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Case Report

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Pancreatic Cystic Neoplasms: The Challenges in Diagnosis and Review of Treatment

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1. Abstract

Pancreatic Cystic Neoplasm (PCN) is an asymptomatic tumor which is increasingly being found by advanced imaging techniques in recent years. We report the case of a 56-year old man who presented as an outpatient with intermittent abdominal pain and abdominal distension for one year. His blood tumor markers showed that serum cancer antigen (CA) 19-9 is 1000.00U/ml. Gastro scope found severe gastric varices and Computed Tomography (CT) of portal vein revealed multiple low density foci occur in the pancreas and splenomegaly. Endoscopic ultrasonography (EUS), Magnetic Resonance Cholangiopancreatography (MRCP) and computed tomography (CT) of pancreas suggested that the lesions in the body and tail of pancreatic are cystic neoplasm, which involved the splenic vein. After removing the pancreas (body tail) and spleen of this patient, it is pathologically confirmed as moderately/poorly pancreatic ductal adenocarcinoma (PDAC). The published date was reviewed to settle the issues of optimal diagnostic methods and therapies of PCN.

2. Introduction

Due to the wide application of imaging, the diagnosis frequency of pancreatic cysts is increasing. This condition increases with age. Pancreatic cystic lesions include true cysts, pseudocysts, and pancreatic cystic neoplasm (PCNs). Due to the malignant potential of PCNs, differentiating them from non-neoplastic cysts and benign PCNs is critical. We present a case about pancreatic cystic neoplasms and review the published literatures to investigate the optimal diagnostic and treatment strategies.

3. Case Report

A 56-year old man who presented as an outpatient with intermittent abdominal pain and abdominal distension. His other medical history included a surgery for varicose veins of both lower limbs. Two years prior to this presentation, he was diagnosed with a gastric ulcer.

When he presented at the hospital as an outpatient, he complained of abdominal pain and abdominal distension. The pain was located in the upper abdomen. These symptoms persist for about a year and become increasingly severe and unbearable. But no obvious abnormality was found in physical examination except varicose veins in the lower extremities. His gastro scope found severe gastric varices with red-color sign, and no active bleeding (Figure 1). So he was admitted to the department of gastroenterology. For the isolated gastric varices (IGV), he was first suspected of liver cirrhosis, but he had no medical history of viral hepatitis, no drinking, and the liver fibrosis determination was normal. Laboratory findings were showed: carcinoembryonic antigen (CEA), 185.7ng/ml and CA19-9,1000.00U/ml. Due to the increase of CA19-9 and no evidence of liver cirrhosis, we did more examinations. The investigation revealed multiple low density foci occur in the pancreas on computed axial tomography (CT) of portal vein (Figure 2). These low density foci are multiple cystic lesions of the pancreas, with the largest lesions about 1.6 cm in diameter. In order to get a clear view, the patient did EUS and MRCP. The images of them revealed that the cystic lesions

are distributed in the pancreatic body and tail, and suspected involvement of the splenic artery. He was taken a laparotomy and surgical treatment. After laparotomy, a 4*5cm tumor was found in the body of the pancreas. The tumor was hard, adherent to the surrounding tissues, and the para-aortic lymph nodes were enlarged. The surgical team resection the para-aortic lymph nodes and send intraoperative frozen. Freezing results are reported as adenocarcinoma. So they decided to perform distal pancreatectomy and splenectomy. After the operation, the specimens were sent to the pathology examination to be confirmed as moderately/poorly differentiated pancreatic ductal adenocarcinoma, and partly mucinous adenocarcinoma. The fibrous tissue of the spleen was infiltrated by cancerous tissue. The fibrous tissue of the spleen capsule and one of five lymph nodes sent for examination were infiltrated by cancerous tissue. (Figure 3, 4) He was continued on intravenous antibiotics and recovered well postoperatively and was discharged home 2 weeks later. The patient had an uneventful postoperative course (3 month after the procedure).

Figure 1: The gastro scope image shows severe gastric varices with red-color sign (September 7, 2018).

Figure 2: Computed axial tomography of portal vein reveals multiple low density foci occur in the pancreas (September 11, 2018).

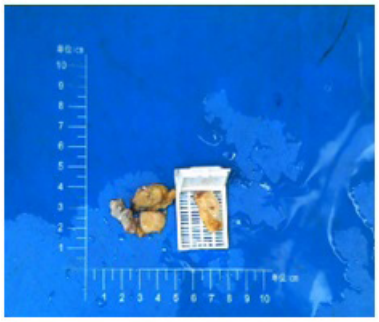

Figure 3: Distal pancreatectomy and splenectomy found a 4*5cm tumor in the body of the pancreas (September 28, 2018).

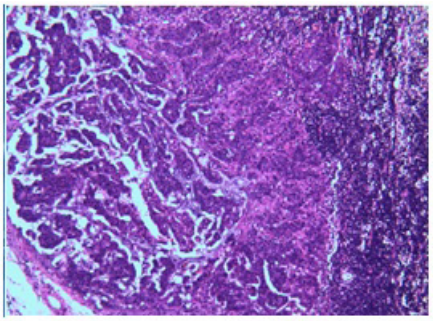

Figure 4: Pathological images of specimens to be confirmed as moderately/poorly differentiated pancreatic ductal adenocarcinoma, and partly mucinous adenocarcinoma (September 28, 2018).

4. Discussion

In clinical practice, most patients of PCNs are asymptomatic, and only a few presented with non-specific abdominal pain, nausea and vomiting, marasmus, obstructive jaundice, and new-onset diabetes mellitus. Symptoms of abdominal pain, jaundice, weight loss, and anorexia had associations with malignant tumors [1]. SEER database analysis showed that malignant transformation of these cysts is very rare.

IGV can occur in patients who has left-sided portal hypertension (LSPH) resulting from splenic vein occlusion caused by thrombosis or stenosis. That leads to IGV [3]. The most common diseases that cause splenic vein occlusion are pancreatic diseases, such as pancreatic cancer, pseudocyst, or a pancreatitis [2].

Examination of the patient can be performed from imaging, biomarkers, endoscopy, pathology and other aspects. In order to evaluate PCNs, kinds of imaging examination can be used, including US, CT, MRI, PET and MRCP. Endoscopic examination includes EUS, FNA. The accuracy of distinguishing mucinous from non-mucinous PCN by EUS alone is low, and the accuracy can be greatly improved by combining with cystic fluid test [3].The cystic fluid can be extracted by FNA, and its complication rate is low.

Biomarkers are useful for the clinical identification of pancreatic cyst types or for the identification of highly dysplasia or cancer. In IPMN, if you are worried about malignant transformation, you can refer to ca19-9.The recognized threshold for cyst fluid CEA is 192ng/mL, with an accuracy ranging 70-86%, sensitivity of 61-89%, and specificity of 63-77%, superior to all other tests (EUS, cytology, CA125, and CA 19-9) [4]. Other biomarkers include amylase, which can exclude pancreatic pseudocyst when cyst fluid amylase <250U/L.

Differentiation of pancreatic pseudocyst from PCN is the key to treatment. There are four main types of PCNs: SCNs, MCNs, IPMNs, and SPNs. The surgical indications suggested of SCNs are symptomatic and greater than 4 cm in size. If there is no symptom or the size is less than 4cm, the patient should be re-examined for once a year. If there are signs of malignant transformation, the patient needs immediate surgical treatment [5]. The risk of malignancy

in MCNs was 17.8%. All current guidelines recommend that MCNs should be surgically removed regardless of the presence or absence of malignancy. IPMNs has obvious tendency to become invasive carcinoma. According to the involvement of pancreatic ductal system, IPMNs are classified as either main-duct IPMN (MD-IPMN) or branch-duct IPMN (BD-IPMN) [6]. All guidelines consider MD-IPMN is an indication for surgery. For the patients, which is definitely or presumed to be BD-IPMN, cysts greater than 3 cm, or for cysts less than 3 cm with symptoms, and the pancreatic duct dilation is greater than 6mm or nodule, suggesting surgical resection [7]. For cysts smaller than 3cm without symptoms, a semiannual review should be taken. If the cyst growth rate ≥5mm/year, cyst diameter ≥4cm, MPD diameter between 5 and 9.9mm, serum CA 19-9 level >37U/ mL in the absence of jaundice, new-onset diabetes mellitus or acute pancreatitis, and contrast- enhancing mural nodules <5mm can be considered for surgical resection [8]. SPN is a low-grade malignant tumor, and the main treatment is surgical resection.

With the development of imaging technology, the diagnostic rate of PCN is obviously improved. FNA should be performed for patients who are difficult to be recognized by CT, MRI or EUS at the first diagnosis. If FNA find malignant cells or elevated CA19-9, the cyst should be resected. If FNA could not be performed or could not be confirmed after FNA, the cyst with symptoms or size ≥3cm, or malignant, should be resected. The patient of this case who has a pancreatic cyst did not receive FNA, but he had intermittent abdominal pain and abdominal distension, marked elevation of CA 19-9, and invasion of the splenic artery. So we sent him for a surgery, and pathology confirmed our decision. We should select appropriate examination tools and measures to diagnose pancreatic cystic neoplasm.

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