultrasound is a first-line imaging technique used to diagnose pancreatic diseases. Contrast ultrasound allows you to determine the perfusion of education in real time. According to the guidelines of the European Federation of Ultrasound Societies in Medicine and Biology (EFSUMB) on the use of contrast enhanced ultrasound (CEUS) for extrahepatic lesions, this method is useful in differentiating vascularized solid structures from avascular fluid and/or necrotic components of the lesion, in a more accurate determination of the sizes and boundaries of the tumor, including the ratio with adjacent vessels, and also helps in choosing the next imaging method [11]. In the meta-analysis of D’Onofrio M., which included 23 studies, the sensitivity and specificity of contrast ultrasound for differentiating neoplastic from neoplastic formations was 95% and 72% [11].

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Unfortunately, in the described case, it was the differential diagnosis of tumor and non-tumor pathology of the pancreas that took almost five years. This clinical case shows several signs uncharacteristic of SPO: the location in the head, not in the tail, the presence of pain and visualization features: with CT, even with a size of about 8 cm, there were no necrotic and cystic changes. Despite the benign nature of the tumor, pronounced negative dynamics is shown: over 5 years, the diameter has increased almost 2 times, signs of compression of the portal vein system have appeared. The complexity of diagnosing SPEN was shown: in the described case, even a biopsy with cytological examination performed in 2013 did not answer the question about the tumor nature of the process. Also, the article is one of the first publications, where the images and semiotics of SPEN with contrast-enhanced ultrasound (CEUS) are presented.

Conflict of Interest

The authors of this article declare no conflicts of interest.

Bibliography