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Pancreatic Perivascular Epithelial Cell Tumor (PEComa) with Liver Metastasis: a Case Report and Literature Review

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Aim: to present a clinical case of pancreatic PEComa with liver metastases with an emphasis on morphological features, radiology diagnostic methods and features of treatment tactics, allowing practitioners to get an idea about this rare mesenchymal tumors composed of "perivascular epithelioid cells", and a review of literature data on pancreatic PEComa, including 32 author's cases.
General statements. A 22-year-old woman who was surgically treated for pancreatic head PEComa with infiltration of the duct of Wirsung, common bile duct, duodenal wall, focal invasion into the blood and lymphatic vessels, and perineural space infiltration. Synchronously, metastases were detected in both liver lobes, for which she took Everolimus for 6 years under the control of radiology methods with dose adjustment and frequency of administration. At the A.V. Vishnevsky National Medical Research Center of Surgery, the patient underwent right-sided hemihepatectomy and atypical resection of II–III liver segments.
Conclusion. To determine clearer criteria for the diagnosis and differential diagnosis of PEComa, to identify criteria for the malignancy of these tumors, to develop treatment tactics and further dynamic monitoring, a set of statistical data of significant group and randomized clinical trials are needed. Pancreatic PEComas in this group are extremely rare and often have a benign course. The presented clinical case demonstrates the most malignant form of this tumor with localization in the pancreas and liver metastases.
Keywords: perivascular epithelial cell tumor, pancreas, PEComa, diagnosis, treatment
Conflict of interest: the authors declare no conflict of interest.

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Периваскулярная эпителиально-клеточная опухоль (ПЭКома) поджелудочной железы с метастазами в печень: клиническое наблюдение и обзор литературы

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Цель исследования: представить клиническое наблюдение ПЭКомы поджелудочной железы (ПЖ) с метастазами в печень с акцентом на морфологические особенности, данные лучевых методов исследования и особенности тактики лечения, позволяющие практическим врачам составить представление об этой редкой опухоли, состоящей из «периваскулярных эпителиоидных клеток», а также данных литературы ПЭКом ПЖ, включающий 32 наблюдения.
Основные положения. Женщина 22 лет перенесла хирургическую операцию по поводу ПЭКомы головки ПЖ с инфильтрацией вирсунгова протока, общего желчного протока, стенки двенадцатиперстной кишки, очаговой инвазией кровеносных и лимфатических сосудов, инфильтрацией периневрального пространства. Синхронно были выявлены метастазы в обе доли печени, по поводу которых в течение 6 лет принимала Эверолимус под контролем лучевых методов исследования с коррекцией дозы и кратности приема. В НМИЦ хирургии им. А.В. Вишневского пациентке выполнена правосторонняя гемигепатэктомия и атипичная резекция II–III сегментов печени.
Заключение. Для определения более четких критериев диагностики и дифференциальной диагностики ПЭКом, выявления критериев злокачественности этих опухолей, выработки тактики лечения и дальнейшего динамического наблюдения необходим набор статистически значимой группы и проведение рандомизиро-

ванных клинических исследований. ПЭКомы ПЖ встречаются крайне редко и чаще имеют доброкачественное течение. Представленное клиническое наблюдение демонстрирует наиболее злокачественную форму этой опухоли с локализацией в ПЖ и метастазами в печень.
Ключевые слова: периваскулярная эпителиально-клеточная опухоль, поджелудочная железа, ПЭКома, диагностика, лечение
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Introduction

The International Classification of Bone and Soft Tissue Tumors identified a family of mesenchymal tumors, designated by the general term PEComas in 2002 [1]. Perivascular epithelial cell tumors (PEComs) belong to the category of mesenchymal tumors consisting of epithelioid or spindle-shaped cells with a transparent or focal-granular eosinophilic cytoplasm, with round or oval nuclei and inconspicuous nucleoli in the center [2]. The location of epithelial cells is typical closer to the vessels for PEComa; spindle-shaped cells, similar to smooth myocytes, as a rule, are determined at a distance from the vessels [3]. The tumor stroma may be hyalinized. PEComs express both melanocytic (HMB-45, Melan-A, MiTF) and muscle markers (SMA, desmin, myosin), as well as vimentin, estrogen receptors, etc. PEComs differ in the level of marker expression. Spindle cell-dominated PEComas show high expression of muscle markers and limited expression of melanocytic markers. In ordinary epithelioid cells, the expression of markers is opposite [4].

The PEComas group includes [5]:
– angioleiomyolipomas;
– clear-cell "sugar" tumor (CCST) of the lung;
– lymphangioleiomyoma;
– clear-cell myomelanocytic tumor of the uterus round ligament / liver falciform ligament;
– abdominopelvic sarcoma of perivascular epithelioid cells (unusual clear-cell tumor of the pancreas, rectum, peritoneum, uterus, vagina, thigh and heart).

The lesion most often occurs sporadically, but may also be associated with tuberous sclerosis (tuberous sclerosis complex, TSC). Tuberous sclerosis is a genetic disease that causes mental retardation, seizures, and the development of benign tumors in many organs [5, 6]. It is believed that about 10 % of PEComss are associated with tuberous sclerosis [7].

For the first time, according to G. Martignoni et al., special clear cell tumor cells were described in the work of K. Apitz in 1943 [2]. In a study by F. Bonetti et al. an important feature was emphasized — the growth of tumor cells occurs around thin-walled branching vessels [8]. The term "perivascular epithelioid cell" (PEC) has appeared. Further clinical studies have shown that in addition to the classical localization — lungs, kidneys and liver — tumors from perivascular epithelioid cells can be located in the pancreas. The first publication describing such a tumor localized in the pancreas was made by G. Zamboni et al. in 1996 [9]. The authors formulated a position on the family of mesenchymal tumors consisting of perivascular epithelioid cells and designated this group of tumors under the general term "PEComas".

The type of cells from which these tumors originate remains unknown. Normally, perivascular epithelioid cells are absent; the name refers to the characteristics of the tumor when examined under a microscope [5]. Early studies suggested that vascular walls [8] or specific muscle cells [10, 11] could be the source of PEComa development. Modern advances in embryology have shown that neural crest stem cells, capable of differentiating into muscle and melanocytic cells during development, can be a source of tumor cells [12]. Research in this area is ongoing.

Based on the characteristics of epithelial cells described above, PEComa is characterized by perivascular localization, often with a radial arrangement of cells around the lumen of the vessel [13]. PEComas have been found in the uterus, liver falciform ligament, gastrointestinal tract, kidneys, pancreas, pelvis, skull, vulva, prostate, thigh soft tissues, common bile duct, and heart. It is believed that PEComas can be localized in any organ but are most often found in the uterus. According to the literature, by the end of 2007, only 51 cases of PEComas had been described, 46 % of them were in the uterus body [3]. The final diagnosis of PEComa can only be made after a histological and often immunohistochemical study.

Some PEComas have malignant features, while others may be cautiously labeled as having "undetermined malignant potential". Establishing the malignant potential of these tumors remains

challenging, although criteria have been proposed [14]: size > 5 cm, infiltrative growth, hypercellularity, high-grade atypia, high mitotic index (> 1/50 HPF), invasion into vascularity and necrosis, as well as other findings that may indicate the possibility of malignant growth. When considering the criteria, all tumors were divided into three groups: benign, tumors with an unclear growth potential, and malignant. Tumors that do not have any risk factor are benign; tumors with one of these features are classified as tumors of unclear prognosis; tumors with two or more factors are malignant [15]. J.S. Bleeker et al. after analyzing 234 cases of PEComas described in the English-language literature, concluded that only tumor sizes over 5 cm and high mitotic activity (1/50 HPF) were significant recurrence factors after tumor removal [16]. The same authors, in continuation of their studies, report that in the presence of criteria for malignancy, 71 % of patients develop metastases or recurrence [17]. J.K. Schoolmeister et al. indicate the importance of four or more factors for determining a malignant lesion [18].

Due to the extremely rare occurrence of pancreatic PEComa, we consider it appropriate to present our own clinical case of a patient with this tumor.

Clinical case

Patient T., 22-years-old woman.

Upon admission to the clinic, the patient does have any complains.

Anamnesis. In September 2016, the patient complained of weakness and occasional tarry stools. On examination, pallor of the skin was noted, general blood test showed hemorrhagic anemia. Gastroscopy revealed bleeding in the upper gastrointestinal region.

Pancreatic head region MSCT revealed a hypervascular solid tumor with clear and even contours, with a maximum size of 5.4 cm. Enlarged parapancreatic lymph nodes up to 4.5 cm in size with an altered structure were determined. There were multiple hypervascular metastases in both lobes of the liver: small — in left lobe segments, the largest — in segments VI–VII of the liver.

The biopsy of pancreatic head lesion was performed. According to the histology: perivascular epithelioid cell tumor.

The patient applied to the University Medical Clinic of the Johannes Gutenberg University of Mainz (Germany). On November 23, 2016, a ***pylorus-preserving pancreaticoduodenal resection (pPDR) and diagnostic atypical resection of the liver IV segment were performed.***

Diagnosis according to histological examination of the removed organocomplex: primary perivascular epithelioid cell tumor of the pancreatic head with infiltration of the Wirsung duct, common bile duct, duodenal wall, focal invasion into blood and lymphatic vessels, infiltration of the perineural space, 9/20 positive local-regional metastases in the lymph nodes, liver metastases. Staging: pT3: duodenal infiltration; pN1: 9/20 resected lymph nodes affected; pM1: multiple liver metastases; resection status of the primary tumor R0 with ongoing multiple liver metastases. Immunohistochemical examination: tumor cells show a well-defined nuclear positivity for TFE3; some cells with AMACR expression positivity, while CK7, pan-cytokeratin, vimentin were negative; small clusters with CD10 positivity; pronounced positivity of HBM45 and E-caderin. Conclusion: Primary Perivascular Epithelioid Cell Tumor (PEComa).

Oncoconsilium dated December 20, 2016: PECs are stably resistant to both traditional radiation therapy and chemotherapy. In the postoperative period, targeted therapy with Everolimus is indicated (current level — 7.1 ug/L, target — 8–15 ug/L). The patient is currently taking the drug.

MSCT of the abdomen and chest were performed annually. Since 2021, the stabilization of the process has been noted in the form of a persistent absence of the growth of liver metastases. MRI dated June 21, 2022: multiple hypervascular metastases in both lobes of the liver. Against the background of taking Everolimus, minimal regressive dynamics was noted in relation to the size of metastases, however, active vascularization of the foci is still noted as a manifestation of the vital tumor tissue.

The patient was hospitalized for planned surgical treatment of liver metastases at the A.V. Vishnevsky National Medical Research Center of Surgery.

On examination. The general condition is satisfactory. Consciousness is clear, the patient is contact, adequate, oriented in place, time and self. The skin is moderately pale, clean. In the lungs, vesicular breathing, no wheezing. The number of respiratory movements — 16 per minute. Heart sounds are muffled, rhythmic. Heart rate — 72 beats per min. Blood pressure — 120/70 mmHg.

Family anamnesis: the father's family history is unknown, the mother's side is known to have a chromosomal abnormality (X-chromosome trisomy), grandmother and great-grandmother had cervical cancer and thrombophlebitis.

Laboratory data. Indicators of blood biochemistry, coagulograms within the physiological norm. Complete blood count: ESR — 29 mm/h, leukocytes — 4.3×10^9/L, hemoglobin — 108 g/L.

Description of MRI data performed on an outpatient basis in another institution. A trace amount of free fluid in the right lateral canal and along the contour of the liver. No encysted fluid was found in the abdominal cavity. The liver is proportionally enlarged in size, the right lobe craniocaudally — 17 cm, moderate steatohepatosis (HFF 12 %). Accumulation of the contrast agent in the unchanged liver parenchyma in the extracellular phases is typical, accumulation and excretion of the hepatospecific contrast agent by the liver in the hepatobiliary phase is normal. In both lobes of the liver, multiple foci are determined, in the segments of the left lobe — small, no more than 4 mm, in segments VI–VII — two large, 77 mm (series 9 slice 87) and 65 mm (series 9 slice 92), and multiple small similar foci — in segments of the right lobe (Fig. 1). These foci have an increased MR signal in T2-weighted images (WI) reduced in T1-WI (large lesions in the right lobe with a heterogeneous MR signal). During intravenous contrasting, large foci in the right lobe (VI–VII segments) accumulate contrast substance mainly along the periphery, in the arterial phase of the study, the area in the center of the foci with little or no accumulation of contrast agent (decay). Smaller lesions are hypervascular and wash out occurs in the venous phase of the study. In the hepato-specific phase of the scan, all secondary lesions showed no signs of contrast agent accumulation. Subocclusion of the posterior sectoral branch of the portal vein, early arteriovenous shunting of blood in the VI–VII segments of the liver.

The gallbladder is not visualized. Intrahepatic bile ducts are not dilated.

In the subhepatic space, a loop of the small intestine is traced, carrying a hepaticojejunostomy, a common hepatic duct has a diameter of 3 mm.

Condition after pPDR, pancreatic stump up to 18 mm thick, without obvious focal changes. The accumulation of contrast agent is typical. Pancreatic duct — 2 mm. The area of pancreaticojejunostomy without features.

The spleen is enlarged (92 × 60 × 111 mm), the parenchyma is not changed, small additional lobules of the spleen.

The walls of the stomach, small intestine, colon without signs of focal / infiltrative changes in the walls, the area of gastrojejunostomy without features.

Kidneys and adrenal glands are not changed.

Contrasting of the great vessels at the study level is homogeneous.

Lymph nodes at scan levels are small.

Bones at the study level without obvious changes of a metastatic nature.

Conclusion. Condition after pPDR. Multiple bilobar hypervascular liver metastases. Subocclusion of the posterior sectoral branch of the portal vein. Hepatomegaly. Moderate steatohepatosis. Splenomegaly.

Ultrasound of the liver. The liver is not enlarged in size: the anterior-posterior size of the right lobe is 133.7 mm, the left lobe is 54.5 mm, the contours are clear, even, the structure of the liver parenchyma is diffusely compacted. Intrahepatic and extrahepatic bile ducts are not dilated.

In both liver lobes multiple focal lesions are determined. The largest is localized in the VI–VII liver segments. The lesion is located subcapsular, two-nodular, solid with clear, somewhat bumpy contours, with a total size of 103.0 × 74.2 mm. The lesion has heterogeneous echo density, there is a slight decrease in echogenicity towards its periphery (Fig. 2A). The course of the right hepatic vein is traced to it, as well as segmental branches of the right branch of the portal vein and the proper hepatic artery involved in its blood supply (Fig. 2B). Arterial and venous signals are located in the lesion structure at duplex scanning, localized mainly at the pole, where the feeding vessels are traced (Fig. 2C).

Conclusion. Condition after pPDR for perivascular epithelioid cell tumor in 2016. Considering the history data, metastases of PEComa in both lobes of the liver.

Gastroduodenoscopy. The duodenal bulb was shortened due to a previous operation. Duodeno-enteral anastomosis is located immediately behind the pylorus, is not available for inspection. The adductor loop of the small intestine is not visualized. The efferent loop of the small intestine was examined at a distance of 30 cm: the lumen was not deformed, it contained a small amount of mucous content. Peristalsis is active, can be traced along all walls, the mucous membrane of the small intestine is pink, velvety.

Conclusion. Condition after pPDR regarding PEComa from 2016.

Based on the survey data, the diagnosis was made: bilobar metastases of perivascular epithelioid cell tumor of the pancreas.

Surgery was performed: *right-sided hemihepatectomy, atypical resection of II–III liver segments.*

Operation description. J-incision access with excision of a part of the old postoperative scar, the abdominal cavity is opened in layers. There is a moderate adhesive process after the previous operation. Pathological changes in the stomach, pancreas, spleen, small and large intestine were not detected. Kidneys without features. The

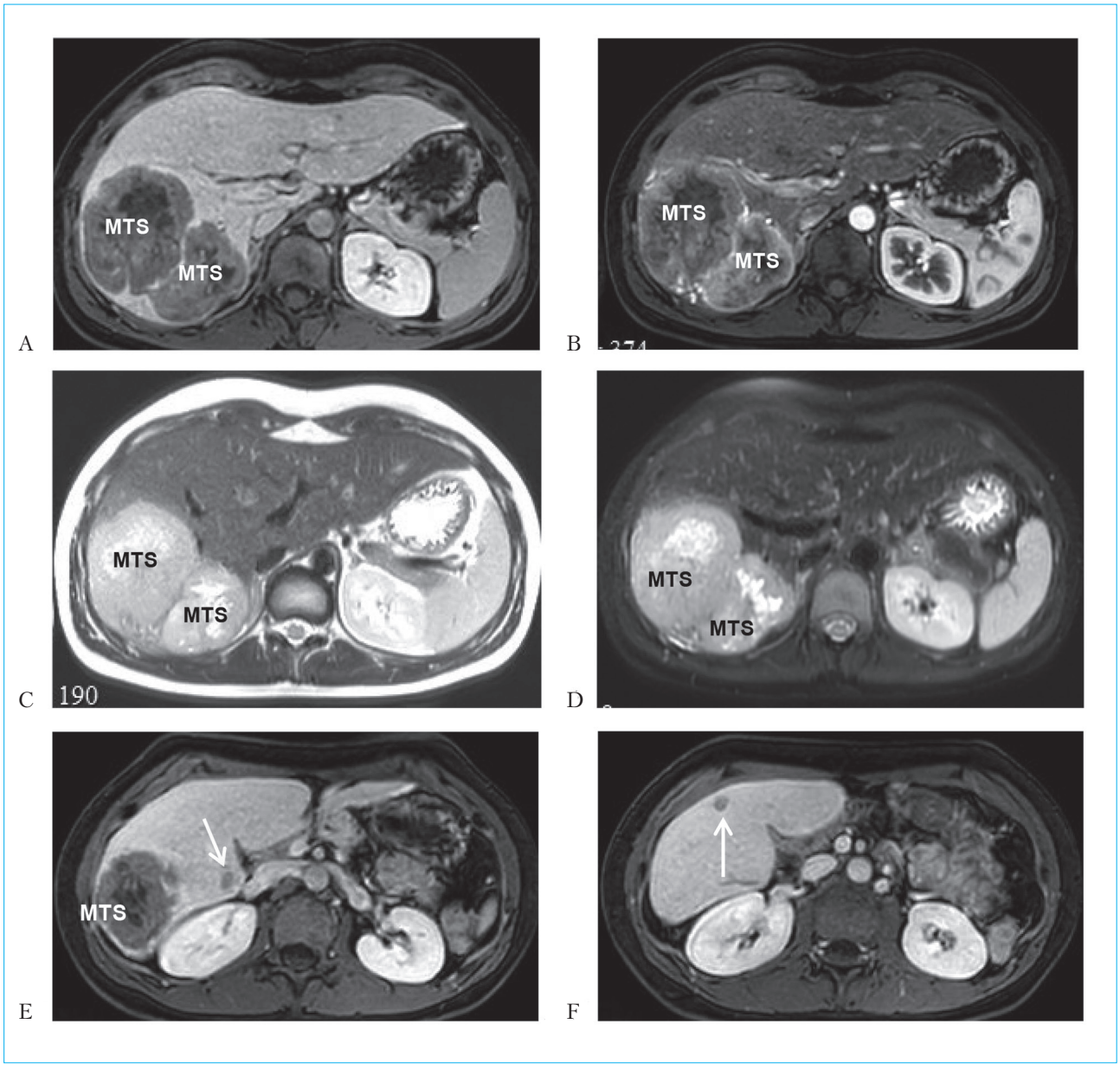


Fig. 1. MRI of liver metastases, two large foci in the VI–VII liver segments: A — T1-WI; B — T1-WI with contrast enhancement; C — T2-WI; D — T2-WI in fat suppression mode; E — large focus of VII and small focus of VIII liver segments, T1-WI; F — small focus of V liver segment, T1-WI

Рис. 1. МРТ-изображения метастазов в печени (MTS) — два крупных очага в VI–VII сегментах печени: A — T1-ВИ; B — T1-ВИ с контрастным усилением; C — T2-ВИ; D — T2-ВИ в режиме жироподавления; E — крупный очаг VII и мелкий очаг VIII (указан стрелкой) сегментов печени, T1-ВИ; F — мелкий очаг V сегмента печени (указан стрелкой), T1-ВИ

para-aortic lymph nodes were not enlarged. The liver is of normal size and color. |In VI–VII liver segments there are two dense nodes merging with each other, 4 × 3 and 5 × 5 cm in size (Fig. 3A). There are also small foci on the border of II–III liver segments. The gallbladder was previously removed. An intraoperative ultrasound was performed. In the VI–VII liver segments there are focal lesions of a solid structure, heterogeneous echo density (mainly increased echogenicity with a decrease in density towards the periphery). Multiple small foci are also determined in the V and VIII liver segments.

Atypical resection of II–III liver segments with metastatic foci was performed. An urgent histological examination was performed: tumor foci, most likely belonging to metastases of a perivascular epithelioid cell tumor. Liver tissue outside

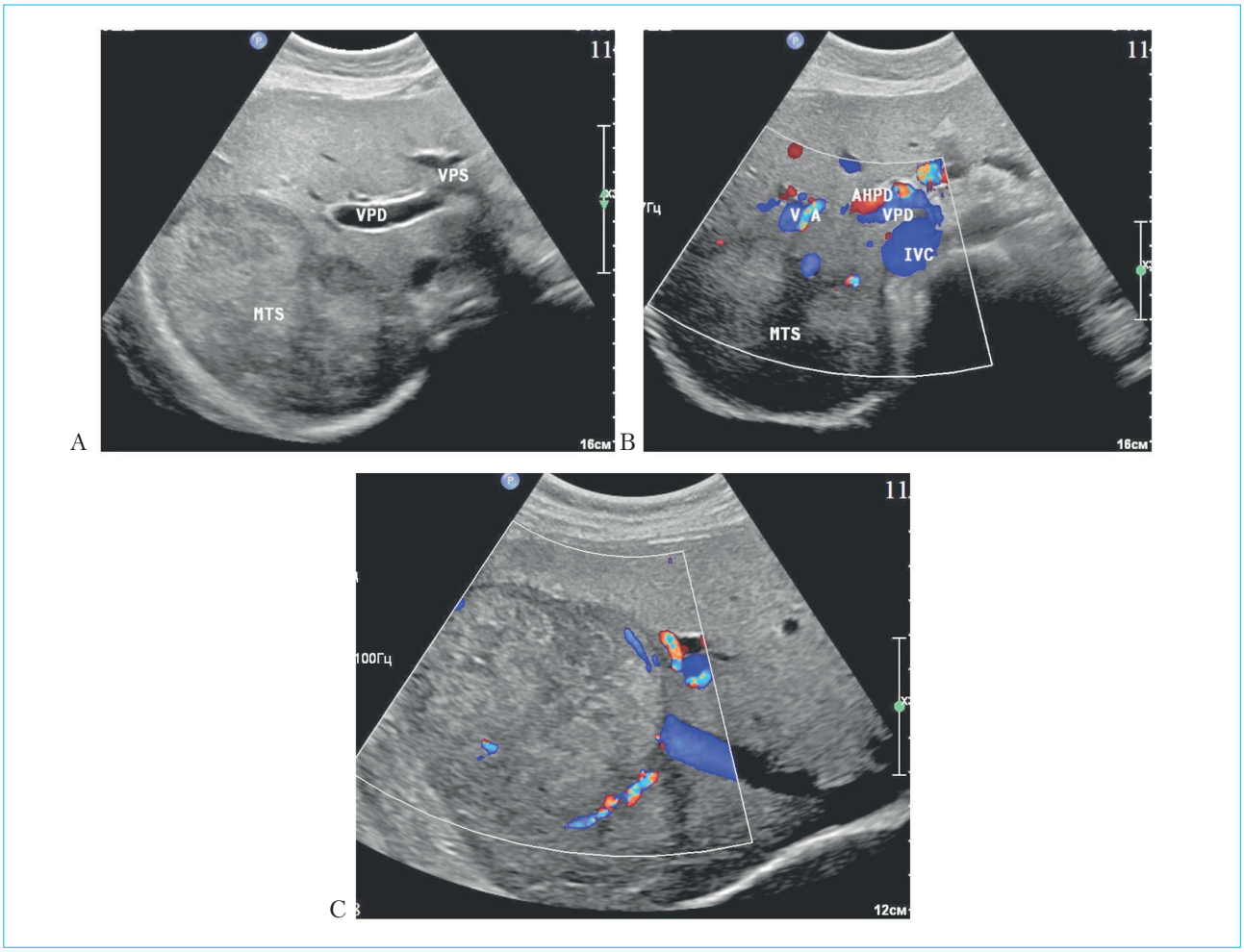


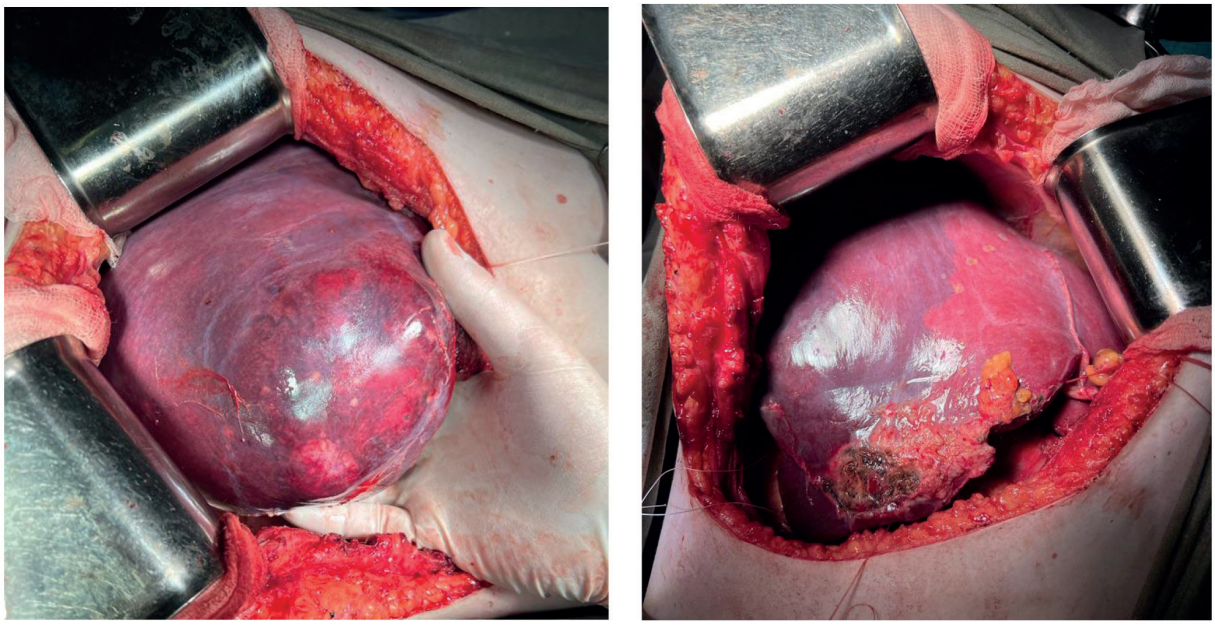
Fig. 2. Ultrasound images of metastasis (MTS) of VI–VII liver segments: A — image in B-mode; B — Color Doppler Imaging, segmental veins and arteries (VA) are determined from the right branch of the portal vein (VPD) and the right own hepatic artery (AHPD), supplying the metastasis (IVC — inferior vena cava); C — Color Doppler Imaging, arterial and venous signals are located in the lesion structure, localized mainly at the pole, where the feeding vessels are traced

Рис. 2. УЗ-изображения метастаза (MTS) VI–VII сегментов печени: A — изображение в B-режиме; B — в режиме цветового допплеровского картирования, определяются сегментарные вены и артерии (VA) от правой ветви воротной вены (VPD) и правой собственной печеночной артерии (AHPD), кровоснабжающие метастаз (IVC — нижняя полая вена); C — в режиме цветового допплеровского картирования, в структуре образования лоцируются артериальные и венозные сигналы, локализованные преимущественно у полюса, где прослеживаются питающие сосуды

the tumor with large droplet fatty degeneration of hepatocytes.

With the help of ultrasound, a projection of the middle vein on the surface of the liver is planned. The right hepatic artery is tortuous, the right segmental arteries depart early from it. The right anterior and posterior hepatic arteries and the right branch of the portal vein were isolated, tied, and transected, after which a distinct demarcation appeared between the liver lobes (Fig. 3B). The right hepatic ducts were isolated and ligated. The right liver lobe was mobilized, while 5 short hepatic veins were ligated and crossed. The right hepatic vein was isolated, occluded, divided and sutured with a 4-0 twist prolene. Five millimeters away from the demarcation line (the latter fully corresponded to the borders of the middle hepatic vein outlined under ultrasound), the liver capsule was incised with an electric knife, the tubular structures in the resection plane were partially coagulated, clipped and ligated. A right-sided hemihepatectomy was performed. Parenchymal hemostasis by coagulation and stitching.

Histology. On microscopic examination, the liver tumor nodule was composed of epithelioid pronouncedly polymorphic cells with light



A B

Fig. 3. Intraoperative field view: A — metastases in the VI–VII liver segments; B — marking of the median hepatic vein and a demarcation line

Рис. 3. Вид в интраоперационном поле: А — метастазы в VI–VII сегментах печени; В — разметка срединной печеночной вены и линией демаркации

eosinophilic cytoplasm and polymorphic hyperchromic nuclei, sometimes containing eosinophilic nucleoli (Fig. 4A). Tumor cells form solid and alveolar structures (Fig. 4B), spreading infiltratively to the underlying liver tissue and overgrowing the bile ducts. There are foci of necrosis, occupying up to 50 % of the area of the tumor in the section. The number of mitoses, including pathological ones, was 5 per 10 representative fields of view at a magnification of 400 (Fig. 4C). Surrounding liver tissue with preserved histoarchitectonics. Liver resection margin without tumor elements (Fig. 4D).

For the purpose of differential diagnosis, an immunohistochemical study was performed. Tumor cells exhibit the following reactions: HMB-45 (clone HMB-45, Cell Marque) — pronounced cytoplasmic expression (Fig. 5A); S100 (polyclonal, DAKO) — weak cytoplasmic expression; Ki67 (clone MIB-1, DAKO) — nuclear expression at 15 %.

Tumor cells are negative for: Arginase-1 (clone SP156, Cell Marque) (Fig. 5B); PanCytokeratin (clone AE1/AE3, Cell Marque) (Fig. 5C); CD34 (clone QBEnd/10, Cell Marque); Cytokeratin 19 (clone A53-B1/A2.26, Cell Marque); TFE3 (clone MRQ-37, Cell Marque).

Conclusion. Taking into account the clinical data, as well as the results of histological and immunohistochemical studies, the morphological picture of metastasis of a malignant perivascular epithelioid cell tumor (PEComa) to the liver.

The postoperative period proceeded without complications. The hydrothorax on the right side identified before the operation and a small amount of free fluid in the subhepatic space regressed independently. The department carried out analgesic, symptomatic therapy, bandaging of postoperative wounds with antiseptic solutions. In a stable satisfactory condition, the patient was discharged for outpatient observation on the 14th day after surgery.

At the control study a month after surgery, no pathological changes were detected.

Discussion

PEComas that occur in the pancreas are extremely rare [2]. To date, only 32 such cases have been identified [9, 14, 19–47]. All of them are described in the form of clinical cases. In the

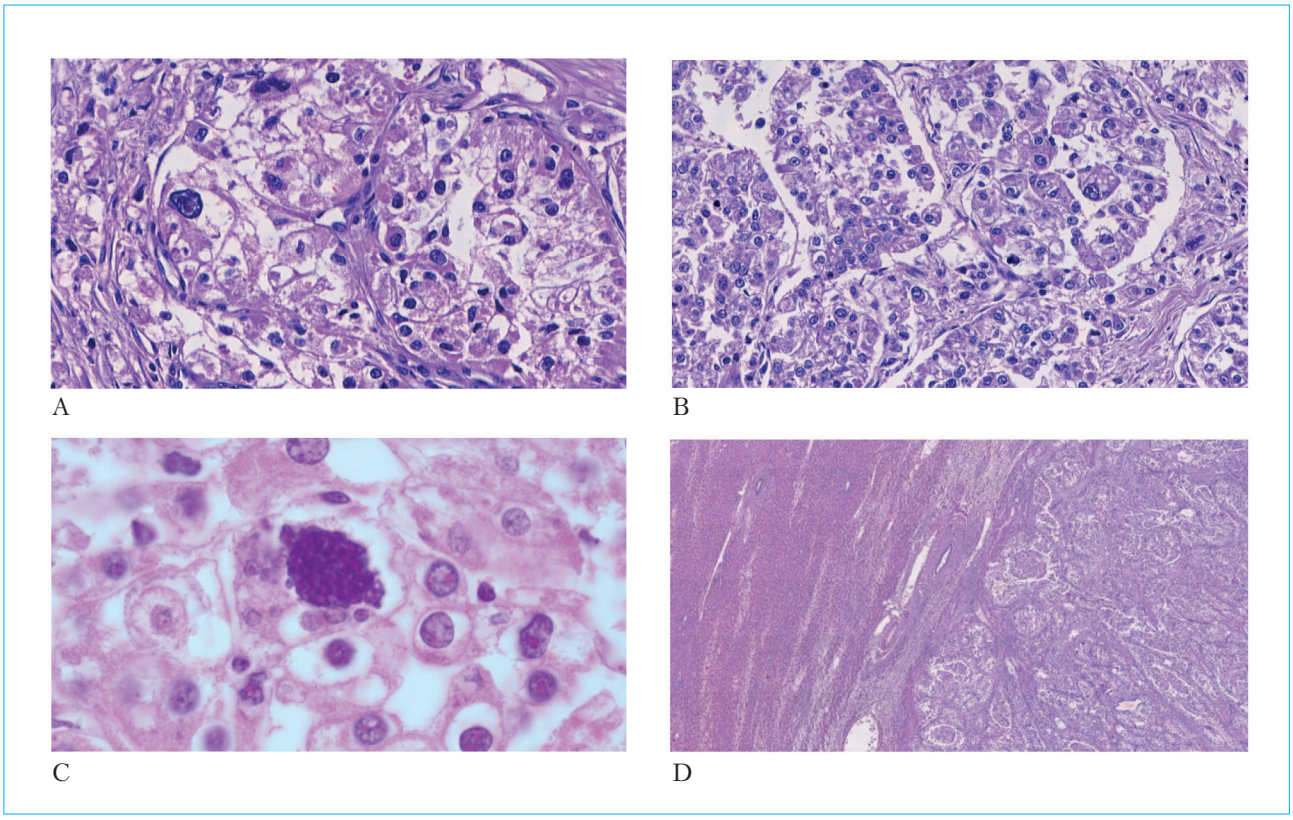


Fig. 4. Micropreparations, stained with hematoxylin and eosin: A — polymorphic tumor cells with granular eosinophilic cytoplasm, part with light cytoplasm and hyperchromic polymorphic nuclei, sometimes containing eosinophilic nucleoli, tumor cells form alveolar structures (magnification 630); B — the tumor is built from epithelioid cells with abundant eosinophilic, sometimes granular cytoplasm, partly with light nuclei, forming alveolar structures (magnification 430); C — atypical mitosis and nuclei containing eosinophilic nucleoli (magnification 1000); D — tumor node with a smooth edge without signs of the presence of a pseudocapsule (magnification 50)

Рис. 4. Микропрепараты, окраска гематоксилин и эозин: А — клетки опухоли полиморфные с гранулярной эозинофильной цитоплазмой, частью со светлой цитоплазмой и гиперхромными полиморфными ядрами, местами содержащими эозинофильные ядрышки, клетки опухоли формируют альвеолярные структуры (увеличение 630); В — опухоль построена из эпителиоидных клеток с обильной эозинофильной, местами гранулярной цитоплазмой, частью светлыми ядрами, формирующих альвеолярные структуры (увеличение 430); С — атипичный митоз и ядра, содержащие эозинофильные ядрышки (увеличение 1000); D — узел опухоли с ровным краем без признаков наличия псевдокапсулы (увеличение 50)

publication of C. Geng et al. two more similar cases are also presented in the review part [46], however, we could not confirm these sources with publications. More recent publications are literature reviews summarizing data on PEComas localized in the pancreas [14, 39, 44]. Table 1 presents a summary of all 32 clinical cases, as well as our own case.

Pancreatic PEComas can occur in patients of any age (ages 17 to 74 years, mean age — 48.3 years). Mostly they are detected in women (78.8 %). In almost half of the cases (54.5 %), patients complained of abdominal pain. No complaints were noted in 18.1 % of patients. The size of the tumors varied from 0.38 to 11.5 cm. It should be noted that tumor sizes of 0.38 cm were found in a tumor localized in the heterotopic pancreas [37]. Most often, the tumor was localized in the pancreatic head (16 patients; 48.5 %). The combination with tuberous sclerosis was revealed in one case (data were not available in 11 cases). Most pancreatic PEComas are benign (90.6 %). Malignant PEComas were identified in three cases. In two of them, liver metastases were detected after 6 and 8 months [25, 30]. In the third (this is our own case), metastases to the lymph nodes and liver were detected synchronously with the primary tumor. Necrosis in the tumor tissue, as one of the signs of malignancy, was detected in 4 cases. In two of them, with a follow-up period of 7 to 18 months, no tumor recurrence and/or distant metastases were detected (50.0 %). In one of them, liver metastases were detected in the long-term period. In our own case, in the structure of

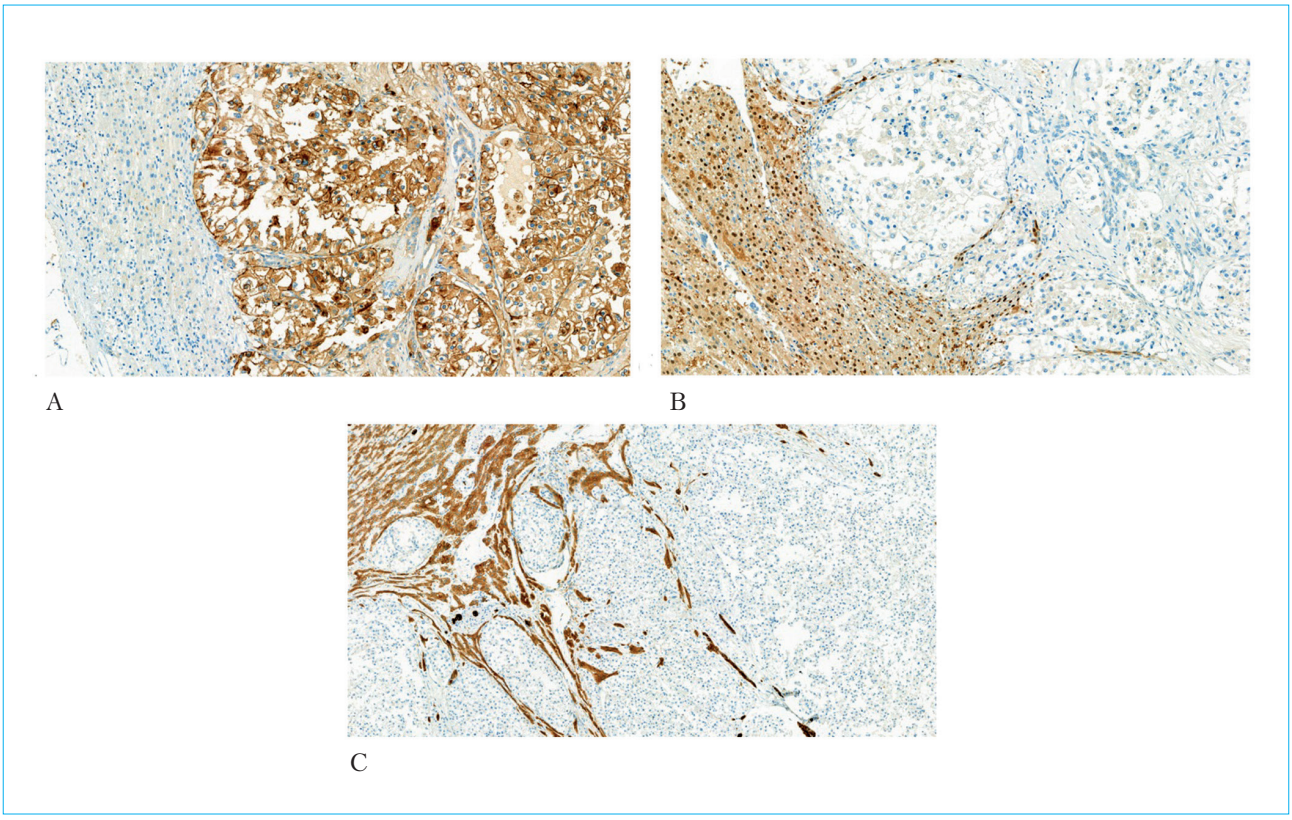


Fig. 5. Micropreparations, DAB and hematoxylin: A — expression of the HMB-45 melanosome marker in tumor cells (magnification 200); B — expression of the marker of hepatocellular differentiation Arginase-1 in the liver tissue surrounding the tumor (magnification 200); C — expression of total cytokeratin in hepatocytes and ducts of the liver tissue indicates a wide front of tumor invasion (magnification 100)

Рис. 5. Микропрепараты, DAB и гематоксилин: А — экспрессия маркера меланосом HMB-45 в клетках опухоли (увеличение 200); В — экспрессия маркера гепатоцеллюлярной дифференцировки Arginase-1 в окружающей опухоль ткани печени (увеличение 200); С — экспрессия общего цитокератина в гепатоцитах и протоках ткани печени указывает на широкий фронт инвазии опухоли (увеличение 100)

synchronously detected liver metastases, there were foci of necrosis, occupying up to 50 % of the tumor area in the section. The number of mitoses, including pathological ones, in our case was 5 per 10 representative fields of view at magnification 400. The data presented demonstrate that we have encountered the most malignant form of this tumor.

The macroscopically solid component of PEComa has a grayish-white color and a fragile texture, cystic inclusions contain a small amount of bloody fluid. The tumor had a clear border with the surrounding tissues of the pancreas [46].

Ultrasound is most often mentioned as a non-specific research method at PEComas diagnosis. However, summarizing the literature data and our own experience, we can say that these tumors are usually visualized as an echo-dense lesion, clearly delimited from the parenchyma of the organ, most often with attenuation of the echo signal to the periphery. Smaller lesions have a more homogeneous structure, in large ones there may be areas of reduced echo density in the center and/or cystic inclusions. Duplex scanning registers blood flow with an arterial spectrum in the tumor structure [48, 49].

With MSCT, PEComa is defined as a heterogeneous solid hypodense lesion with an X-ray density of less than –20 HU (–20...–120 HU). With the introduction of a contrast agent, the lesion is hypervascular, the accumulation of the contrast agent occurs in the arterial phase of the study with its discharge to the delayed phase, with a hypervascular capsule [40, 46].

Despite the fact that MRI is a method specific for the differentiation of adipose tissue, in the case of PEComa, verification is not always obvious. This situation is due to the different content of adipose tissue in different types of angiolipomas (for example, with a myomatous (monomorphic) type, no more than 10 %). On MR imaging, angiomyolipomas appear hyperintense on T1-weighted images (WI) and hypointense on T1-WI with fat

Table 1. Summary data on pancreatic PEComas presented in the literature to date [9, 14, 19–47]

Таблица 1. Сводные данные о ПЭКомах поджелудочной железы, представленные в литературе к настоящему времени [9, 14, 19–47]

№	Authors Авторы	Year Год	Cases Наблюдения	Age Возраст	Gender m/f Пол м/ж	Symptoms Симптомы	Localization Локализация	Dimensions, cm Размеры, см	TS ТС	B/M Д/З	Necrosis Некроз	Observation period Срок наблюдения	Process progression Прогрессирование процесса
1	Zamboni G., et al. [9]	1996	1	60	f ж	abdominal pain боль в животе	body тело	2	–	B Д	ND ОД	3	–
2	Heywood G., et al. [19]	2004	1	74	f ж	abdominal pain боль в животе	uncinate process крючковидный отросток	4.7	–	B Д	–	69	–
3	Ramuz O., et al. [20]	2005	1	31	f ж	abdominal pain боль в животе	body тело	1.5	–	B Д	–	9	–
4	Perigny M., et al. [21]	2008	1	46	f ж	diarrhea диарея	body тело	1.7	ND ОД	B Д	–	3	–
5	Hirabayashi K., et al. [22]	2009	1	47	f ж	abdominal pain боль в животе	head головка	1.7	–	B Д	–	12	–
6	Baez J.C., et al. [23]	2009	1	60	f ж	deformity of the abdomen in the form of a bulge in the epigastrium деформация живота в виде выпуклости в эпигастрии	body тело	3.5	–	B Д	–	7	–
7	Zemet R., et al. [24]	2011	1	49	m м	fever, cough, malaise лихорадка, кашель, недомогание	head головка	4	–	B Д	–	10	–
8	Nagata S., et al. [25]	2011	1	52	m м	abdominal pain боль в животе	head головка	4	–	M З	ND ОД	37	mts in the liver метастазы в печень
9	Xie F.L., et al. [26]	2011	1	58	m м	abdominal pain, dyspepsia боль в животе, диспепсия	head головка	2.2	ND ОД	B Д	ND ОД	5	–
10	Finzi G., et al. [27]	2012	1	62	f ж	none отсутствуют	head головка	2.5	–	B Д	ND ОД	5	–
11	Al-Haddad M., et al. [28]	2013	1	38	f ж	abdominal pain боль в животе	uncinate process крючковидный отросток	1.8	ND ОД	B Д	ND ОД	ND ОД	–
12	Okuwaki K., et al. [29]	2013	1	43	f ж	abdominal pain боль в животе	body and tail тело и хвост	10	–	B Д	+	7	–
13	Mourra N., et al. [30]	2013	1	51	f ж	abdominal pain, jaundice боль в животе, желтуха	head головка	6	–	M З	+	8	mts in the liver метастазы в печень
14	Tummala P., et al. [31]	2013	1	ОД ND	ОД ND	ND ОД	head and isthmus головка и перешеек	ND ОД	ND ОД	B Д	ND ОД	ND ОД	ND ОД
15	Kim J.Y., et al. [32]	2014	1	31	f ж	none отсутствуют	tail хвост	3.3	ND ОД	B Д	ND ОД	63	–

16	Petrides C., et al. [33]	2015	1	17	f ж	melena, anemia мелена, анемия	head головка	4.2	ND ОД	B Д	ND ОД	18	–
17	Mizuuchi Y., et al. [34]	2015	1	61	f ж	abdominal pain боль в животе	body тело	7	–	B Д	–	144	–
18	Wei J., et al. [35]	2016	1	58	f ж	none отсутствуют	body тело	2	ND ОД	B Д	–	18	–
19	Jiang H., et al. [36]	2016	1	50	f ж	none отсутствуют	head and uncinate process головка и крючковидный отросток	2	–	B Д	ND ОД	14	–
20	Kiriyama Y., et al. [37]	2016	1	57	m м	lower back pain боль в пояснице	heterotopic гетеротопическая	0.38	ND ОД	B Д	ND ОД	56	–
21	Hartley C.P., et al. [38]	2016	1	31	f ж	abdominal pain боль в животе	tail хвост	6.5	+	B Д	–	ND ОД	–
22	Collins K., et al. [39]	2017	1	54	f ж	abdominal pain боль в животе	head and body головка и тело	2.6	ND ОД	B Д	–	27	–
23	Zizzo M., et al. [40]	2017	1	68	m м	pain in the upper abdomen боль в верхней половине живота	head головка	2.8	–	B Д	–	13	–
24	Zhang S., et al. [14]	2017	1	43	f ж	abdominal pain боль в животе	head головка	11.5	–	B Д	+	18	–
25	Sangiorgio V., et al. [41]	2019	2	47	f ж	ND ОД	isthmus перешеек	30	–	B Д	ND ОД	ND ОД	ND ОД
26				70	f ж	ND ОД	body тело	10	–	B Д	ND ОД	ND ОД	ND ОД
27	Gondran H., et al. [42]	2019	1	17	m м	weight loss, asthenia, flu-like syndrome потеря веса, астения, гриппоподобный синдром	head головка	5.5	–	B Д	–	42	–
28	Uno K., et al. [43]	2019	1	49	f ж	none отсутствуют	tail хвост	4	–	B Д	–	12	–
29	Colón Rodríguez A., et al. [44]	2020	1	50	f ж	abdominal pain, anemia боль в животе, анемия	body тело	1.5	–	B Д	–	18	–
30	Ulrich J.D., et al. [45]	2020	1	49	f ж	pain in the upper abdomen боль в верхней половине живота	body тело	2.5	ND ОД	B Д	ND ОД	ND ОД	–
31	Geng C., et al. [46]	2021	1	40	f ж	none отсутствуют	body and tail тело и хвост	2.3	ND ОД	B Д	ND ОД	30	–
32	Wang X., Xue H. [47]	2022	1	62	m м	abdominal pain боль в животе	head головка	5	–	B Д	–	ND ОД	–
33	Own case Собственное наблюдение	2022	1	22	f ж	weakness, anemia слабость, анемия	head головка	5.4	–	*M + mts in the liver and lymph nodes	+	69	–

Note: ND — no data; TS — tuberous sclerosis; B — benign; M — malignant; mts — metastases; * — simultaneous detection of malignant pancreatic PEComa and liver metastases.
Примечание: ОД — отсутствие данных; ТС — туберозный склероз; Д — доброкачественное; З — злокачественное; * — одновременное выявление злокачественной ПЭКомы ПЖ и метастазов печени.

suppression, and moderately hyperintense on T2-WI. The dynamics of contrast agent accumulation in MR examination is similar to that in CT [40, 50].

As for the pancreatic PEComas metastases, then, unfortunately, their radiology pattern is nonspecific, similar signs will also be with metastases of a neuroendocrine tumor and renal cell carcinoma. Most of the clinical cases presented in the literature focus on the morphological, immunohistochemical, and genetic aspects of pancreatic PEComas. Considering the rarity of occurrence of malignant pancreatic PEComas, we did not find a separate focus on metastases and their signs. In this regard, we consider it appropriate in the presented clinical observation to provide a description and illustrations of the ultrasound and magnetic resonance characteristics of these metastases. It should be noted that PEComa metastases contain an extremely small amount of fat and the T2-WI sequence in the fat suppression mode in this situation is not effective for differential diagnosis.

Surgical treatment remains the main method of treating patients with PEComas of various localizations. Unfortunately, chemotherapy and radiotherapy have not shown any significant results in either adults or children. Also, surgical treatment remains the main method, both in the treatment of relapses and metastases [51–54]. The rarity of these tumors makes it difficult to study the use of new drugs and their regimens in patients with PEComas of various localizations. The role of neoadjuvant therapy is still unclear due to the small number of cases. Attempts have been made to use mTOR inhibitors in the treatment of PEComas. Thus, H. Gondran et al. in 2019 described a case of pancreatic head PEComa in a 17-year-old man who complained of weight loss, asthenia, and influenza-like syndrome. The lesion had a size of 55 mm and did not compress the bile and pancreatic ducts. In this case, the patient usually performs PDR. But the authors decided that it would be too mutilating for a relatively benign tumor (a biopsy was abandoned because the result of a single biopsy may be questionable). The patient was treated with Sirolimus (an mTOR inhibitor) in tablet form for 42 months, monitoring the condition according to the data of radiology methods and adjusting the dose and frequency of administration. After 42 months, MRI showed a decrease in the size of the tumor and stabilization of the condition. The authors plan to perform RFA of the tumor under ultrasound control in the future [42]. In the clinical case presented above, a patient with liver metastases received Everolimus therapy, which allowed her to somewhat stabilize her condition for 6 years.

It should be noted that, in general, PEComas have a low malignant potential. Thus, a clinical case of a 49-year-old patient with retroperitoneal localization of a 7.0 cm PEComa, who has been alive for 4 years after tumor removal in the presence of metastases in the brain and lungs, has been described [55].

Conclusion

Perivascular epithelioid cell tumor, also known as PEComa, is a family of mesenchymal tumors composed of perivascular epithelioid cells (PECs). These are rare tumors that can occur anywhere in the human body. PEComa is a heterogeneous and usually benign tumor with a favorable prognosis. Malignant neoplasms are rare and often present with local recurrences and distant metastases.

In the literature, there are reports of only 32 cases of pancreatic PEComas. Our own clinical case demonstrates that we have encountered the most malignant form of this tumor. Further studies on large series are needed to better understand the biological features of PEComa, to be able to develop recommendations for diagnosis, treatment and follow-up, which today seems extremely difficult due to the rarity of this tumor.

Литература / References

1. *Fletcher C.D., Unni K.K., Mertens F.* World health organization classification of tumors: Pathology and genetics of tumors of soft tissue and bone. *Lyon: IARC Press*, 2002.
2. *Martignoni G., Pea M., Reghellin D., Zamboni G., Bonetti F.* PEComas: The past, the present and the future. *Virchows Arch.* 2008;452(2):119–32. DOI: 10.1007/s00428-007-0509-1
3. *Fang S.H., Zhou L.N., Jin M., Hu J.B.* Perivascular epithelioid cell tumor of the liver: A report of two cases and review of the literature. *World J Gastroenterol.* 2007;13(41):5537–9. DOI: 10.3748/wjg.v13.i41.5537
4. *Conlon N., Soslow R.A., Murali R.* Perivascular epithelioid tumours (PEComas) of the gynaecological tract. *J Clin Pathol.* 2015;68(6):418–26. DOI: 10.1136/jclinpath-2015-202945
5. *Folpe A.L., Kwiatkowski D.J.* Perivascular epitheloid cell neoplasms: Pathology and pathogenesis. *Human Pathology.* 2009;41(1):1–15. DOI: 10.1016/j.humpath.2009.05.011
6. *Crino P.B., Nathanson K.L., Henske E.P.* The tuberous sclerosis complex. *N Engl J Med.* 2006;355(13):1345–56. DOI: 10.1056/NEJMra055323
7. *Bennett J.A., Braga A.C., Pinto A., Van de Vijver K., Cornejo K., Pesci A., et al.* Uterine PEComas: A morphologic, immunohistochemical, and molecular analysis of 32 tumors. *Am J Surg Pathol.* 2018;42(10):1370–83. DOI: 10.1097/PAS.0000000000001119
8. *Bonetti F., Pea M., Martignoni G., Zamboni G.* PEC and sugar. *Am J Surg Pathol.* 1992;16(3):307–8. DOI: 10.1097/00000478-199203000-00013

9. *Zamboni G., Pea M., Martignoni G., Zancanaro C., Faccioli G., Gilioli E., et al.* Clear cell "sugar" tumor of the pancreas. A novel member of the family of lesions characterized by the presence of perivascular epithelioid cells. *Am J Surg Pathol*. 1996;20(6):722–30. DOI: 10.1097/00000478-199606000-00010
10. *Bonetti F., Pea M., Martignoni G., Doglioni C., Zamboni G., Capelli P., et al.* Clear cell ("sugar") tumor of the lung is a lesion strictly related to angiomyolipoma — the concept of a family of lesions characterized by the presence of the perivascular epithelioid cells (PEC). *Pathology*. 1994;26(3):230–6. DOI: 10.1080/00313029400169561
11. *Fernandez-Flores A.* Evidence on the neural crest origin of PEComas. *Rom J Morphol Embryol*. 2011;52(1):7–13.
12. *Armah H.B., Parwani A.V.* Perivascular epithelioid cell tumor. *Arch Pathol Lab Med*. 2009;133(4):648–54. DOI: 10.5858/133.4.648
13. *Давыденко П.И., Кармазановский Г.Г., Широков В.С., Паклина О.В., Вишневский В.А.* Опухоли из периваскулярных эпителиоидных клеток: проблемы диагностики. *Хирургия. Журнал им. Н.И. Пирогова.* 2013;7:4–12. [*Davydenko P.I., Karmazanovsky G.G., Shirokov V.S., Paklina O.V., Vishnevsky V.A.* Tumors from perivascular epithelioid cells: Diagnostic problems. Pirogov Russian *Journal of Surgery = Khirurgiya. Zurnal im. N.I. Pirogova.* 2013;7:4–12 (In Russ.)].
14. *Zhang S., Chen F., Huang X., Jiang Q., Zhao Y., Chen Y., et al.* Perivascular epithelial cell tumor (PEComa) of the pancreas: A case report and review of literature. *Medicine (Baltimore)*. 2017;96(22):e7050. DOI: 10.1097/MD.0000000000007050
15. *Нечушкина И.В.* Пекомы. *Онкопедиатрия*. 2016;3(4):267–76. [*Nechushkina I.V.* Pekoma. *Onkopediatria*. 2016;3(4):267–76 (In Russ.)]. DOI: 10.15690/onco.v3i4.1630
16. *Bleeker J.S., Quevedo J.F., Folpe A.L.* "Malignant" perivascular epithelioid cell neoplasm: Risk stratification and treatment strategies. *Sarcoma*. 2012;2012;541626. DOI: 10.1155/2012/541626
17. *Bleeker J.S., Quevedo J.F., Folpe A.L.* Malignant perivascular epithelioid cell tumor of the uterus. *Rare Tumors*. 2012;4(1):e14. DOI: 10.4081/rt.2012.e14
18. *Schoolmeester J.K., Howitt B.E., Hirsch M.S., Dal Cin P., Quade B.J., Nucci M.R.* Perivascular epithelioid cell neoplasm (PEComa) of the gynecologic tract: Clinicopathologic and immunohistochemical characterization of 16 cases. *Am J Surg Pathol*. 2014;38(2):176–88. DOI: 10.1097/PAS.0000000000000133
19. *Heywood G., Smyrk T.C., Donohue J.H.* Primary angiomyolipoma of the pancreas. *Pancreas*. 2004;28(4):443–5. DOI: 10.1097/00006676-200405000-00014
20. *Ramuz O., Lelong B., Giovannini M., Delpero J.R., Rochaix P., Xerri L., et al.* "Sugar" tumor of the pancreas: A rare entity that is diagnosable on preoperative fine-needle biopsies. *Virchows Arch*. 2005;446(5):555–9. DOI: 10.1007/s00428-005-1216-4
21. *Périgny M., Larochelle O., Hammel P., Sauvanet A., Dokmak S., Belghiti J., et al.* Pancreatic perivascular epithelioid cell tumor (PEComa). *Ann Pathol*. 2008;28(2):138–42 (In French). DOI: 10.1016/j.annpat.2007.11.005
22. *Hirabayashi K., Nakamura N., Kajiwara H., Hori S., Kawaguchi Y., Yamashita T., et al.* Perivascular epithelioid cell tumor (PEComa) of the pancreas: Immunoelectron microscopy and review of the literature. *Pathol Int*. 2009;59(9):650–5. DOI: 10.1111/j.1440-1827.2009.02421.x
23. *Baez J.C., Landry J.M., Saltzman J.R., Qian X., Zinner M.J., Mortelé K.J.* Pancreatic PEComa (sugar tumor): MDCT and EUS features. *JOP*. 2009;10(6):679–82.
24. *Zemet R., Mazeh H., Neuman T., Freund H.R., Eid A.* Asymptomatic pancreatic perivascular epithelial cell tumor (PEComa) in a male patient: Report and literature review. *JOP*. 2011;12(1):55–8.
25. *Nagata S., Yuki M., Tomoeda M., Kubo C., Yoshizawa H., Kitamura M., et al.* Perivascular epithelioid cell neoplasm (PEComa) originating from the pancreas and metastasizing to the liver. *Pancreas*. 2011;40(7):1155–7. DOI: 10.1097/MPA.0b013e318221fc0e
26. *Xie F.L., Song Y.N., Qu L.J., Yu Y.H., Zheng Z.Y.* Clinicopathologic features of perivascular epithelioid cell tumor of the pancreas. *Shijie Huaren Xiaohua Zazhi*. 2011;19:964e8.
27. *Finzi G., Micello D., Wizemann G., Sessa F., Capella C.* Pancreatic PEComa: A case report with ultrastructural localization of HMB-45 within melanosomes. *Ultrastruct Pathol*. 2012;36(2):124–9. DOI: 10.3109/01913123.2011.642463
28. *Al-Haddad M., Cramer H.M., Muram T., Wang X., Pitt H.A.* Perivascular epithelioid cell tumor: An unusual pancreatic mass diagnosed by EUS-FNA. *Gastrointest Endosc*. 2013;78(1):165. DOI: 10.1016/j.gie.2013.02.011
29. *Okuwaki K., Kida M., Masutani H., Yamauchi H., Katagiri H., Mikami T., et al.* A resected perivascular epithelioid cell tumor (PEComa) of the pancreas diagnosed using endoscopic ultrasound-guided fine-needle aspiration. *Intern Med*. 2013;52(18):2061–6. DOI: 10.2169/internalmedicine.52.0746
30. *Mourra N., Lazure T., Colas C., Arrive L., de Gramont A.* Perivascular epithelioid cell tumor: The first malignant case report in the pancreas. *Appl Immunohistochem Mol Morphol*. 2013;21(3):e1–4. DOI: 10.1097/PAI.0b013e3182392bb6
31. *Tummala P., Rao S., Agarwal B.* Differential diagnosis of focal non-cystic pancreatic lesions with and without proximal dilation of pancreatic duct noted on CT scan. *Clin Transl Gastroenterol*. 2013;4(11):e42. DOI: 10.1038/ctg.2013.15
32. *Kim J.Y., Song J.S., Park H., Byun J.H., Song K.B., Kim K.P., et al.* Primary mesenchymal tumors of the pancreas: Single-center experience over 16 years. *Pancreas*. 2014;43(6):959–68. DOI: 10.1097/MPA.0000000000000130
33. *Petrides C., Neofytou K., Khan A.Z.* Pancreatic perivascular epithelioid cell tumour presenting with upper gastrointestinal bleeding. *Case Rep Oncol Med*. 2015;2015:431215. DOI: 10.1155/2015/431215
34. *Mizuuchi Y., Nishihara K., Hayashi A., Tamiya S., Toyoshima S., Oda Y., et al.* Perivascular epithelial cell tumor (PEComa) of the pancreas: A case report and review of previous literatures. *Surg Case Rep*. 2016;2(1):59. DOI: 10.1186/s40792-016-0186-x
35. *Wei J., Liu X., Fu Y., Gao W., Jiang K., Zhang Z., et al.* Primary perivascular epithelioid cell tumor of the pancreas: A case report. *Pancreatology*. 2016;16:S30. DOI: 10.1016/J.PAN.2015.12.094
36. *Jiang H., Ta N., Huang X.Y., Zhang M.H., Xu J.J., Zheng K.L., et al.* Pancreatic perivascular epithelioid cell tumor: A case report with clinicopathological features and a literature review. *World J Gastroenterol*. 2016;22(13):3693–700. DOI: 10.3748/wjg.v22.i13.3693
37. *Kiriyama Y., Tsukamoto T., Mizoguchi Y., Ishihara S., Horiguchi A., Tokoro T., et al.* Intrahepatic peribiliary perivascular epithelioid cell tumor (PEComa) associated with heterotopic pancreas: A case report. *Diagn Pathol*. 2016;11(1):81. DOI: 10.1186/s13000-016-0528-9
38. *Hartley C.P., Kwiatkowski D.J., Hamieh L., Lefferts J.A., Smith K.D., Lisovsky M.* Pancreatic PEComa is a novel member of the family of tuberous sclerosis complex-associated tumors: Case report and review of the literature. *Virchows Arch*. 2016;469(6):707–10. DOI: 10.1007/s00428-016-2032-8
39. *Collins K., Buckley T., Anderson K., Karasik M., Ligato S.* Perivascular epithelioid cell tumor (PEComa) of pancreas diagnosed preoperatively by endoscopic ultrasound-guided fine-needle aspiration: A case report and review of literature. *Diagn Cytopathol*. 2017;45(1):59–65. DOI: 10.1002/dc.23599
40. *Zizzo M., Castro Ruiz C., Ugoletti L., Bonacini S., Giunta A., Valli R., et al.* Primary pancreatic PEComa: Is surgery always the best choice? *JOP*. 2017;18(1):241–3.
41. *Sangiorgio V., Daniele L., Gallo T., Rocca R., Fava C., Campisi P., et al.* Ultrasound-guided fine needle aspira-

tion cytology in the diagnosis of hepatic and pancreatic perivascular epithelioid cell tumors: A case series. *Diagn Cytopathol*. 2019;47(4):315–9. DOI: 10.1002/dc.24111
42. *Gondran H., Thebaud E., Moreau A., Le Rhun M., Touchefeu Y., Regenet N., et al.* First pancreatic perivascular epithelioid cell tumor (PEComa) treated by mTOR inhibitor. *Pancreatology*. 2019;19(4):566–8. DOI: 10.1016/j.pan.2019.05.459
43. *Uno K., Shimizu S., Hayashi K., Yamada T., Kusakabe A., Kanie H., et al.* Perivascular epithelial cell tumor of the pancreas diagnosed preoperatively by endoscopic ultrasound-guided fine-needle aspiration. *Intern Med*. 2019;58(17):2515–21. DOI: 10.2169/internalmedicine.2265-18
44. *Colón Rodríguez A., Calleja Kempin J., Quirós Higueras E., Ramón Botella E., Castillo J.* Pancreatic perivascular epithelial cell tumour (PECOma). Case report and literature review. *Gastroenterol Hepatol*. 2020;43(5):257–9. DOI: 10.1016/j.gastrohep.2019.11.012
45. *Ulrich J.D., Specht K., Schlitter A.M., Ceyhan G.O., Quante M., Schmid R.M., et al.* A rare case of perivascular epithelioid cell tumor (PEComa) of the pancreas diagnosed by endoscopic ultrasound. *Endosc Int Open*. 2020;8(1):E25–8. DOI: 10.1055/a-1038-3852
46. *Geng C., Cao Z., Bacacao B., Cao Z.* Perivascular epithelioid cell tumor of the pancreas: Case report and literature review. *Int J Clin Exp Pathol*. 2021;14(5):653–61.
47. *Wang X., Xue H., Chang X., Jin Z.* Gastrointestinal: Epithelioid angiomyolipoma of the pancreas. J *Gastroenterol Hepatol*. 2022;37(5):781. DOI: 10.1111/jgh.15739
48. *Jiang T.A., Zhao Q.Y., Chen M.Y., Wang L.J., Ao J.Y.* Diagnostic analysis of hepatic angiomyolipoma. *Hepatobiliary Pancreat Dis Int*. 2005;4(1):152–5.
49. *Степанова Ю.А., Кармазановский Г.Г., Щеголев А.И., Журенкова Т.В., Косова И.А., Паклина О.В.* и др. Ангиомиолипома (ПЕКома) печени: обзор литературы и клиническое наблюдение. *Медицинская визуализация*. 2011;6:30–7. [*Stepanova Yu.A., Karmazanovsky G.G., Shchegolev A.I., Zhurenkova T.V., Kosova I.A., Paklina O.V., et al.* Angiomyolipoma (PEComa) of the liver: A review of the literature and clinical case. *Medical Visualization*. 2011;6:30–7 (In Russ.)].
50. *Tan Y., Zhang H., Xiao E.H.* Perivascular epithelioid cell tumour: Dynamic CT, MRI and clinicopathological characteristics — analysis of 32 cases and review of the literature. *Clin Radiol*. 2013;68(6):555–61. DOI: 10.1016/j.crad.2012.10.021
51. *Zekry N., Rettenmaier M.A., Abaid L.N., John C.R., Micha J.P., Brown J.V.* 3rd, et al. Perivascular epithelioid cell neoplasms: A systematic review of prognostic factors. *J Minim Invasive Gynecol*. 2009;16(5):527–32. DOI: 10.1016/j.jmig.2009.04.017
52. *de León D.C., Pérez-Montiel D., Bandera A., Villegas C., Gonzalez-Conde E., Vilchis J.C.* Perivascular epithelioid cell tumor of abdominal origin. *Ann Diagn Pathol*. 2010;14(3):173–7. DOI: 10.1016/j.anndiagpath.2010.02.007
53. *Peng J., Yu B., Wang J., Hong X., Xu Y., Cai S.* An unresectable malignant perivascular epithelioid cell tumor resistant to multiple combined chemotherapies: A unique case report. *J Pediatr Hematol Oncol*. 2010;32(4):e136–8. DOI: 10.1097/MPH.0b013e3181b8700b
54. *Wu J.H., Zhou J.L., Cui Y., Jing Q.P., Shang L., Zhang J.Z.* Malignant perivascular epithelioid cell tumor of the retroperitoneum. *Int J Clin Exp Pathol*. 2013;6(10):2251–6.
55. *Weinreb I., Howarth D., Latta E., Ghazarian D., Chetty R.* Perivascular epithelioid cell neoplasms (PEComas): Four malignant cases expanding the histopathological spectrum and a description of a unique finding. *Virchows Arch*. 2007;450(4):463–70. DOI: 10.1007/s00428-007-0378-7

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