Short Communication

Gastroesophageal Varices (Bleeding) and Splenomegaly: The Initial Manifestations of Some Pancreatic Body and Tail Carcinoma

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Introduction

The most common symptoms of the pancreatic body and tail carcinoma are epigastric pain, asthenia and back pain. However, these symptoms are nonspecific, which as a consequence leads to late diagnosis as well as a low resection rate between 10% and 30%, with a 5 years overall survival rate below 10%.[1] That how to increase early diagnosis of the pancreatic body and tail carcinoma has been a difficult problem since then. Sometimes the pancreatic body and tail carcinoma can induce splenic vein occlusion, leading to pancreatic sinistral portal hypertension (PSPH). PSPH is defined as a pathological condition of splanchnic venous hypertension localized within the left-sided gastrosplenic region in pancreatic diseases. As a kind of extrahepatic portal hypertension, the main manifestations of PSPH are splenomegaly and gastrosplenic varices, without abnormalities in liver function and portal vein. PSPH is rare and curable, found in about 5–10% of the extrahepatic portal hypertension cases. The pancreatic body and tail carcinoma is a main cause for PSPH, and in some cases gastroesophageal variceal hemorrhage and splenomegaly are noted as the initial symptoms. Unfortunately, the correlation between pancreatic body and tail carcinoma and PSPH has not been fully recognized, which sometimes results in misdiagnosis and delay in treatment. In this study, we report our experience of 18 patients confirmed of pancreatic body and tail carcinoma with gastroesophageal varices and splenomegaly as initial clinical manifestations from 2009 January to 2012 December in Peking Union Medical Collage Hospital.

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Methods

Patient characteristics

Eighteen patients were enrolled in this study. There were 11 males and 7 females, with a mean age of 47.1 ± 6.2 years (range 23–65). The inclusion criteria are: (1) Definitive evidence of the pancreatic body and tail carcinoma; (2) portal hypertension symptoms, such as splenomegaly, gastric varices with or without esophageal varices, plus positive findings shown by computed tomography or gastroscope; (3) finding early presence of gastroesophageal varices, hematemesis, melena or splenomegaly, often as initial clinical manifestations. The exclusive criteria are: (1) Abnormal liver function, (2) with hepatitis, hepatocirrhosis, blood system diseases, or schistosomiasis, etc., (3) with regional portal hypertension caused by nonpancreatic diseases.

The initial manifestations of these patients were gastroesophageal varices in 6 cases (33%), hematemesis and melena in 5 cases (28%) and splenomegaly in 7 cases (39%). The lab tests showed normal liver function in all 18 patients, hypersplenism (white blood cell [WBC] and platelet were below the normal range) in 15 cases, and serum CA199 level elevation in 16 cases. The Doppler ultrasound showed splenomegaly (Stage I in 13 cases, Stage II in 5 cases, Stage III in 0 cases) and splenic vein occlusion in all 18 patients. Three-dimensional reconstruction of the portal vein by computed tomography found neither dilatation nor thrombosis of the portal vein, but had splenic vein occlusion and gastric varices in all 18 cases [Figure 1]. Gastroscope or upper gastrointestinal radiography found gastric varices in all 18 cases, and esophageal varices in 4 cases.

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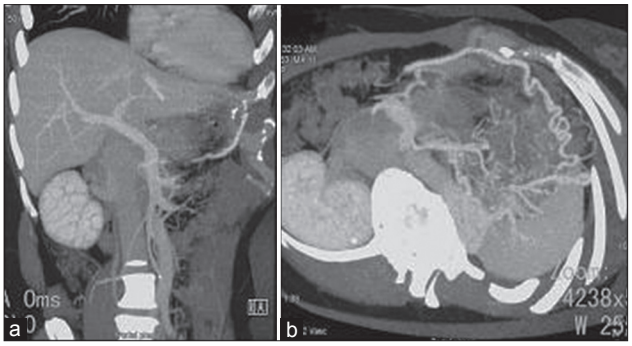

Figure 1: (a) Portal vein and superior mesenteric vein are normal, no dilatation and thrombosis (b) splenic vein occlusion, gastric varices and collateral circulation formed.

Therapeutic options

Ten of the 18 patients underwent surgical operations. Eight patients underwent distal subtotal pancreatectomy, gastric devascularization, and splenectomy while the other two got distal subtotal pancreatectomy, gastroesophageal devascularization combined with splenectomy. In 8 cases, radiotherapy and chemotherapy were applied, using gemcitabine chemotherapy in 5 cases and gemcitabine chemotherapy plus radiotherapy in 3 cases, because the tumors were found to have metastasized, or because the patients refused surgery. During the follow-up period, five of these patients who underwent conservative therapy had upper digestive tract hemorrhage, and they were treated by medication that is, hemostatics, somatostatin and acid suppressive medications in 5 cases, endoscopic variceal ligation and sclerotherapy sessions in 3 cases, and splenic artery embolization in 1 case.

Follow-up

Follow-up was achieved by outpatient visit and telephone communication. We used pathological diagnosis as the start point of follow-up and survival time. Seventeen of the 18 patients got followed-up in a period of 12–60 months.

Statistic analysis

Data were expressed as the mean ± standard error of the mean. Survival analysis was performed using Kaplan–Meier method. All statistical analyses were performed with a computer program Statistical Package for Social Sciences (SPSS) version 13.0 (SPSS Inc., Chicago, IL, USA). In all cases, the values of $P \leq 0.05$ were considered as statistically significant.

Results

Surgical data

Ten of the patients underwent surgical resection with a resection rate of 55.5% (10 out of the 18) in this study. We found that the pancreatic body and tail carcinomas were all located within the pancreatic capsule and no sign of metastasis, normal liver, splenomegaly, and short gastric vein, gastroepiploic vein, gastric coronary vein dilated in gastrosplenic region. Surgeries in all patients were successful, and there were no mortalities. The postoperation length of stay was 15.3 ± 4.9 days (range 12–45 days), with pancreatic fistula in 1 case and delayed gastric emptying in 1 case. Thrombocytosis was found in all the operation cases, and the platelet count was controlled in normal range after aspirin intake. In eight nonoperative cases, two patients who had hematemesis or melena history before were found with upper digestive tract hemorrhage during radiotherapy. After conservative treatment that was of little effect, the hemorrhage was controlled by endoscopic and interventional therapy. Three patients had upper digestive tract hemorrhage during radiotherapy or chemotherapy for the first time, and one of them underwent conservative therapy successfully, while the other two underwent endoscopic therapy after the failure of conservative therapy.

Pathological features

The pathology of the 10 operative patients was: Duct adenocarcinoma in 9 cases, cystadenocarcinoma in 1 case, and the spleen all showed chronic congestive splenomegaly. The pancreatic puncture pathology of the eight nonoperative patients was all adenocarcinoma.

Follow-up results

The patients were followed-up for a duration range 12–60 months, and only one nonoperative patient lost of follow-up (follow-up rate 94.4%). All operative patients found no upper digestive tract hemorrhage and disappearance of hypersplenism after surgery. Gastric varices also disappeared after 3 months confirmed by endoscope or computed tomography. Hypersplenism was not worse in the nonoperative patients during the follow-up. However, five patients had upper digestive tract hemorrhage and managed by drug therapy, endoscopic therapy or interventional therapy. Two patients survived until now, and the others died because of recurrence or complications, with a median survival time of 18.0 ± 2.1 months.

Discussion

Pancreatic portal hypertension is a special form of portal hypertension. As reported by previous literatures, pancreatic body and tail carcinoma is the second major cause of PSPH.[2] The nonspecific nature of these symptoms often contributes to a delay in diagnosis that may lead to poor outcomes. Thus, early diagnosis and therapy is of great significance to improve the prognosis of pancreatic body and tail carcinoma. In 1952, Marks *et al*.[3] first reported a pancreatic body and tail carcinoma presenting gastric varices bleeding because of splenic vein thrombosis as initial symptoms. Since then, there have been several other case reports of pancreatic carcinoma with gastroesophageal varices bleeding and splenomegaly as initial symptoms. In pancreatic body and tail carcinoma and PSPH patients, the early nonspecific symptoms such as abdominal discomfort and anorexia may mislead the physicians to gastritis or other diseases, while it would be too late for them when weight loss, abdominal

mass or back pain were present. PSPH mainly manifest splenomegaly, hypersplenism and gastroesophageal varices bleeding specifically and is easy to diagnose. We believe that if we can be sensitive with these manifestations of PSPH, which is possibly caused by pancreatic body and tail carcinoma, early detection and diagnosis as far as possible may be achieved.

Routine blood test, liver function test, tumor markers and radiography are of great significance to the diagnosis and differential diagnosis of PSPH and pancreatic body and tail carcinoma. The WBC and platelet number can tell us the degree of hypersplenism, and liver function can help us exclude hepatic diseases, which can also cause portal hypertension, while the tumor markers are important hints in diagnosis pancreatic body and tail carcinoma. Ultrasound examination and computed tomography can find pancreatic diseases as well as splenic vein thrombosis, collateral circulation, and gastric varices. Gastroscope and alimentary canal visualization can evaluate the degree of gastric varices and are also of use in the treatment of gastric varices bleeding.

The treatment of PSPH caused by pancreatic body and tail carcinoma is usually composed of two aspects. Clinically the pancreatic body and tail carcinoma should be resected if allowed. Otherwise, comprehensive treatment that is, radiotherapy, chemotherapy and biotherapy is available when it is too late to remove the tumor by surgery. Academically, there are no criteria in the treatment of PSPH. In our opinion, we should choose different methods according to different situations. If there were evidence of splenic vein thrombosis, gastric varices with or without splenomegaly, we choose distal pancreatectomy, gastric devascularization plus splenectomy. While for those who still had esophageal varices we add esophageal devascularization as well. In our study all the 10 patients followed these criteria and they showed no upper digestive tract bleeding and disappearance of hypersplenism as well as gastric varices after 3 months. For those whose tumors may not be resected by surgery, our aim is to improve the quality of life and increase the life span. Therefore, we should focus on prevention and management of the complications caused by portal hypertension. The complications by PSPH are mainly gastroesophageal varices bleeding and low WBC and platelet number in case of hyperspenism. Ku *et al*.[4] divided the hemodynamic changes in PSPH into three stages. In his system, at first there are no gastric varices and splenomegaly when splenic vein is partly occluded. Then the splenic vein is totally occlusive while the splenic artery remains open, resulting in gastric varices and splenomegaly. At last the splenic artery is invased and obstructed, while the gastric varices and splenomegaly get relieved. In that case the second stage is the most dangerous and clinically obvious time in PSPH. If the patients can get diagnosed according to gastric varices bleeding and splenomegaly at this time, most of them may still have operation opportunity. The resection rate in our study was 55.5%, which was higher than other reports, indicating that early diagnosis and treatment are possible for the patients with pancreatic body and tail carcinoma manifesting gastroesophageal varices bleeding and splenomegaly.

In PSPH hypersplenism gradually develops and can be relieved later in case of splenic artery occlusion, so patients seldom have a severe hypersplenism. In our study, the splenomegaly was mainly in Stage 1, accounting for 70% of the cases and no Stage 3 splenomegaly was found. So we can pay less attention in the hypersplenism in these patients. Nevertheless, the gastroesophageal varices bleeding more urgent and may cause death when bleeding happens. We conclude that the prevention and treatment of upper digestive tract bleeding in PSPH are more important in these cases. The patients can take soft diet or nonselective beta receptor blocking drugs to prevent gastroesophageal varices bleeding, while to those who have had bleeding, they should acquire more active treatment. Surgical operation is the best method to cure and prevent bleeding thoroughly, including splenectomy and gastroesophageal devascularization. However, for patients in poor conditions such as with instable hemodynamic status or advanced malignancy, who are ineligible for surgical management, there are many conservative therapies instead. Medications, including hemostasis, acid suppressive, antibiotics and somatostatin medications can be used. Some minimal invasive procedures, such as endoscopic ligation or sclerotherapy,[5] or splenic artery embolization can be selected when drug therapy failed. We recommend treating patients first with medication or medication plus endoscope, and then splenic artery embolization when the above therapies are ineffective. There were 5 cases who were ineligible for surgical management had upper digestive tract bleeding, and all of them achieved good results through conservative methods.

Pancreatic body and tail carcinoma is one of the major causes of PSPH. Some patients may show portal hypertension as initial manifestations. In that case, we should be alert of this possibility. Therefore, we should aim to achieve early diagnosis and treatment as far as possible of pancreatic body and tail carcinoma patients through clinical manifestations of gastroesophageal varices (bleeding) as well as splenomegaly. Surgical treatment may be the best method to prevent and treat upper digestive tract bleeding in PSPH, and we should also pay attention to the prevention and treatment of complications of portal hypertension during the treatment of the primary carcinoma.

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