

The Clinical Case of Surgical Treatment of Giant Pancreatic Lymphangioma with the Involvement of the Portal Vein and Extrahepatic Bile Ducts in the Pathological Process

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Abstract

Lymphangioma of the abdominal cavity and retroperitoneal space is very rare surgical disease, which has a little description in the world literature. Lymphangiomas of the retroperitoneum occur in 1% of all cases of lymphangiomas of the human body. Untimely diagnosis and inadequate treatment of the disease leads to reduce the quality of life. **Objective.** To describe the rare clinical case of pancreatic lymphangioma. **Materials and methods.** The subject of the study is a clinical case of patients treated in Neftyanik hospital (Tyumen), where the therapy and surgical intervention was made. Laboratory tests were used: a general blood test, a general urine test, a biochemical blood test (glucose, total bilirubin, direct bilirubin, urea, creatinine, aspartate aminotransferase, alanine aminotransferase, amylase, protein), coagulogram, HIV test, a study of antibodies to hepatitis B, a study of antibodies to hepatitis C, a blood group, electrocardiography, examination of the therapist. Special diagnostic methods include the ultrasound of the abdominal cavity, multispiral computed tomography (MSCT) with intravenous contrasting of the abdominal organs, alpha-fetoprotein, oncomarkers, histological examination of the operating material. **Conclusion.** Previously unknown information was obtained about the possible location of lymphangioma in the head and body of the pancreas with the growth of the portal vein and the development of portal hypertension syndrome. Pancreatoduodenal resection with removal of the head and body of the pancreas with part of the affected portal vein is the optimal variant for surgical correction.

Keywords: cystic lymphangioma, mechanical jaundice, pancreas, pancreatoduodenal resection, portal hypertension syndrome.

INTRODUCTION

Lymphangioma of the pancreas refers to rare human tumors which dramatically reduces the duration and quality of the patients' life without adequate treatment [1-3]. There are single descriptions of this pathology in the world literature [4, 5]. More often, this tumor is found in the neck and mediastinum. Cases of localization in the mesentery of the small intestine in the abdominal cavity have been described by some authors [6-8]. Lymphangiomas of the retroperitoneal space in frequency constitute 1% of all cases of lymphangiomas of the human body [9-11]. The purpose of this article is to inform about the rare clinical case of intraorganic lymphangioma of the pancreas.

MATERIALS AND METHODS

The subject of the study is a clinical case of patients treated in Neftyanik hospital (Tyumen), where the therapy and surgical intervention was made. Laboratory tests were used: a general blood test, a general urine test, a biochemical blood test (glucose, total bilirubin, direct bilirubin, urea, creatinine, aspartate aminotransferase, alanine aminotransferase, amylase, protein), coagulogram, HIV test, a study of antibodies to hepatitis B, a study of antibodies to hepatitis C, a blood group, electrocardiography, examination of the therapist. Special diagnostic methods include the ultrasound of the abdominal cavity, multispiral computed tomography (MSCT) with intravenous contrasting of the abdominal organs, alpha-fetoprotein, oncomarkers, histological examination of the operating material.

This patient gave written voluntary consent to participate in the clinical trial, which was performed in accordance with the Declaration of Helsinki of the World Medical Association.

RESULTS

A 47 years old patient (born in 1969) entered the Neftyanik hospital with complaints of tumor-like formation of the abdominal cavity, icterus of the skin. She's going to get surgical treatment.

From the anamnesis it is known that an examination was performed 2 years ago (2015) when there was pain in epigastrium and discomfort during physical activity: ultrasound of the

abdominal cavity and the subsequent MSCT of the abdominal cavity with intravenous contrast during the venous phase, 1.5 mm step - a volume formation 7.93 x 6.82 cm with multiple septa and calcium inclusions was revealed in the head and the body of the pancreas (Fig. 1). The formation was classified as a pseudotumorous cyst of the pancreas, clinical group 1B. Surgical treatment was not required because of the absence of complications.

In August 2016 the patient's condition worsened because of increased pain syndrome and the development of mechanical jaundice. The level of direct bilirubin in blood was 260 $\mu\text{mol/l}$.

Endoscopic retrograde cholangiopancreatography (ERCP) was performed. The intra-pancreatic part of the common bile duct was deformed and narrowed over 40 mm long with a supragenetic dilatation up to 20 mm and an expansion of the intrahepatic ducts to 8 mm. Stenting of choledoch was performed.

During the physical examination, 2 weeks later: soft-elastic, painless, not mobile formation up to 10 cm in diameter was palpated in the epigastrium and in the right hypochondrium. The main laboratory indicators were without pathological changes. Normalization of bilirubinemia was noted. The level of alpha-fetoprotein - 4.2 Units; CEA - 2.6 Units; MUC16 - 19.9 - (21.9 Units) indicates that the indicators of oncomarkers were not increased.

In control MRI with intravenous contrast in T2 mode (October 2017), was observed that the size of the formation increases up to 10.2x5.9x7.2 cm (Figs. 2, 3). The tumor completely replaced the head, isthmus and most of the body of the pancreas with compression of the intrapancreatic part of the common bile duct (previously stented), the superior mesenteric and portal veins. The formation did not accumulate the contrast.

On the basis of the above data, the diagnosis is Giant cystic pancreatic tumor complicated by mechanical icterus (choledoch stent in August 2016) with growing into the portal vein.

The course of this disease with complications was an indication for the operation.

November 21, 2016, under endotracheal anesthesia a Rio Branco laparotomy was performed by MD Zaitsev E.Y.

Intraoperative: during revision, the formation 15x10 cm was found in the projection of the pancreas, little mobile; the entire gland head was replaced by a tumor that extends to the isthmus and body of the gland. There were no metastatic foci in the liver and other organs. A biopsy of the formation was taken and an urgent histological examination of the tumor showed no signs of malignant growth.

After mobilizing of the pancreatoduodenal complex, it was found out that tunneling between the isthmus of the gland and the portal vein is impossible because of the tumor growing into the portal and upper mesenteric veins. We decided to perform pancreatoduodenal resection with the resection of the affected veins. After cholecystectomy, the elements of the hepatoduodenal ligament were isolated and taken on the holders: choledoch (up to 1.2 cm in diameter), portal vein, common hepatic artery. Choledoch was cut immediately above the place of the connection with the cystic duct, the stent was extracted. The duodenum was crossed 2 cm below the pylorus by the GIA 50 Premium. The pancreas was crossed by a scalpel in the area of the body left to the superior mesenteric vein, laterally 1 cm from the tumor. Pancreatic duct was not enlarged (0.2 cm), gland was "juicy". The jejunum was crossed 10 cm below the Tretyc ligament, was mobilized and held to the right of the superior mesenteric vessels together with the ascending part of the duodenum. The splenic vein was ligated. The portal vein was crossed above the gland and the superior mesenteric vein was also crossed. The pancreatoduodenal complex was removed. An end-to-end venovenous anastomosis was performed. The pancreatojejunal anastomosis was performed with a continuous suture PDS 4-0, then a single-row continuous suture was applied to the end-to-side hepaticojejunal anastomosis. Then, an anterior-transversal duodenojejunal anastomosis was formed end-to-side. The abdominal cavity was closed.

Accumulation of 500 ml non-drained fluid in the area of the gland bag was noted In the postoperative period - treatment by a puncture under the ultrasound navigation was made. There were no other complications. The blood flow through the portal vein is satisfactory (controlled by ultrasound). The patient in a satisfactory condition was discharged on the 32nd day after hospitalization. The dynamic observation during 9 months: the patient's condition is stable, no complications were detected, no recurrence of the disease.

In morphological research: the macro preparation is a multi-compartment cystic formation, constructed from different sizes and forms closely located vascular cavities, which were separated by fibrous tissue in the head and body of the pancreas (Fig. 4).

During the microscopic examination, as shown in Figs. 5-8: the inner surface of the cystic cavities was lined with flattened endothelial cells. A large number of collagen fibers were determined in the cyst walls, single islets of Langerhans were found in some places. There were focal lymphoplasmic infiltrates with an admixture of hemosiderophages and local calcifications. There were the chylous and hemorrhagic contents in the lumen of the cavities; There were wall clots with signs of organization in the part of the cavities. There was stromal sclerosis with focal lymphoid infiltration in the head of the pancreas on the border with the tumor. There were islets of Langerhans as well as tubular structures of various sizes, with no signs of atypia, in some places. There was a positive reaction determined with antibodies CD 31 and ERG in immunohistochemical studies in endothelial lining cells. The morphological picture corresponds to the cavernous lymphangioma of the pancreas.

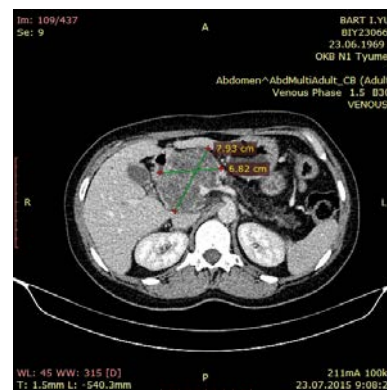


Figure 1 – MSCT with intravenous contrast during the venous phase with 1.5 mm step - volume formation of the pancreas 7.93x6.82 cm with multiple septa and calcium inclusions are visualized



Figure 2 – Reconstruction of MRI with intravenous contrast in T2 mode - a polycystic formation 10.2 x 5.9 cm



Figure 3 – Reconstruction of MRI with intravenous contrast in T1 mode - a polycystic formation 7.25x5.32 cm is visualized



Figure 4 – Macropreparation of the head and body tumor of the pancreas

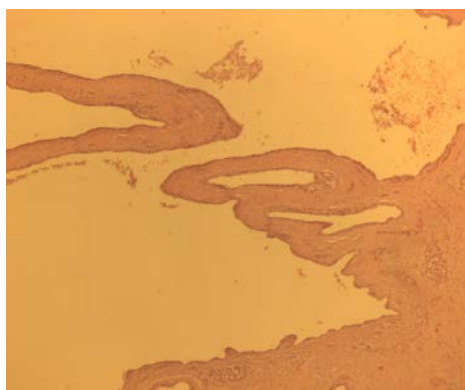


Figure 5 – Micropreparation of lymphangioma colored by Hematoxylin-Eosin under 50-fold increase – there are islets of Langerhans determined in the stroma of the cyst



Figure 6 – Micropreparation of lymphangioma colored by Hematoxylin-Eosin under 100-fold increase

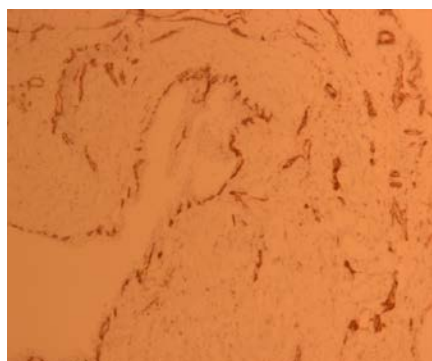


Figure 7 – Micropreparation of lymphangioma. The immunohistochemical study with CD31 expression, 100-fold increase - the lining of the cysts by the endothelium is determined

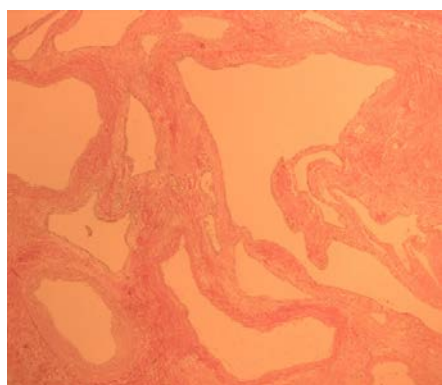


Figure 8 – Micropreparation of lymphangioma colored according to Van-Gieson - defining the connective tissue in the preparation

DISCUSSION

We have treated a patient with a rare form of pancreatic neoplasm - lymphangioma. In publications devoted to rare cases of localization of lymphangioma in the pancreas, it has been established, that the formation has an external localization rather than penetrating deeply into tissue in the relation to the gland [12-14]. In certain cases of operative treatment lymphangioma had a definite border separating the formation from healthy pancreatic tissues and in this cases, the disease usually proceeds asymptotically and does not lead to damage of the ductal system and surrounding organs, including the vessels. Surgical treatment with the external location of the tumor is not accompanied by technical difficulties and it was performed laparoscopically in all known cases [15, 16].

In this specific clinical case, the patient's lymphangioma developed from lymphatic structures which lay deep in the gland tissues. Its growth was rather infiltrative, with the growing into all tissues of the pancreas head. This localization of the formation ultimately led to compression of the common bile duct throughout its pancreatic part with the development of mechanical jaundice. The venous structures located near the head of the pancreas, including the portal vein, also underwent compression.

Despite the unequivocally benign nature of this cystic formation and its localization in the head of the pancreas can carry a threat to the life and health of the patient. The large-scale surgical interventions like pancreatoduodenal resection are required to overcome this situation. Involvement of large venous structures in the process (primarily the portal vein), makes possible to perform a resection of the pancreas head only with a part of the affected vein and the creation of portal venous anastomosis. The benign nature of the tumor allows us to perform the resection only of that part of the PDC that directly interacts with the gland head and tumor. Gastrectomy was not performed. Such volume of the resection allows us to restore the continuity of the digestive tract in a more physiological way with the creation of end-to-end gastroenteroanastomosis with the turned off jejunum loop by Ru.

The correctly selected volume of surgical intervention, the involvement of vascular surgeons and other technical issues leads to complete cure of the patients. The radical nature of the intervention completely eliminates the risk of recurrence of the disease in the future.

CONCLUSION

Thus, previously unknown information was obtained about the possible location of lymphangioma in the stroma of the pancreas with the growing into its structures. In this case, the course of the disease with the growth of the portal vein wall was established for the first time. The only radical method of treatment of the disease, in the development of this scenario, is pancreatoduodenal resection with removal of the affected part of the pancreas together with part of the portal vein. Restoration of bile outflow was made by creating an anastomosis between the common hepatic duct and jejunum, the integrity of the portal vein was restored by the imposition of venous (v.porta) anastomoses. Despite the benign nature of lymphangioma, the location of the tumor deep in the tissues of the head of the pancreas, the involvement of bile ducts and large vessels in the process makes this volume of operation the optimal.

REFERENCES

- [1] Artemieva, N.N., Kokhanenko, N.Yu., True pancreatic gland cysts, *XVI international congress of hepatology CIS countries "Actual problems of surgical hepatology"*, Ekaterinburg 2009.
- [2] Borisov, A.E., Kubachev, K.G., Yakhonov, S.P., Kachabekov, M.S., Cysts of the pancreas, *Bulletin of St. Petersburg MAPO* 2011, 3(4), 33-37.
- [3] Vinogradov, V.V., Aripov, U.A., Danilov, M.V., *Cysts of the pancreas glands*, Medicine, Tashkent 1975.

- [4] Kloppel, G., Pseudocysts and non-neoplastic cysts of the pancreas, *Semin. Diagn. Pathol.* 2000, 17(1), 7-15.
- [5] Rosien, U., Layer, P., Cystic lesions of the pancreas, *Med. Klin.* 1999, 94(7), 377-385.
- [6] Artemyeva, N.N., Savinov, I.P., Savrasov, V.M., et al., Surgical tactics for pseudocysts of the pancreas, *Annals of the surgical hepatology* 1997, 2, 74-75.
- [7] Krasilnikov, D.M., Fedorov, V.V., Minigaleev, M.M., Siraziev, I.Sh., Salimzyanov, Sh.S., Surgical tactics for chronic benign pancreatic diseases, *Annals of surgical hepatology* 1999, 4(2), 161-162.
- [8] Tokin, A.N., Chistyakov, A.A., Mamalygina, L.A., et al., Treatment of pseudocyst of pancreas, *Annals of surgical hepatology* 1999, 2, 176-177.
- [9] Bagnenko, S.F., Kurygin, A.A., Rukhlyada, N.V., Smirnov, A.D., *Chronic pancreatitis: A guide for doctors*, Peter, St. Petersburg 2000.
- [10] Vashko, R.V., Tolstoy, A.D., Kurygin, A.A., et al., *Acute pancreatitis and pancreatic injury. A guide for doctors*, Peter, St. Petersburg 2000.
- [11] *Information materials on emergency surgical care in acute surgical diseases of the abdominal cavity in St. Petersburg*, SRI Research Institute. I.I. Dzhanelidze, St. Petersburg 2011.
- [12] Brekhov, E.I., Kalinnikov, V.V., Izotov, R.A., Minimally invasive methods of treatment of false pancreatic cysts, *Annals of surgical hepatology* 2008, 13(3), 153-154.
- [13] Karpanova, N.I., Kashirskaya, N.Yu., *Cystic fibrosis: Monograph*, Medpraktika, Moscow 2014.
- [14] Shalimov, A.A., *Diseases of the pancreas and their surgical treatment*, Meditsina, Moscow 1970.
- [15] Rybachkov, V.V., Shvetsov, R.V., Utkin, A.K., Damage of pancreas and its consequences, *Annals of surgical hepatology* 2004, 9(2), 191.
- [16] Saveliev, B.C., Filimonov, M.I., Gelfand, B.R. et al., Destructive pancreatitis. Standards of diagnosis and treatment, *Annals of surgical hepatology* 2000, 6(2), 115-122.