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Research Article

Differential Radiology Diagnostics of Rare Cystic Pancreatic Lesions

Yu. A. Stepanova*, A.G. Krieger and D.V. Kalinin

A.V. Vishnevsky, National Medical Research Center of Surgery of the Ministry of Health of Russia, Russia

*Address for Correspondence: Stepanova Yulia Aleksandrovna, Academic Secretary of A.V. Vishnevsky National Medical Research Center of Surgery, Bolshaya Serpukhovskaya st., 27, Moscow, 117997, Russia, Tel: +7 (916) 654-84-85; E-mail: stepanovaua@mail.ru

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Abstract

The purpose: to analyze the criteria and assess the possibilities of radiology methods in the diagnosis of rare cystic pancreatic neoplasms.

Materials and methods: From 2004 to 2019 at A.V. Vishnevsky NMRC of Surgery 222 patients with suspected cystic pancreatic lesions were examined and treated, 15-80 years old, men - 59.9%. Radiology examination: ultrasound, multislice computed tomography (MSCT) and magnetic resonance imaging (MRI). 182 patients (82.0%) were operated. Morphology: intraductal papillary mucinous neoplasms (IPMN) - 96 (43.2%), solid pseudopapillary tumors (SPPT) - 50 (22.6%), cystic form of duodenal dystrophy (DD) - 72 (32.4%), cystic teratoma - 1 (0.45%), cystic lymphangioma – 1 (0.45%), echinococcosis – 2 cases (0.9%).

Results: The article provides the differential diagnostic criterias for rare cystic pancreatic lesions. Complete radiology examination (ultrasound, MSCT, and MRI) makes it possible to diagnose and differentiate various forms of pancreatic cystic lesions. At suspected presence of a rare pancreatic lesion, the radiology preference should be given to MRI. Criteria for radiology diagnostics of the IPMN and DD cystic form have been well defined. The formulation of the SPPT diagnostic criteria requires the accumulation of a greater number of observations in one place and an analysis of the corresponding information. Anamnesis of the disease is an important supplement to the SPPT radiology diagnostics.

Conclusions: A.V. Vishnevsky NMRC of Surgery (Moscow) has fifteen years experience in diagnosing and treating such rare forms of pancreatic cystic neoplasms as IPMN, SPPT, and the cystic form of DD. It should be pointed out that, due to the accumulation of knowledge and the expert analysis, preoperative diagnostic accuracy has been growing.

Keywords: Cystic pancreatic neoplasm, US, MSCT, MRI, Radiology, IPMN, SPPT, Diagnostics

Introduction

Due to development and promotion of new methods for abdominal diagnostic imaging, different specialists meet cystic lesions of the pancreas more often. By late 1970s, the range of known cystic pancreatic neoplasms was represented mainly by serous and mucinous tumors. In the 80s and 90s, due to the rapid development of diagnostic and surgical techniques, it has considerably grown. In the past 20 years, it has been reported on previously undifferentiated diseases of the pancreas, i.e. intraductal papillary mucinous neoplasms (IPMN), solid pseudopapillary tumors (SPPT), cystic form of duodenal dystrophy (DD) and others. The classification of cystic tumors of the pancreas published by The European Study Group on Cystic Tumors of the Pancreas in 2018 is currently used [1]. The differential diagnostic criteria for these pathologies are not always obvious. The accuracy of preoperative diagnosis is undoubtedly important as it defines the tactics of patients' treatment. Thus, differential diagnostics of these lesions and determination of the optimal treatment tactics are still questionable.

The purpose of the study is to analyze the criteria and assess the possibilities of radiology methods in the diagnosis of rare cystic pancreatic neoplasms.

Materials and Methods

From 2004 to 2019, at A.V. Vishnevsky National Medical Research Center of Surgery (A.V. Vishnevsky NMRC of Surgery), 222 patients with suspected cystic lesions in the pancreas were examined and treated, aged from 15 to 80, men slightly predominated (59.9%). Patients were subjected to the complete radiology examination: ultrasound examination (in B-mode and duplex scanning, with 3D reconstruction, if it's necessary), multi slice computed tomography (MSCT) and magnetic resonance imaging (MRI) with contrast enhancement. 182 patients (82.0%) were operated. Morphological diagnosis was defined as follows: IPMN - 96 cases (43.2%), SPPT - 50 cases (22.6%), cystic DD form - 72 cases (32.4%), cystic teratoma - 1 case (0.45%), cystic lymphangioma – 1 case (0.45%), and echinococcosis – 2 cases (0.9%).

Results

Speaking of the results of the survey of patients with IPMN, SPPT, and cystic DD form, it should be noted that, while analyzing data of radiology examination, a diagnostician should also consider the anamnesis of the disease and the results of clinical examination of a patient. This principle coincides with the doctors of the French Medical Association conclusions that were made during their multicenter study of pancreatic cystic neoplasms based on one of the biggest clinical materials (study included nearly 400 patients). Joel Le Borgne et al. believe that it is the combination of radiology diagnostics data, medical history, and complaints that allow defining the correct diagnosis [2].

Intraductal Papillary Mucinous Neoplasm

The IPMN is a rare exocrine pancreatic tumor originating from the main pancreatic duct (MD) or one of its branches and characterized by the secretion of large amounts of extracellular mucus, the papillary type of growth, accumulation of mucus in the pancreatic ducts, a high degree of resectability and good prognosis. Depending on the localization of tumors, three types are defined: main duct type (MD-IPMN), branch duct type (BD-IPMN) and combined type (combined-IPMN) [3].

The first report on IPMN was made more than 70 years ago, but the characteristic features of these neoplasms were not taken into account until K. Ohhashi et al. [4] reported on a series of cases of mucin-producing pancreatic tumors in 1982. They diagnosed a malignant tumor of the pancreas, which was accompanied by a significant expansion of the main pancreatic duct, the accumulation of a large amount of mucin in its lumen and a gaping of the mouth of the large duodenal papilla - the Ohashi triad. This condition has been termed "mucin-producing pancreatic cancer." It was noticed that a similar variant of a tumor lesion of the pancreas is accompanied by high resectability and a rather favorable prognosis for the patient's life. Later it became known that the described changes can occur not only in malignant tumors, but also in borderline, as well as in benign neoplasms of the pancreas [5].At A.V. Vishnevsky NMRC of Surgery, 96 with morphologically verified IPMN patients were examined and treated, aged from 38 to 80, men slightly predominated (57.3%). The following types of IPMN have been diagnosed: MD-IPMN - 19 cases (19.8%), BD-IPMN -

46 cases (47.9%), combined-IPMN – 31 cases (32.3%).

At US IPMN is featured by an extended MD with possible segmental duct extension, walls thickening, parietal papillary excrescencies with varying degrees of severity, possible tumor mass lesion around the MD, as well as an absence of blood flow in the structure of the tumor masses at duplex scanning.

At MSCT, IPMN appears as an extended MD with thickened walls, parietal soft tissue growth with density of up to 43 HU, soft tissue lesion around the MD, accumulating contrast agent through all phases of the examination.

MRI shows the MD extension with wall thickening, as well as thickened segmental ducts of the first order (especially at the magnetic resonance cholangiopancreatography (MRCP), parietal soft tissue growth, soft tissue lesion around the MD, accumulating contrast agent through all phases of the examination.

MD-IPMN. All radiology methods (US, MSCT, and MRI) allow defining diagnosis independently (Figure 1).

BD-IPMN. At US and MSCT, the branch ducts are poorly available to visualization due to their insignificant diameter in unaffected parts, that is why the connection of individual small cystic cavities in the pancreatic parenchyma with the main duct is not always possible to determine. MRCP becomes the method of choice (Figure 2), making it possible to clearly differentiate cystic cavities associated with the main pancreatic duct.

Combined-IPMN (Figure 3). Our experience in diagnostics, treatment and morphological studies of this type of neoplasms suggests that, most likely, this type of IPMN is a progression of either MD-IPMN or BD-IPMN. Despite the fact that all methods of radiology diagnostics allowed to get the diagnostic in lesion at the combined-IPMN, as is the case of MD-IPMN and BD-IPMN, the greatest diagnostic capacity belongs to MRCP, as it allows tracking of both MD and BD-IPMN in three dimensions throughout its extension.

Thus, the criteria of radiology diagnostics of the IPMN are as follows: extended duct throughout its range; thickening of its walls; parietal papillary proliferations of varying degrees of severity; possible single expanded segmental ducts and tumor masses around the main pancreatic duct. The MRCP is the priority method of

Figure 1: MD-IPMN: a — US image (DP — main pancreatic duct, arrows indicated papillary proliferations and extended parietal branch duct of the second order); b — MSCT image; c — MR image

diagnostics.

Tumor malignancy criteria [6]: obstructive jaundice; parietal papillary growths of more than 5 mm / presence of a solid component; the presence of tumor cells during cytological examination; the diameter of the pancreatic duct is more than 10 mm.

In accordance with these criteria, 56 (58.3%) patients were operated on, 40 (41.7%) were under observation:

Type 1: operated on - 17 (89.5%); observation - 2 (10.5%);

Type 2: operated on - 12 (26.1%); observation - 34 (73.9%);

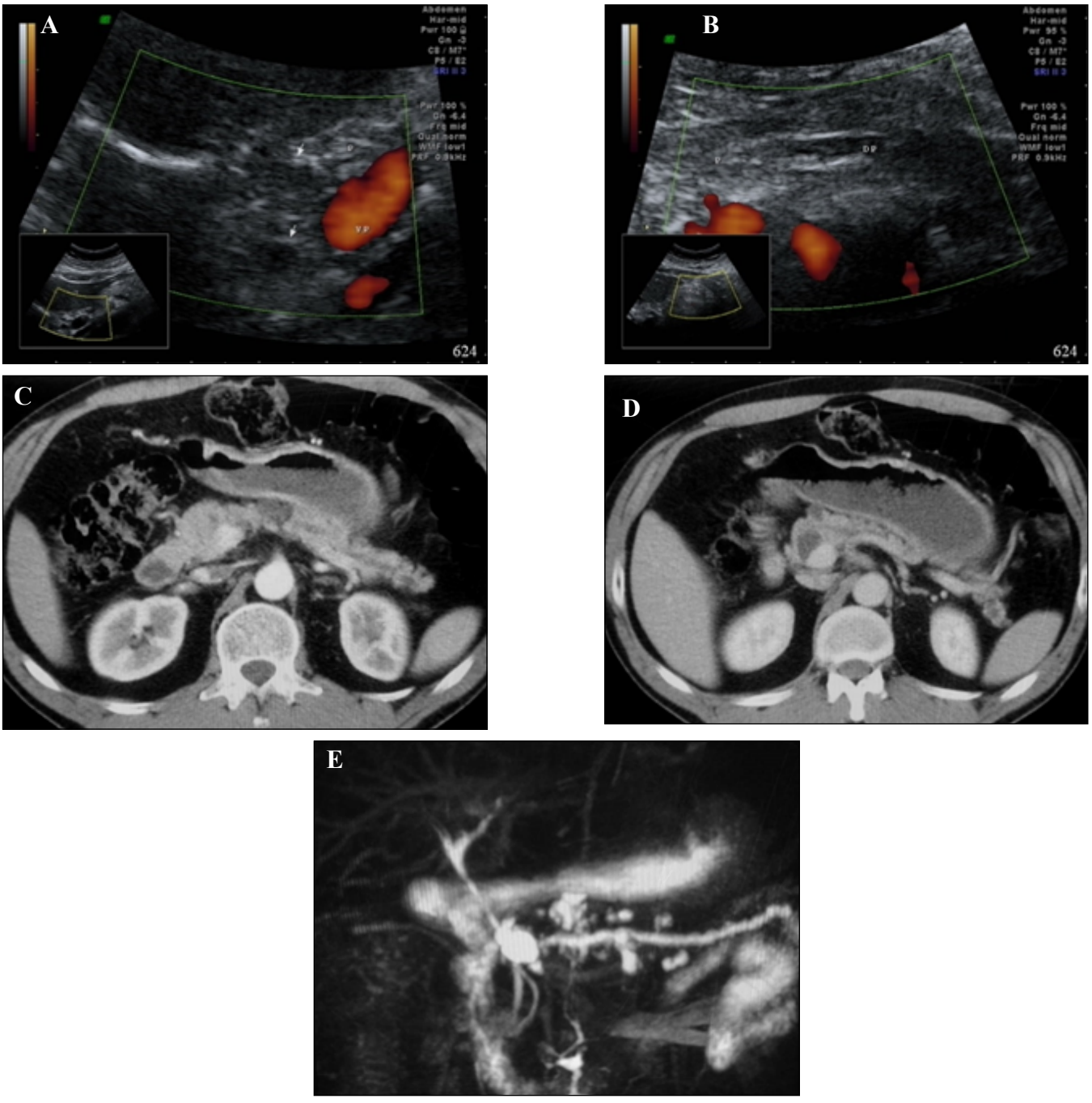

Figure 2: BD-IPMN: a, b — US images (VP – vena portae, P – pancreas, DP — main pancreatic duct, arrows indicated separate small cystic cavities of branch ducts); c,d — MSCT image; e - MRCP image

Type 3: operated on - 27 (87.1%); observation - 4 (12.9%).

Thus, dynamic monitoring is carried out mainly for type II tumors.

The observation is carried out according to the following scheme: after 3 months, after 3 months, then 1 time in 6 months. The scope of the examination during the follow-up: MSCT and / or MRI and the tumor marker CA 19-9.

Criteria for malignancy in dynamic observation: newly detected expansion of the pancreatic duct 5-10 mm; an increase in the cystic component over 5 mm in 1 year; newly identified parietal papillary growths up to 5 mm; the appearance of a solid component; transition to type III; an increase in CA 19-9 [6].

The results of the histological examination of the removal lesion showed that MD-IPMN are mostly invasive, BD-IPMN are mostly non-invasive and combined-IPMN had an undetermined malignancy potential in ¾ of the cases (Figure 4A and B) and carcinoma in ¼. (Figure 4C and D).

Solid-pseudopapillary tumors

SPPT are considered rare and "mysterious" neoplasms of the pancreas. For the first time this tumor was noted in 1927 in a 19-year-old girl, but it was described as a special form only in 1959 by V.K. Frantz [7]. Later, as such neoplasms were identified and described, many terms were proposed to designate these tumors, however, in 1996, WHO experts adopted the term "solid-pseudopapillary tumor", reflecting two main microscopic signs: the presence of areas of solid structure and pseudopapillary lesions [8]. According to the WHO definition, the SPPT is a rare, usually benign, tumor primarily affecting young women, expressed by monomorphic cells with different expression of epithelial, mesenchymal and endocrine markers and forming solid and pseudopapillary structures with frequent development of cystic-hemorrhagic changes [9]. In the international histogenetic classification of tumors, there are two forms: the SPPT in the group of neoplasms and SPP-carcinoma in the group of malignancies. SPPT account for 0.2-2.7% of the total number of pancreatic cancers [10-12]. The female to male ratio is 8-9 to 1 [13].

C.-C. Yu et al. [14] define three types of SPPT MR imaging:

Type 1 corresponds to completely solid neoplasms;

Type 2 is characterized by a combination of solid components with hemorrhages;

Type 3 is featured by extensive hemorrhages and cystic lesions.

This classification is valid for SPPT as a morphological form in

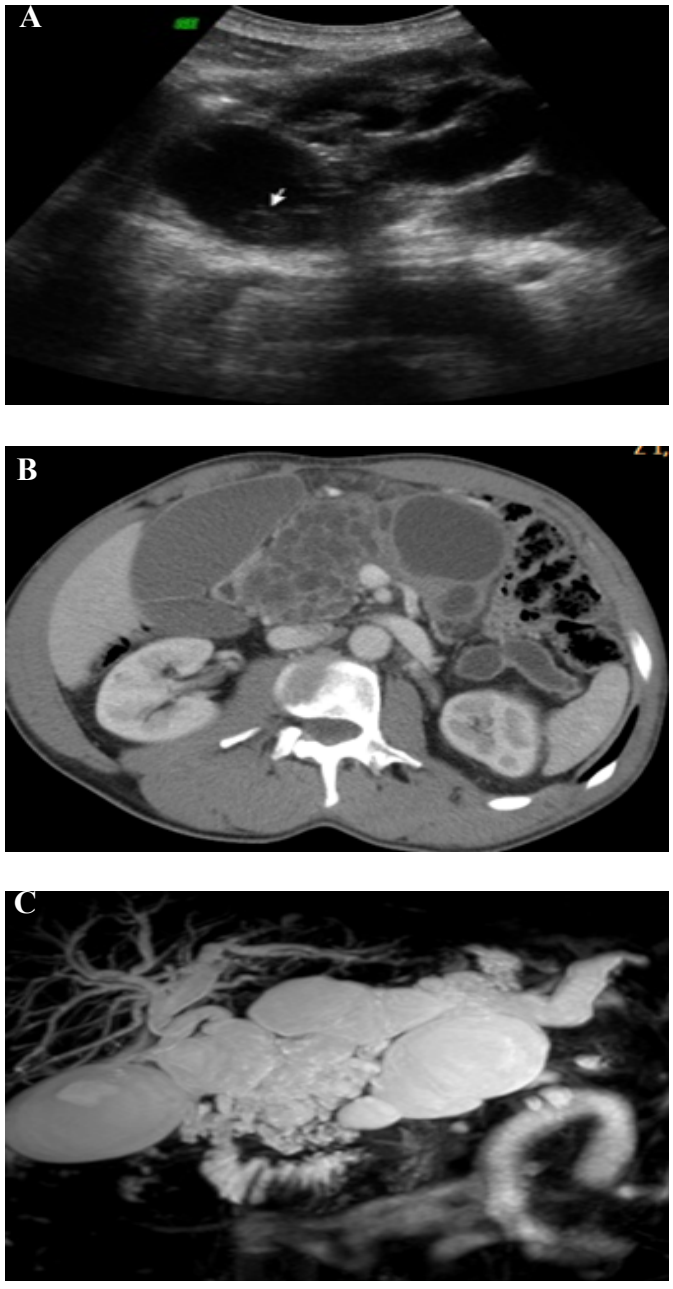

Figure 3: Combined-IPMN: a — US image (arrow indicated papillary proliferations); b — MSCT image; c — MRCP image

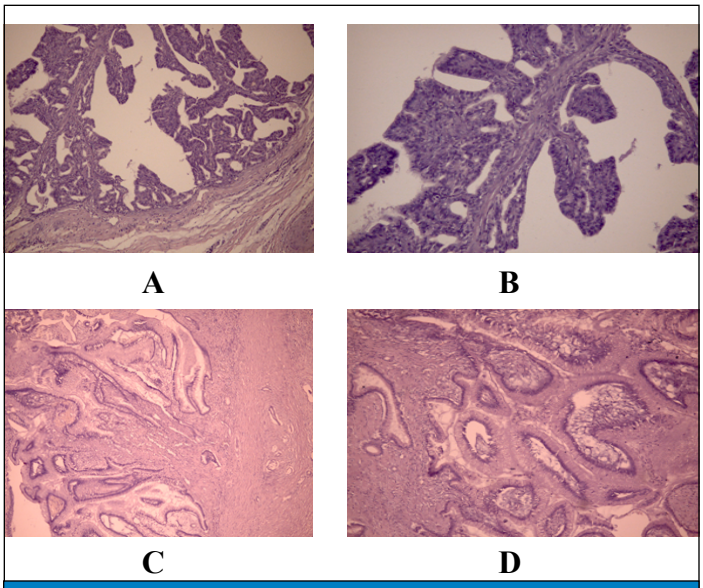

Figure 4: Histology, IPMN with moderate epithelial dysplasia, a - x100 magnification and b - x200 magnification; IPMN-carcinoma, c – x50 magnification and d - x100 magnification. Hematoxylin and Eosin.

general, in our opinion.

Despite the locally aggressive features, the tumor has a low-grade malignant potential and tends to have a favorable prognosis, even in the presence of metastatic disease [15-17]. Common sites of metastasis are liver, peritoneum, omentum and regional lymph nodes. The correlation between tumor size and malignant potential is controversial [16,18] and the blood vessels' invasion, high degree of nuclear atypia, high mitotic count and presence of large necrotic clusters are considered criteria of high malignancy potential. K. Nishihara et al. [19] demonstrated aneuploidy in patients with metastases, whereas patients without any sign of malignancy showed diploidy.

The reported mortality from this tumor is less than 2% [20]. Compared to female patients, males have a twofold higher incidence of metastases and a threefold higher incidence of death [12].

At A.V. Vishnevsky NMRC of Surgery, 50 patients with the mor-

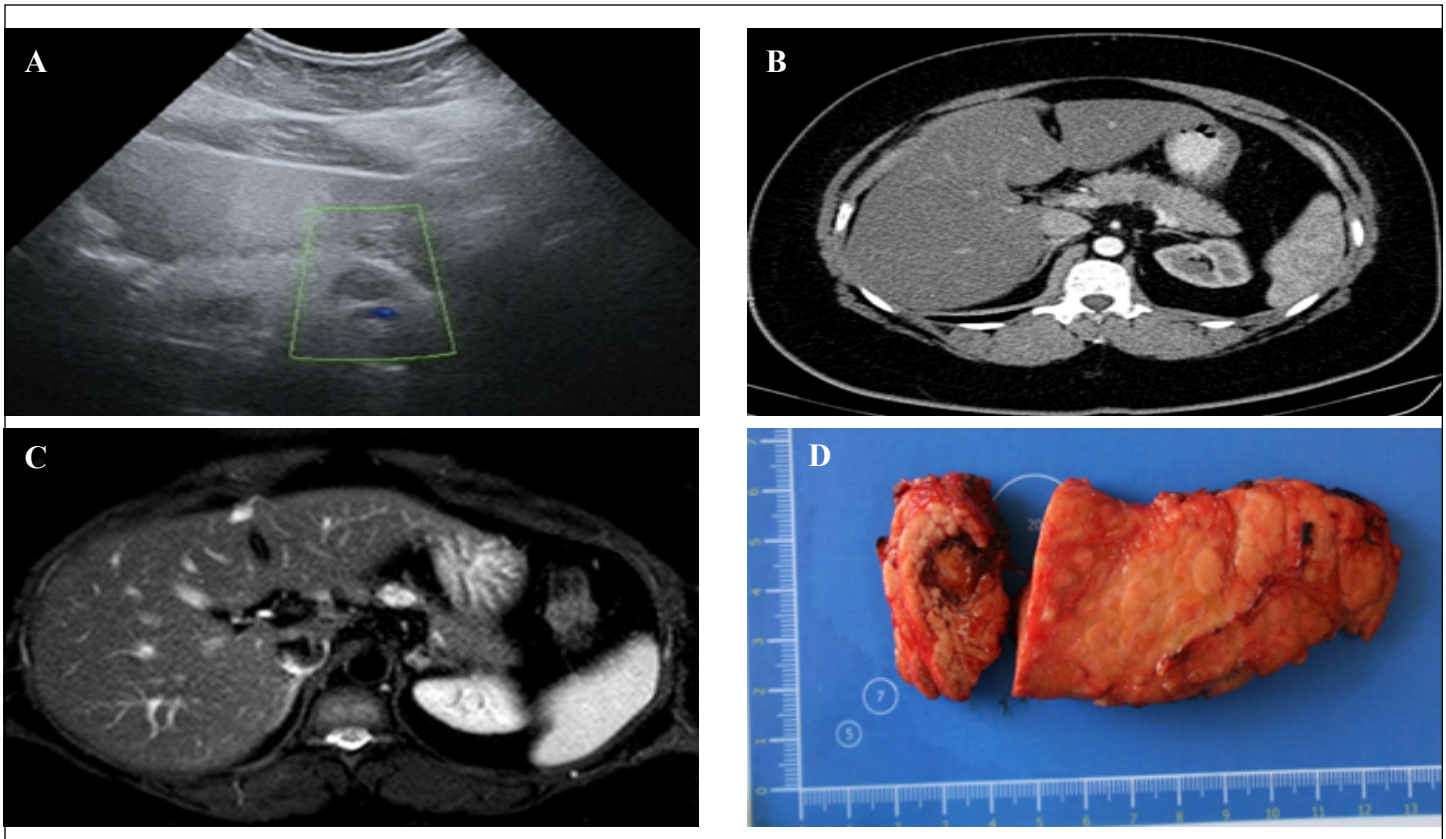

Figure 5: SPPT type 1: a — US image, duplex scanning; b — MSCT image; c — MR image; d - macroscopic appearance of the pancreas with SPPT

phologically verified SPPT were treated. Women dominated with 96.0%. All patients aged between 15 and 68. Distribution of neoplasms according to the classification of C.-C. Yu et al. [14] was as follows: Type 1 - 23 cases (46.0%); type 2 - 19 cases (38.0%), type 3 - 8 cases (16.0%). It should be noted that the presence of a cystic component does not always depend on the size of the tumor.

The greatest difficulties were caused by 1 type SPPT lesions diagnosis.

US is almost impossible to differentiate individual microcavities of a solid lesion. SPPT, as a rule, are well vascularized, however, with lesion sizes up to 3.5 cm in diameter, no data were obtained for the presence of blood flow during duplex scanning, with sizes over 3.5 cm in diameter single arteries and veins were recorded in the lesion structure.

It should be noted that at MSCT, high indicators of HU are possible in the fluid component of the 1 type lesions, which is due to the fact that the cystic cavities are very small and on the CT slices a "clipped" solid component can fall into the measurement point.

MRI makes it possible to identify small fluid lacunae of the lesion and differentiate their hemorrhagic content, which is defined as a hyperintense MR signal in the T1-FFE, which is explained by the significant paramagnetic effect of methemoglobin.

SPPT type 1. The most complex for differential diagnostics; as a result, the highest number of diagnostic errors at US and MSCT were noted (Figure 5).

SPPT type 2. All radiology methods (US, MSCT, and MRI) allow

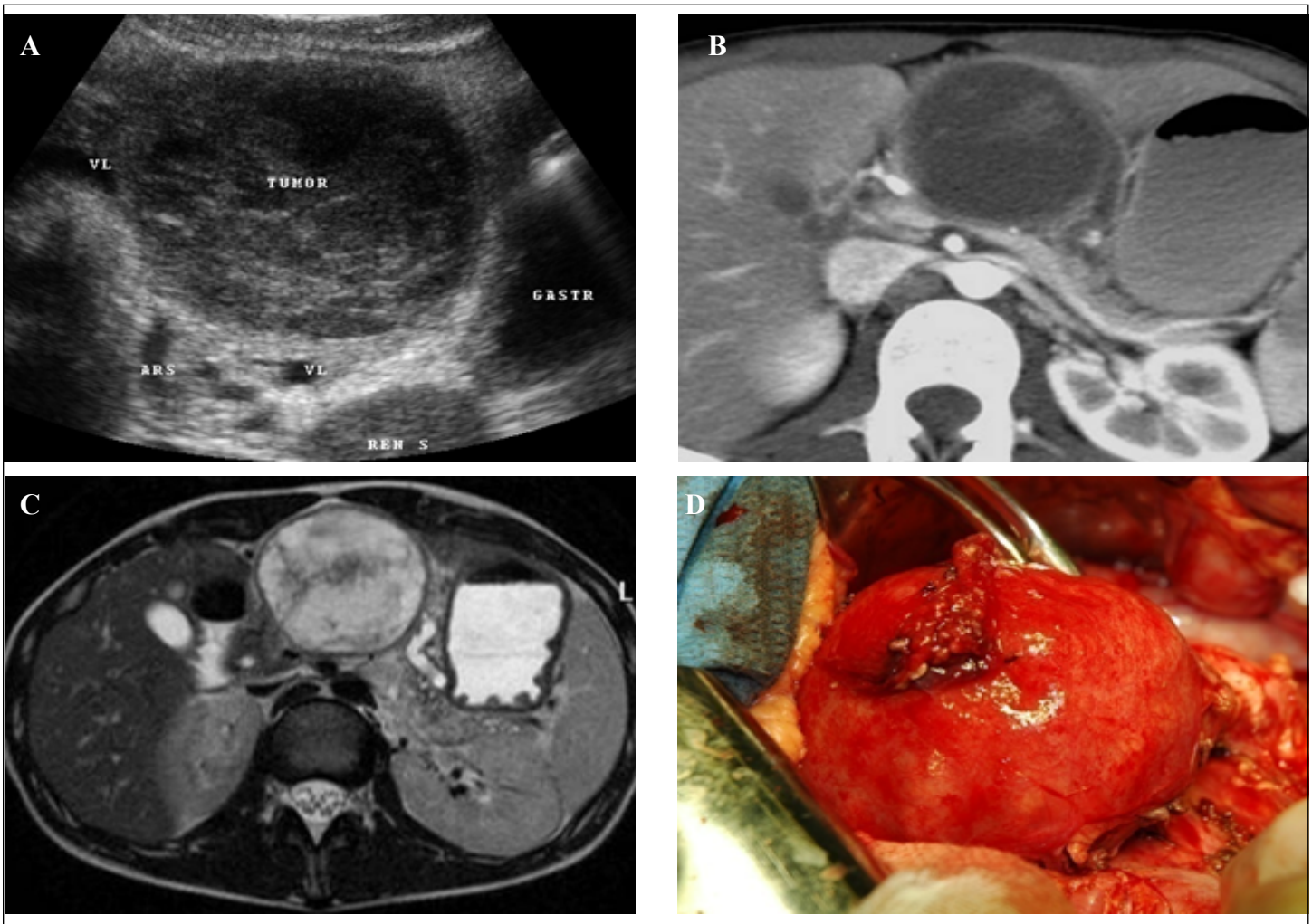

Figure 6: SPPT type 2: a — US image (TUMOR — tumor, VL — spleen vein, GASTR — stomach, ARS — left kidney artery, REN S — left kidney); b — MSCT image; c — MR-image; d - macroscopic appearance

defining the diagnosis independently (Figure 6). At US, it is useful to practice the 3D reconstruction of ultrasonic images to verify the cystic structure of neoplasms more accurately.

SPPT type 3. All radiology methods (US, MSCT, and MRI) also allow defining the diagnosis independently (Figure 7). As a rule, those are large enough tumors, with the presence of calcinates in a capsule and partitions. 3D reconstruction of US images allows tracking of neoplasm's cystic structure more accurately, "removing" acoustic interference from calcinates.

Preoperative verification - 56.0%. In 16 cases (32.0%), the diagnosis was not made preoperatively: neuroendocrine tumor was diagnosed in 5 cases; cystic tumor - 5; retroperitoneal tumor - 3, pancreatic adenocarcinoma - 2; pseudocyst with hemorrhage - 1. Differential diagnosis was made in 6 cases (12.0%), which included, in addition to SPPT, neuroendocrine, cystic tumor and adenocarcinoma. It should be noted that the largest number of misdiagnoses occurred in the first 15 cases. As experience gained in the diagnosis and treatment of this type of tumor, preoperative verification has improved significantly.

Considering the above-mentioned clinical-laboratory and instrumental features of the SPPT, it seems possible to recommend the following criteria for the SPPT identification proposed by T. Asano et al. [21]:

- the overwhelming affliction of young women and girls;
- the abdominal lesion, with or without abdominal pain;
- the absence of deviations in laboratory and hematology indices;
- the combination of solid and cystic areas, including hemorrhages, at US, MSCT, and MRI.

Analysis of the results of our studies has shown that the most effective method of the SPPT diagnostics is MRI, which, due to the paramagnetic effect of methemoglobin, helps to verify neoplasms without the radiation exposure and the introduction of a contrast agent. The US and MSCT allow a clear verification (including anamnesis) only of SPPT types 2 and 3.

On microscopic examination of removed tumors, they consisted of small monomorphic cells forming solid (Figure 8A) and papillary structures (Figure 8B).

Cystic form of duodenal dystrophy

Ectopia, or heterotopia, of the pancreas is an unusual localization of pancreatic tissue with its own blood supply and duct system without a vascular, nervous, or anatomical contact with the typically located (orthotropic) pancreas [23,23]. The ectopic pancreas often resides in abdominal and thoracic organs, most often in the stomach (25-60%), and less frequently in the duodenum (DD) (25-35%) [24]. Rare complications of pancreatic ectopia are cystic dystrophy and malignant transformation [25]. Duodenal dystrophy is a chronic inflammation of the tissues of the pancreas ectopic in the duodenum wall. Due to difficulties in making a diagnosis and vague signs, it was first described only in 1970 by the french authors F. Potet and N. Duclert [26]. In the English literature, the term "groove pancreatitis" is often used [25,27]. DD is a rather rare disease, so according to the multicenter study

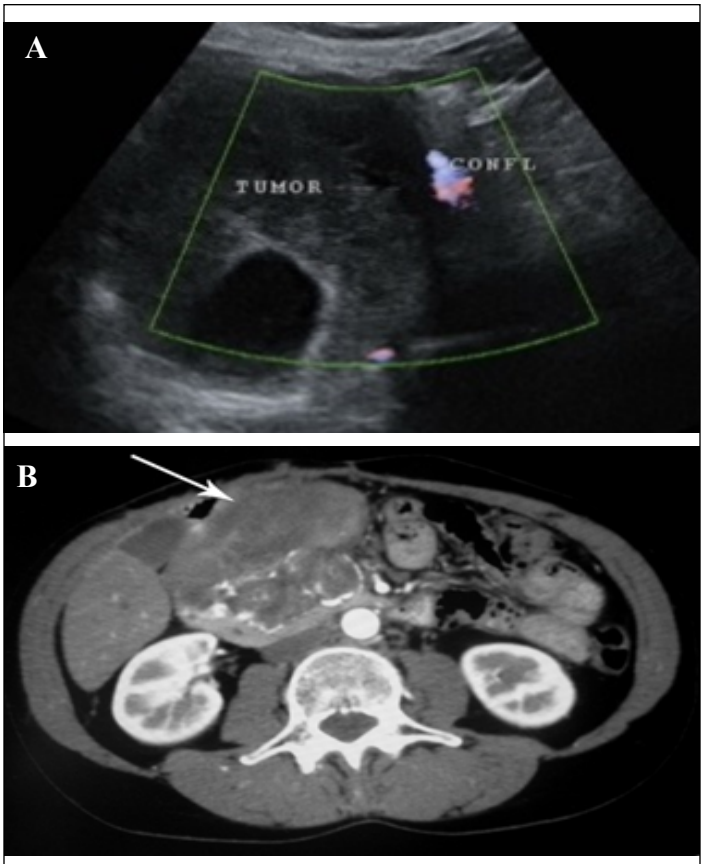

Figure 7: SPPT type 1: a — US image (TUMOR - SPPT, CONFL – confluens of vena portae); b — MSCT image (tumor is indicated by an arrow)

PanCroInfAISP, conducted in Italy among 893 patients with chronic pancreatitis included in the study, DD was detected in 6% of cases [28].

At the A.V. Vishnevsky NMRC of Surgery, 72 patients (93.1% men) with the cystic form of DD were examined and surgically treated.

The examination of the patients with typical DD by radiology methods (US, CT, and MRI) gave the following data. In all cases, the DD wall was irregularly densified and thickened in the range of 16.4 to 67.0 mm due to the cystic lesion with a varying degree of cystic component expression deforming the bowel lumen (Figure 9-11). Intramuscular injection of 1 ml of atropine before examination allows more clearly differentiating changes in the duodenum (Figure 10). Capsule and partitions were avascular. The main pancreatic duct hypertension was diagnosed in 39 cases (54.1%), one of which had the main pancreatic duct cystic expansion. In 19 cases (26.4%), the chronic calculous pancreatitis was identified, with the presence of calcinates both in the gland parenchyma and main duct lumen (Figure 12). The development of the above changes in the pancreas in some cases has led to such complications as biliary and portal hypertension, extravasal compression of veins in the portal system and inferior vena cava system, and the thrombosis (partial or occlusive) of the portal vein system.

In all 72 cases, the gastroduodenal artery forward and leftward displacement was diagnosed radiologically. The localization of this artery is a kind of a beacon in the DD diagnostics. If the gastroduodenal artery is visualized to the right of the cystic lesion, then it is most likely localized in the DD wall.

It should be noted that MRI showed the cystic changes in the DD wall more clearly, even without the contrast agent. In our view,

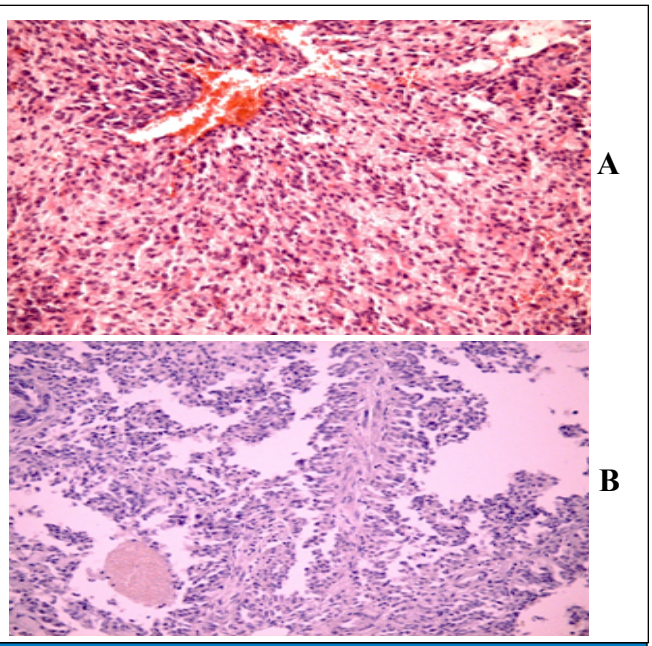

Figure 8: Histology of SPPT of the pancreatic head, Hematoxylin and Eosin, x200 magnification: a - solid structure; b - pseudopapillae.

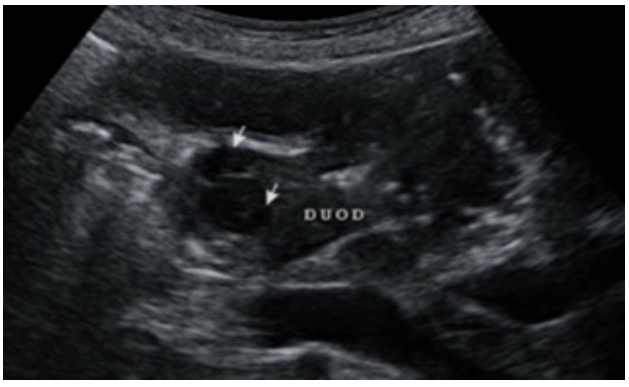

Figure 9: Cystic form of DD, US image, arrows indicate a cyst in the duodenal wall (DUOD)

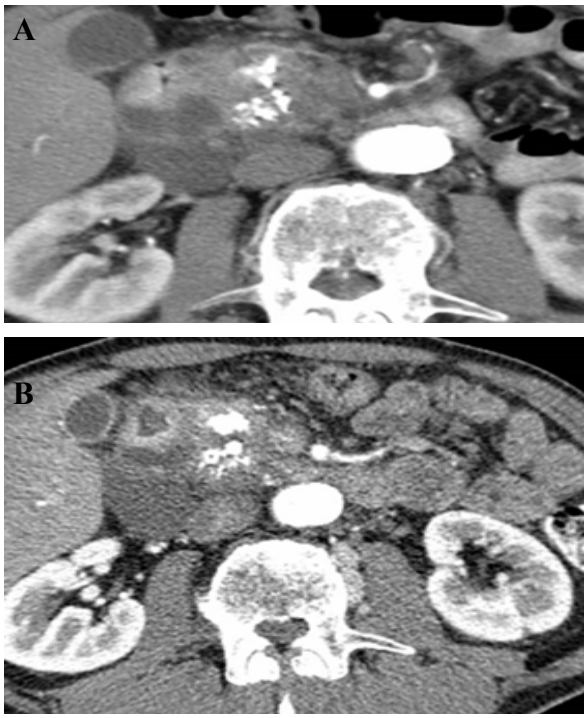

Figure 10: Cystic form of DD, MSCT image, arterial phase: a - contrast agent per os; b - contrast agent per os + 1 ml atropine intramuscular injection

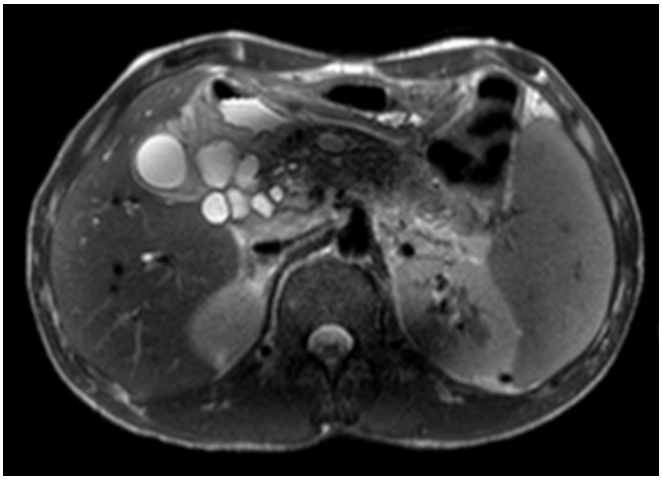

Figure 11: Cystic form of DD, MR image on T2 WI

Figure 12: Cystic form of DD the background of chronic calculous pancreatitis with calcifications of the main duct, MSCT image, native phase.

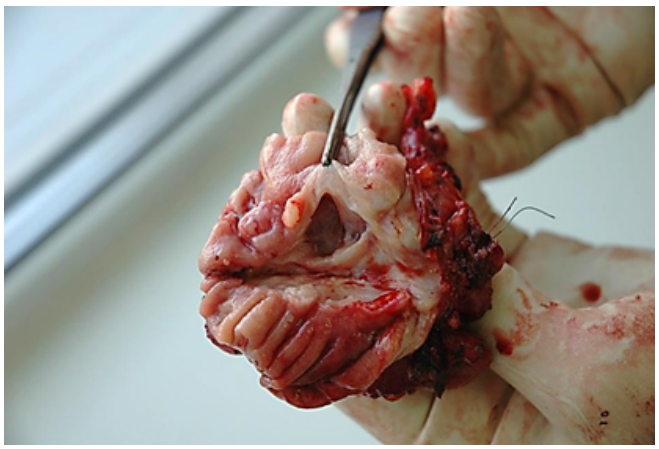

Figure 12: Cystic form of DD, macroscopic appearance

the non-invasive radiology methods provide comprehensive data for the duodenal dystrophy diagnostics (Figure 11). However, in the case of any diagnostic controversies, endosonography is recommended.In all cases, the patients had a long anamnesis of the disease or had complaints, therefore, a decision was made in favor of different kinds of surgical treatment.

Histological examination of remote pancreatoduodenal complexes with (Figure 13) the lesion of the duodenal wall showed lymphoid infiltration in some places of the duodenum mucosa and submucosa. The muscular layer was thickened, there were areas of heterotopia of the pancreas in the form of acini and ducts (Figure 14). The latter were enlarged, with signs of chronic inflammation around them.

In addition, there were detected the cystic lesions which walls did not have an epithelial lining for a greater length, and in some areas were lined with glandular epithelium (Figure 15). Apparently, it was a question of dilated ducts in these cases. The pancreatic tissue was severely sclerosis, dilated ducts, and signs of chronic inflammation.

Cystic lymphangioma

Lymphangioma is a benign tumor growing out of the lymph vessels and, as many researchers believe, occupying an intermediate position between the tumor and malformation. The localization of cystic lymphangioma in the pancreas is extremely rare. Cystic lymphangioma of the pancreas was first identified in 1913 by K. Koch [29] as a form of benign cyst secondary to blocked regional lymphatic ducts. T.S. Lyngdoh et al. [30] in their review of the literature for 2008 showed that since 1913 only 70 cases of cystic lymphangioma of the pancreas have been described. In recent years, these lesions began to come to light somewhat more, even the publication of 5 own cases was noted [31]. We conducted PUBMED research of the world literature from January 1, 2009 to July 31, 2020 using the keywords [Pancreatic lymphangioma], [diagnosis]. During this time, 42 cases of pancreatic cystic lymphangioma have been published. Thus, to date, July 2020, since the first publication in the literature, 112 cases of cystic lymphangioma of the pancreas have been published. Cystic lymphangioma is more common in the tail of the pancreas, in females, with the same frequency in all age groups.

In the present study, one case of the cystic pancreatic lymphangioma was observed. The preoperative diagnosis was not made, with probable presence of the serous cystadenoma.

Cystic lymphangioma is detected by all possible methods of radiology diagnostics. This nosologic form has clear diagnostic criteria well described in the literature. Radiologically it is seen the usually rounded lesion, primarily with the liquid content, which may be mono- or polycyclic form, in all cases a thin capsule is detected, with the lesion structure varying from uniformly liquid to liquid with single or multiple thin hyperechoic partitions (Figure 16 and 17). The bloodstream in the capsule and partitions of the lesion is not detected. It should be noted that the lesion structure is more clearly defined at US and MRI, as thin, "filmy"-like partitions are not always identified by computer tomography.

The lesion revealed in our study was a cystic lymphangioma, which was represented by a fibrous hyalinized wall with diffuse lymphoid infiltration (Figure 18A), in its thickness small compressed acini of the pancreas and cystic cavities with thin walls lined with flattened cells were revealed (Figure 18B).

All these features are characteristic of the lymphangiomas of any localization. However, despite the above clear criteria for radiology diagnostics of this nosological form, the complexity of verification of the disease, with its localization in the pancreas, can be explained by the fact that similar signs are also characteristic for true pseudocysts of the pancreas, as well as for serous and mucinous cystadenomas [32]. At present, there are no clear differen-

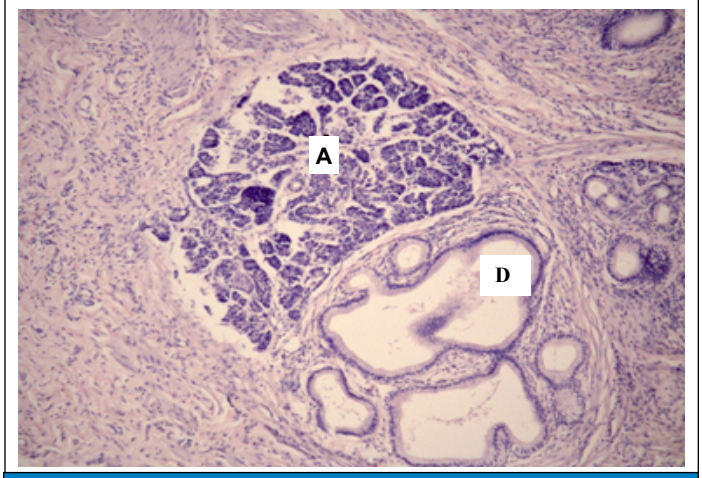

Figure 14: Histology, heterotopia of tissue (acini - A and ducts - D) of the pancreas in the duodenum, Hematoxylin and Eosin, x100 magnification

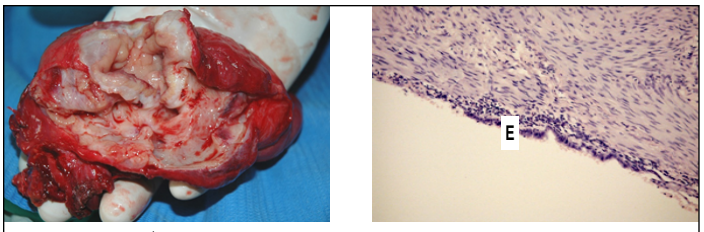

Figure 15: Cystic DD: a - removed specimen, b - micro specimen, dilated duct with sections of intact epithelium (E) - cyst in the duodenal wall, Hematoxylin and Eosin, x200 magnification

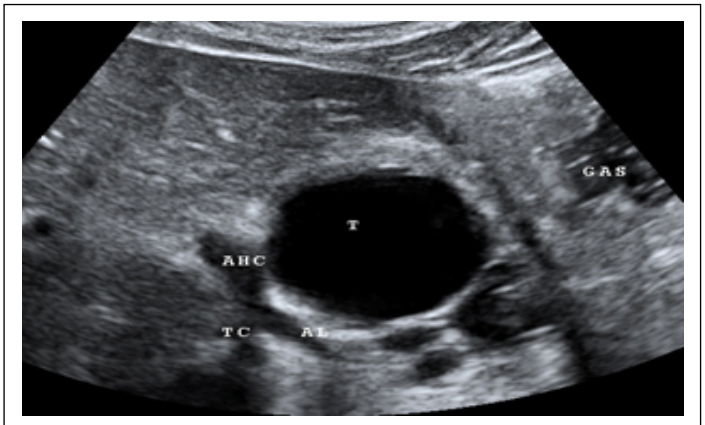

Figure 16: US image of cystic lesion with the celiac trunk and its branches in B-mode (T - tumor, AHC - arteria hepatica communis, AL - arteria lienalis, TC - truncus coeliacus, GAS - stomach)

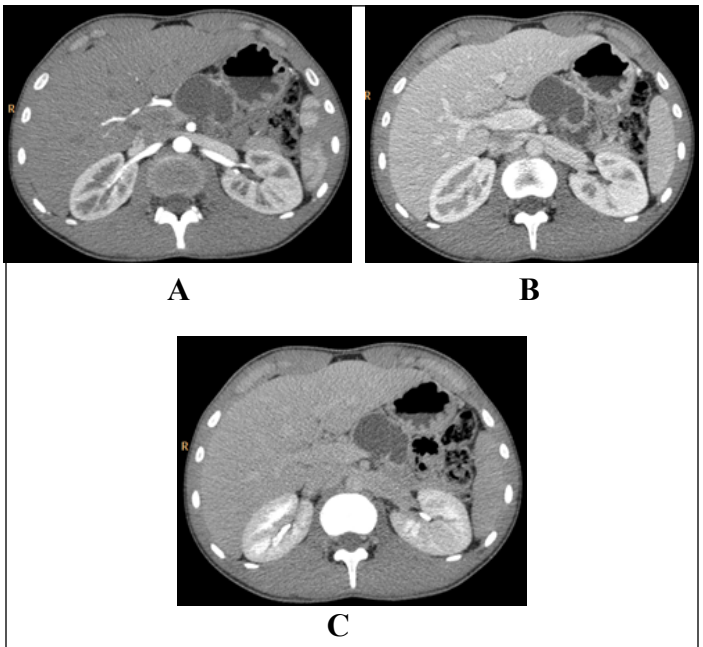

Figure 17: MSCT images of cystic lesion of pancreatic body-tail: a - arterial; b - venous; c - delayed phase

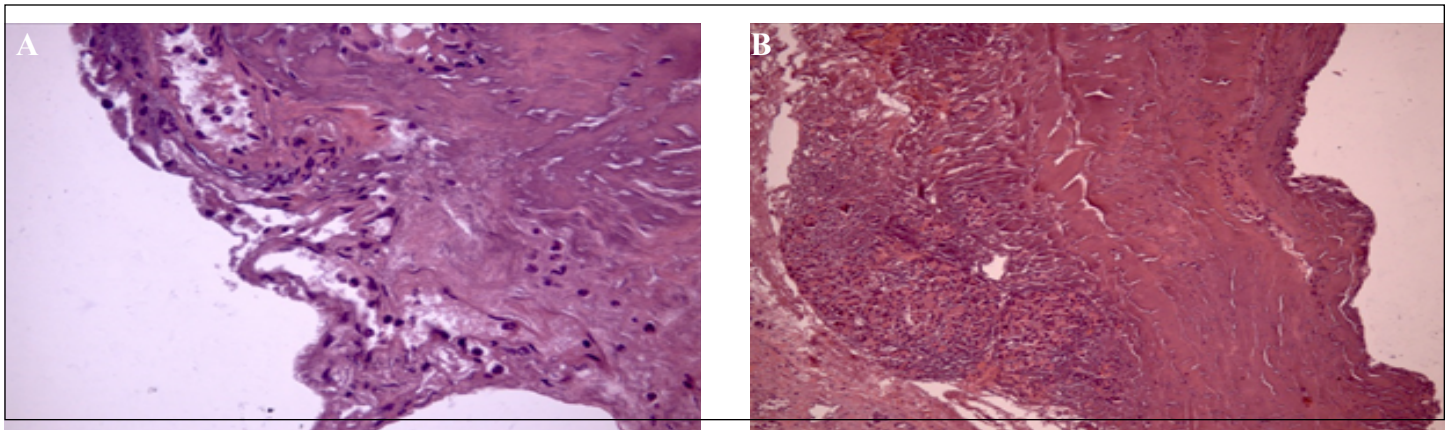

Figure 18: Histology, Hematoxylin and Eosin: a - the wall of cystic lymphangioma, represented by a fibrous hyalinized wall with diffuse lymphoid infiltration (x400 magnification); cyst wall with pancreatic tissue (x100 magnification)

tial diagnostic signs, contributing to the formulation of a correct diagnosis. The accurate verification of lesions can only be based on the results of the morphological examination. The analysis of radiology methods data must be carried out with an account to the anamnestic and clinical data, which, in the case of the pancreatic localization of lesions, can give additional information to verify it.

It should be mentioned that, since the pancreatic localization of cystic lymphangiomas is extremely rare, in most cases described in the literature, the diagnosis has never been preoperative as the doctors forgot to place this lesion in the differential diagnostic series.

Cystic teratoma

Teratoma is a rare tumor arising from the polypotent cells as a result of violations of embryogenesis. It develops on the basis of chipping off one of egg blastomers. Teratomas are composed of derivatives of three embryonic leaves: ectoderm, mesoderm, and endoderm [32]. In the pancreas, cystic teratomas are detected extremely rare.

Teratoma of this localization was first described by A.A. Kerr [33], but only in 1922 it was included by A. Primrose in the classification of cystic lesions of the pancreas [34]. Teratomas of the pancreas develop from embryonic pluripotent cells persisting in the tissue of the pancreas. The lesions can develop anywhere in the pancreas. L. Zhi et al. [35] searched MEDLINE and EMBASE and found only 50 reported cases of pancreatic cystic teratomas. At the moment, our analysis of international databases has revealed 3 such cases more. Thus, as of July 2020, 53 cases of pancreatic cystic teratoma have been published in the literature since the first publication.

Cystic teratomas of the pancreas, in contrast to teratomas of another localization, are often accompanied by clinical symptoms.

When analyzing the literature data on pancreatic cystic teratoma, we noted the fact that all authors point to the heterogeneity of the lesion and the variability of its radiology pattern, however, the description of this picture in the literature is rather scarce.

At A.V. Vishnevsky NMRC of Surgery, there was one case of the pancreatic cystic teratoma. It was diagnosed preoperatively according to all three methods of radiology diagnostics.

The cystic teratomas at US, MSCT, and MRI are characterized by irregular polycyclic shape, sharp and smooth contours and inhomogeneous structure with the presence of partitions of varying severity and dense unequally pronounced capsules. At US part of the chambers can have a homogeneous liquid content, while the other may have jelly-like content with uniformly expressed isoechogenic suspension, as well as the echosolid masses (Figure 19A). At duplex scan of the lesion, there were no blood flow in the capsule and septas (Figure 19B). At MSCT, the lesion may consist of liquid, fat, soft tissue and induration on the periphery, as well as calcinates. At contrast enhancing, the lesion does not accumulate a contrast agent (Figure 20). MRI also reveals the heterogeneity of the lesion structure represented by different tissues: with increased MR signal on T1 WI and sharply reduced on T2 WI and

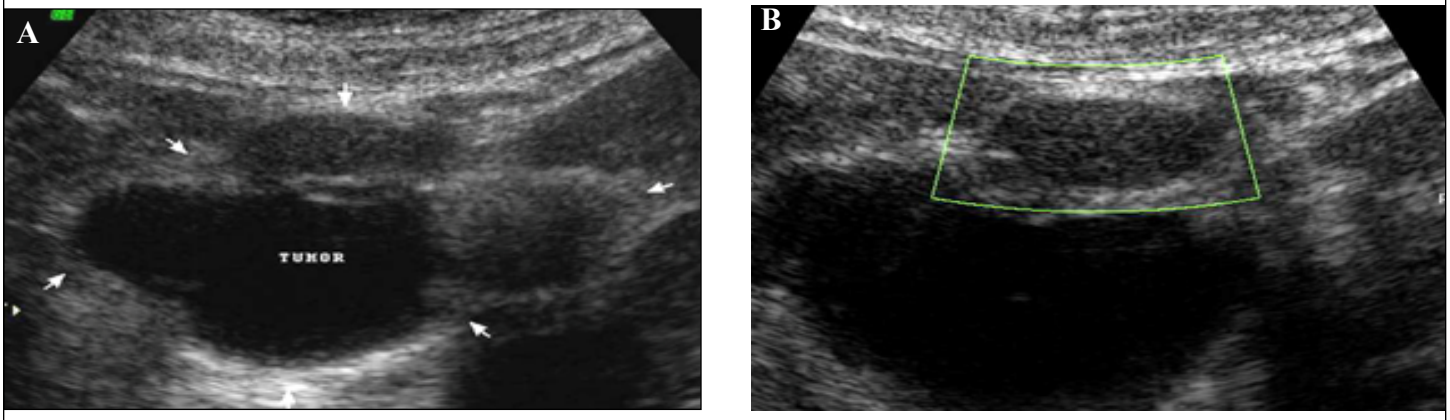

Figure 19: US image of pancreatic cystic teratoma: a - in B-mode (TUMOR indicated by arrows); b - blood flow is not detected either in the capsule and septas or in the echo-dense component at duplex scanning

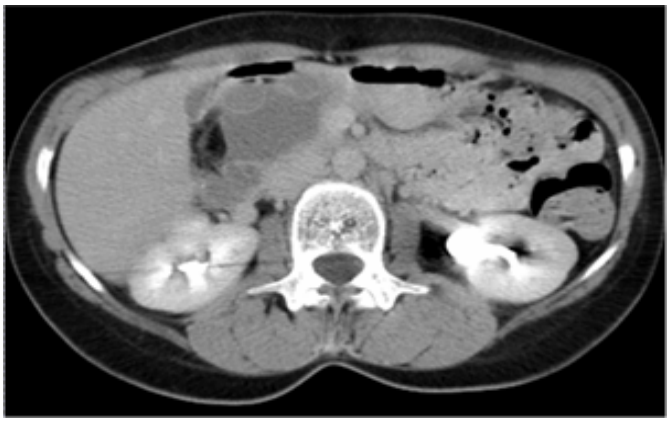

Figure 20: MSCT image of pancreatic cystic teratoma, delayed phase

STIR sequence; the fat component with increased MR signal on T2 WI and STIR and reduced on T1 WI; as well as the soft portions with reduced MR signal on T1 WI and moderately increased on T2 WI and clear on STIR (Figure 21). In contrast enhancement at MRI, the lesion does not accumulate a contrast agent; the accumulation is seen only in partitions in the deferred phase. It should be noted that the MRI enables clear differentiation of the content of various cavities without radiation exposure. MRI image without radiation exposure and the introduction of a contrast medium makes it possible to practically morphological verification of the lesion (Figure 22 and 23).

Considering pure benign nature of mature cystic teratomas, resection could be avoided if accurate diagnoses are made. Preoperative diagnostic imaging, however, is challenging and malignant potential cannot be ruled out [36].

Echinococcosis

Echinococcosis is a chronic and potentially life-threatening disease that often causes abdominal pain syndrome and damages different organs. According to various authors, the localization of echinococcal cysts in the pancreas is extremely rare and occurs in the range of 0.1-0.2% to 1-2% of echinococcal lesions [37,38]. Most of these masses are solitary (90%–91%) [39].

An addition to the rarity of this condition, despite clearly defined radiology criterias for echinococcal cysts, their diagnosis is challenging due to the similarity of imaging with much more common pancreatic cystic masses [40].

So, Y.B. Kattan [41] analyzed the diagnosis and treatment of two patients with morphologically verified echinococcus of the pancreas in 1975. Using the X-ray examination methods available at that time, taking into account the clinic and anamnesis, in the first case, adenocarcinoma of the pancreatic head was diagnosed before the operation, in the second, with the localization of the parasite in the distal part of the pancreas, splenomegaly. Soin et al. [38] could already apply a vast arsenal of the most modern diagnostic imaging methods in 2019 . But, nevertheless, applying at the first stage ultrasound and computed tomography, the results were uncertain and a broad differential diagnosis was given, which did not include echinococcosis. Further, magnetic resonance imaging

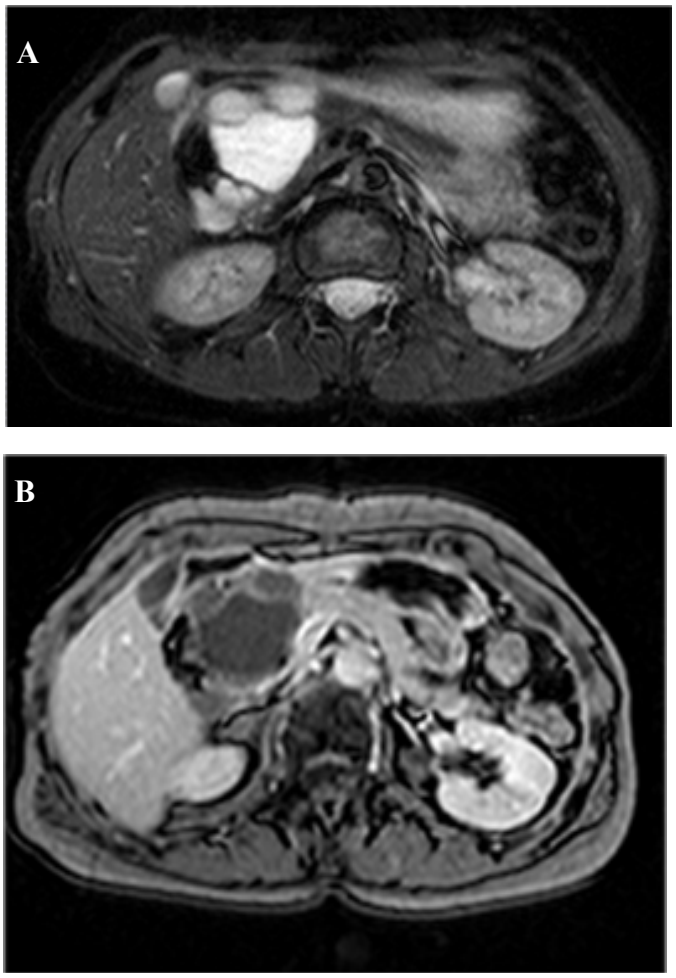

Figure 21: MRI images of pancreatic cystic teratoma: a - T2 WI; b - T1 WI

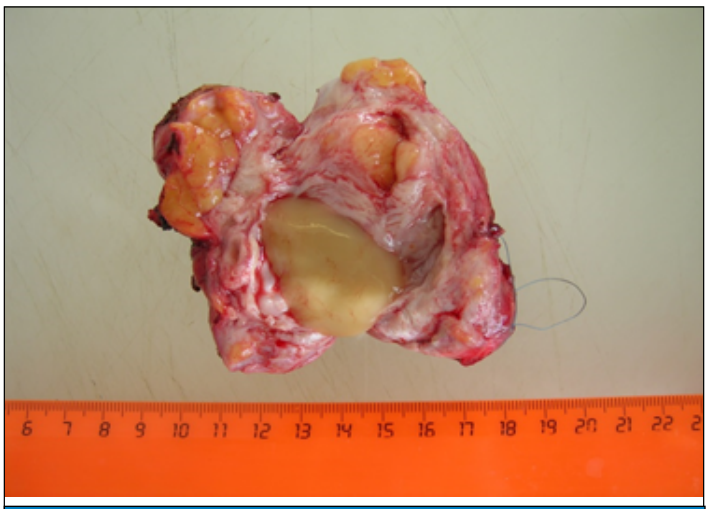

Figure 22: Pancreatic cystic teratoma, sectional view

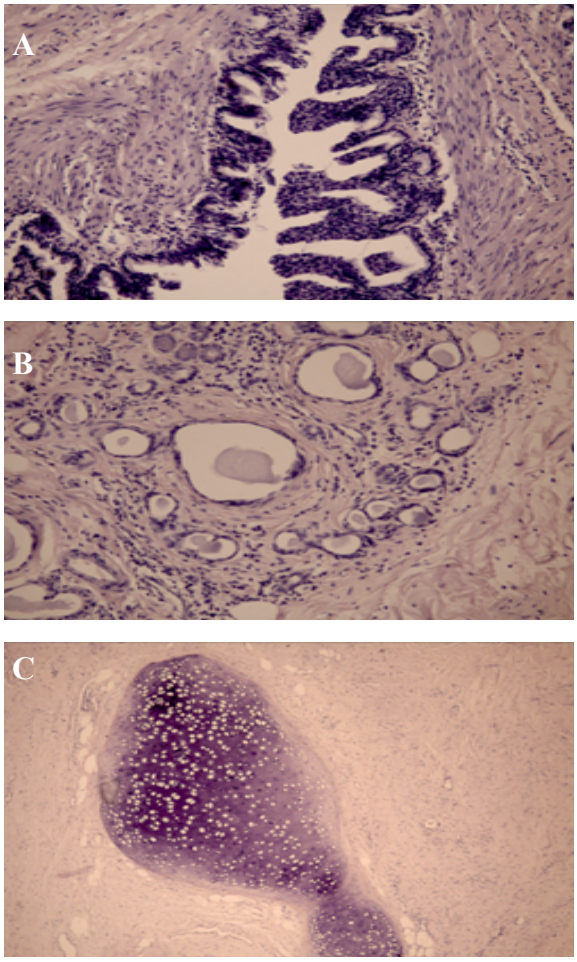

Figure 23: Histology, Hematoxylin and Eosin: a - cyst wall lined with stratified squamous epithelium (x200 magnification); b - mucous and protein glands (x200 magnification); c - areas of cartilaginous tissue (x50 magnification).

recorded a cystic lesion and only endoscopic ultrasound examination showed a cystic structure with thin curvilinear floating membranes corresponding to the "water lily" sign, a classic pathognomonic sign of echinococcosis. Which was confirmed by the results of enzyme-linked immunosorbent assay. Thus, in spite of the fact that 45 years have passed, the arsenal of diagnostic modalities has expanded significantly, the quality of the images obtained at the present time is extremely high; nevertheless, the diagnosis of pancreatic echinococcosis is still difficult. This can be explained by the fact that physicians in the Western world (nonendemic regions) who have not seen a pancreatic hydatid in their practice may not even consider it in the differential diagnosis. In nonendemic areas, a past history of traveling or immigration should make physicians

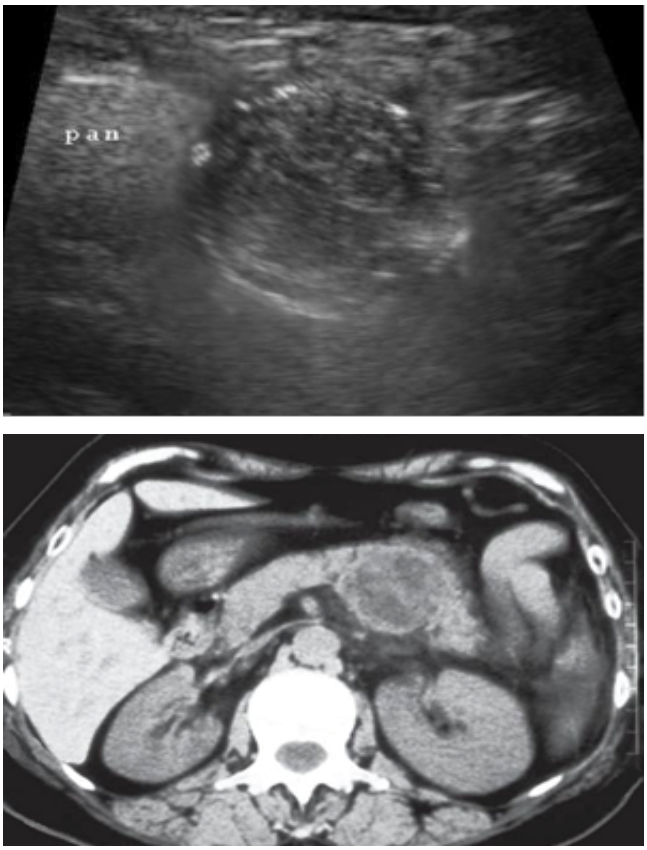

Figure 24: Echinococcus in the tail of the pancreas: a - US image (pan - pancreas); b - MSCT image

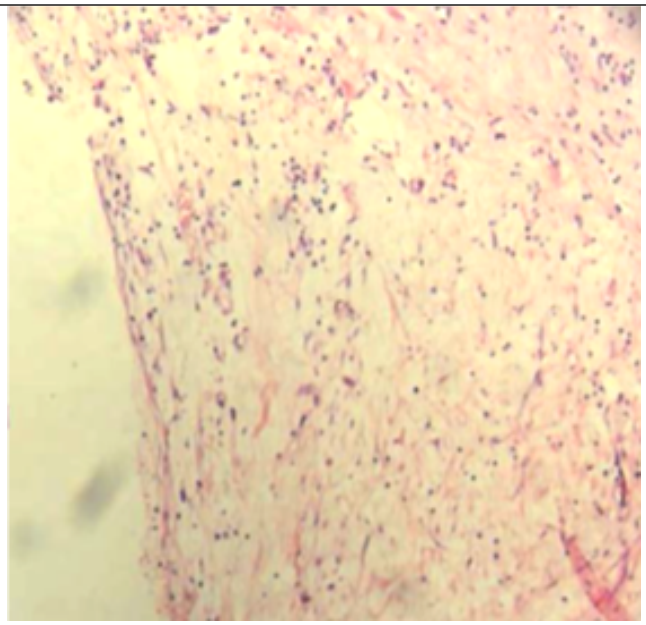

Figure 25: Histology of parasitic cyst with membranes. Hematoxylin and Eosin stain, x50 magnification

suspect the possibility of hydatid disease [42].

At A.V. Vishnevsky NMRC of Surgery, two cases of echinococcosis lesions of the pancreas were observed. In one case, there was a combined damage with the liver; the proper preoperative diagnosis was made. In another case, there was the preoperative diagnosis of pseudocyst in a 73-year-old woman with chronic pancreatitis. Misdiagnosis verification of the diagnosis was due to the presence of dead echinococcus against the background of chronic pancreatitis.

Diagnostic criteria of echinococcal cysts have been analyzed well enough when examining patients with cysts of the typical localization (liver and lungs). Radiologically this lesion is a spherical liquid lesion (at the unilocular echinococcosis), which may be of the heterogeneous content when it is difficult to distinguish it from true or pseudocyst cysts (Figure 24). The preoperative differential diagnostics is most often based on the presence of a dual capsule lesion due chitin (outer) and germinative (inner) shells or identifying typical associated parietal bubbles. Echinococcosis shell calcification is rather common. Morphological verification of echinococcus is also beyond doubt (Figure 25).Thus, the diagnostics of echinococcal cysts of the pancreas is usually troubled by the fact that researchers "forget" to put such a diagnosis as echinococcosis in a diagnostical series due to the rare occurrence of such localization of this disease.

Conclusions

A.V. Vishnevsky NMRC of Surgery (Moscow) has fifteen years of experience in diagnosing and treating such rare forms of pancreatic cystic neoplasms as the IPMN, SPPT, and the cystic form of DD. It should be pointed out that, due to the accumulation of knowledge and the expert analysis, preoperative diagnostic accuracy has been growing.

Thus, the complete radiology examination including US, MSCT, and MRI, makes it possible to diagnose and differentiate various forms of pancreatic cystic diseases. At suspected presence of a rare pancreatic lesion, the radiology preference should be given to MRI. Criteria for radiology diagnostics of the IPMN and DD cystic form have been well defined. The formulation of the SPPT diagnostic criteria requires the accumulation of a greater number

of observations and an analysis of the corresponding information. Anamnesis of the disease is an important supplement to the SPPT radiology diagnostics. The knowledge of the entire spectrum of possible morphological forms of pancreatic cystic lesions by the diagnostician will make it possible to assume, when making a diagnosis, an extremely rare form of the disease.

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