

Case Report

Primary Castleman disease in peripancreatic tissue regarded as pancreatic malignance: a case report

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Abstract: Castleman disease is a rare, usually uncommon benign lymphoproliferative disorder with unknown etiology and pathogenesis. It is commonly found in mediastinum. Peripancreatic Castleman disease is rarely reported. We present a case of peripancreatic tumor of a 58-year-old Chinese woman with chief complaint of 10 kg weight loss, jaundice and poor appetite for one year. The patient had an uneventful postoperative course. Histopathological examination revealed hyaline-vascular Castleman disease. No recurrence was found during the follow-up period.

Keywords: Castleman disease, lymphoproliferative disorder, hyaline-vascular, peripancreatic tumor

Introduction

Castleman disease (CD), first described in 1954 [1], is a rare lymphoproliferative disorder which is commonly found in mediastinum [2, 3]. After then, more and more other locations of CD were found [4, 5]. It is also reported that CD is always preoperatively diagnosed as tumors [6, 7]. Some reports [8-11] show CD is related to pancreas, but CD arising in peripancreas is rarely reported.

CD can be divided into 3 types according to pathologic characteristics: hyaline-vascular type, plasma-cell type and mixed cell type. Hyaline-vascular type is the most common type in CD characterized by hyperplasia of the hyaline-vascular follicles and capillary proliferation. It is usually asymptomatic and presents as a mass. Whereas the less common type is plasma-cell type characterized by plentiful plasma-cell infiltration in the interfollicular tissue [12]. It usually shows systematic symptoms like fever, anemia, weight loss, night sweat and generalized lymphadenopathy [13-15]. The mixed cell type is really rare. It can also be divided into the other 2 subtypes according to clinical characteristics: unicentric CD (UCD) and multicentric CD (MCD). The most commonly seen type is UCD form which is usually asymptomatic and presented as a mass or swelling. Meanwhile, MCD is characterized by fever with chills, ane-

mia, generalized lymphadenopathy and hepatosplenomegaly [1]. CD is often ignored by clinicians because of not frequently encountering in clinical. Here, we report a case of Castleman disease in peripancreatic tissue which was preoperatively diagnosed as pancreatic malignancy.

Case report

A 58-year-old woman with chief complaint of 10 kg weight loss, jaundice and poor appetite for one year was suggested as pancreatic malignancy by the computer tomography (CT) images in local hospital. She was referred to our hospital for further diagnosis and treatment. Routine physical examinations and routine laboratory investigations were normal. Especially, the tumor markers of alpha-fetoprotein (AFP), carcinoembryonic antigen (CEA), and carbohydrate antigen 19-9 (CA19-9) were 4.52 ng/mL, 0.772 ng/mL, and 14.69 U/mL.

The endoscopic ultrasonography and fine needle aspiration (EUS-FNA) of the pancreas revealed a 3.5 × 3.3 cm mild enhancing solitary mass at the pancreas head and neck (**Figure 1**). The aspiration biopsy cytology was performed and showed lymphoid hyperplasia (**Figure 2**).

On the fluorine-18-fluoro-2-deoxyglucose position emission tomography/computer tomogra-

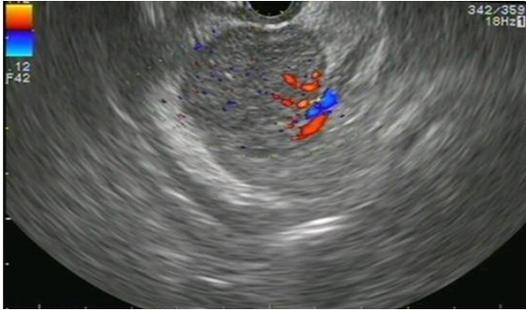


Figure 1. The endoscopic ultrasonography shows mass lesions in the body and neck of the pancreas.

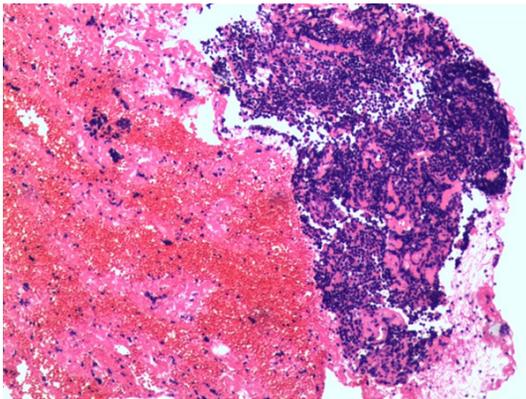


Figure 2. Hematoxylin and Eosin stained slide of pancreatic puncture.

phy (^{18}F -FDG PET-CT) scanning, a localized uplift in the pancreatic neck about 1.7×2.8 cm was supposed as pancreatic malignancy. Lymph nodes less than 0.8 cm adjacent to aortaventrals were visible. In the right lung, two micro nodules appeared in lung apex, one in the lower lobe; while in the left lung, one in left lung apex, one in fissure obliqua pulmonis, which were all less than 0.4 cm and similar to a lymph node. The spleen was in normal size and had homogenous internal structure. The pancreas was normal in size and position, and both kidneys were normal in size and position too (**Figure 3**).

The patient underwent a surgical excision of the mass and dissection of lymph nodes that revealed a lymphadenoid mass on the pancreas superior border and ahead common hepatic artery. Multiple enlarged lymph nodes invaded hepatoduodenal ligament. The pancreas was normal in size and position. Hematoxylin and eosin (HE) staining of specimen in peripancre-

atic tissue and hepatoduodenal ligament revealed essentially preserved lymph node architecture. There was expansion of mantle zone in lymphatic nodules accompanied by burnt out germinal centers. Para cortical areas showed endothelial hyperplasia of vascular channels, some of which encroached the germinal centers (**Figure 4**). Then immunohistochemical (IHC) staining was done in which neoplastic elements expressed the immunophenotypic profile CD20+, Bcl6+, Bcl2-, CD5+, CD21+, CD23+, CD38+. The proliferation index of Ki67 was 2+.

Discussion

CD is a benign and rare vascular lymphoproliferative disorder characterized by enlarged hyperplastic lymph nodes. It is pathologically classified into two major types: hyaline-vascular and plasma-cell CD, first described by Keller, et al. [16]. The most commonly-seen pathological form is the hyaline-vascular type, characterized by small hyaline-vascular follicles and capillary proliferation. Histopathologically, the hyaline-vascular type consists of small lymphocytes, vascular stoma, and plasma cells clustered around multiple germinative centers in the form of an onion skin. In the HE staining of our case, it demonstrates expansion of mantle zone in lymphatic nodules accompanied by burnt out germinal centers like an onion skin. While, in the IHC staining, CD20, CD21 and CD23 were positive indicated B lymphocyte originated, Bcl-6 was positive showed marked expansion of the mantle zone, Ki-67 was mid-strong positive indicated proliferation actively, CD34 was positive hard indicated the endothelial hyperplasia of vascular channels. The definitive diagnosis of hyaline-vascular CD is obviously confirmed. The hyaline-vascular cases are usually asymptomatic. The patient in this case was detected by healthy examination even though she had been weight loss, jaundice and poor appetite for one year.

In this case, the patient was supposed to be pancreatic malignancy at first for the enhancement in CT and no significant symptoms but for weight loss, jaundice and poor appetite. The serum levels of tumor markers were normal, especially CA19-9, which was regarded as the only serum biomarker in routine clinical use for pancreatic cancer [17, 18]. The clinical symptom of pancreatic cancer is weight loss, abdominal pain, abdominal distension, acratia, jaun-

Primary CD

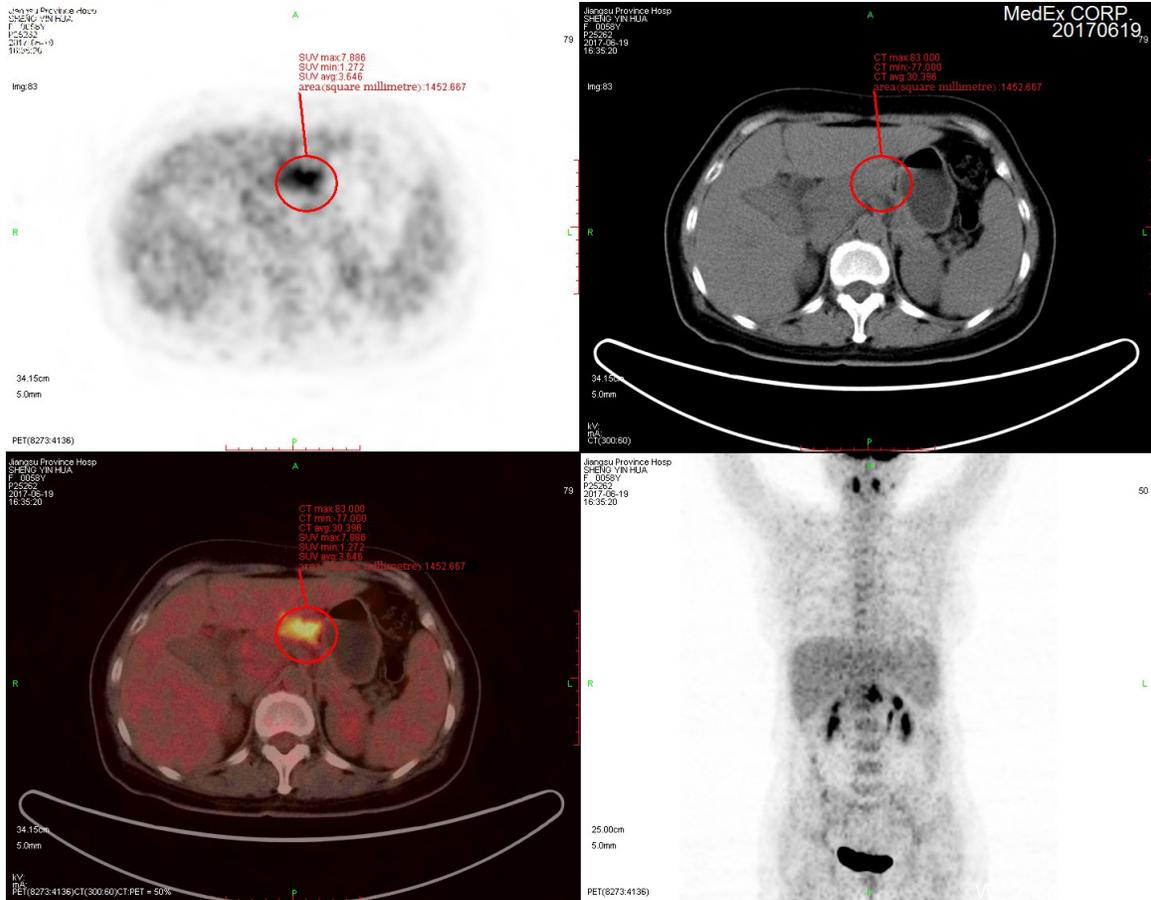


Figure 3. The PET-CT shows mass lesions in pancreatic neck.

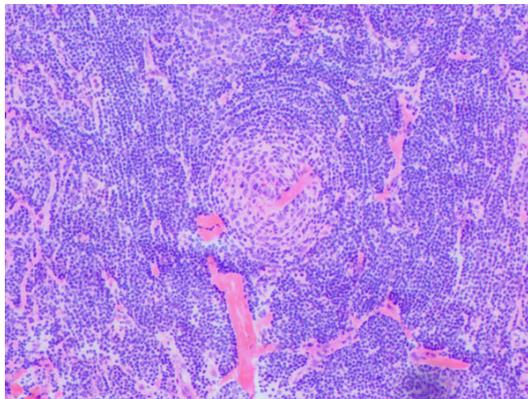


Figure 4. Hematoxylin and Eosin stained slide of peripancreatic tissue and hepatoduodenal ligament ($\times 100$ magnification).

dice and more with no obvious symptoms at an early stage. However, CA19-9 is a sialylated Lewis blood group cell surface carbohydrate antigen expressed around 95% of the population which express the Lewis antigen glycosyltransferase enzyme. That is to say, CA19-9

would not be elevated in about 5% of the population who did not express the Lewis antigen glycosyltransferase enzyme even if in pancreatic cancer patients.

The pathogenesis and etiology of CD is unknown, although most believe that a defect in immunoregulation resulting in excessive proliferation of B lymphocytes and plasma cells in lymphoid organs is responsible for its origin [19]. Our case is the same as it that B lymphocytes originated. CD can develop anywhere lymphoid tissue can be found. So that, the preoperative diagnosis is often difficult. Radiological diagnosis of the CD remains difficult [2, 20]. In our case, CT and PET-CT scan have proven to be less useful in the diagnosis of disease that have misdiagnosed as malignancy mass. The first diagnosis of lymphoid hyperplasia was made based on biopsy samples obtained preoperatively by EUS. However, this technique is still inadequate due to limited implementation and low rate of accurate diagnosis. There is no doubt that histopathology is the gold standard.

Therefore, a decision was also made to perform lesion excision and definite diagnosis was made postoperatively by investigating the specimens obtained during the surgery.

Castleman disease is rare and often ignored in clinical. It still needs to pay more attention when a mass is found in abdomen which may be regarded as the tumor. It is known that surgical removal of the localized type of the mass has long been considered standard therapy for the disease. In our patient, a complete surgical excision was accomplished and up to now, there is no evidence of recurrence.

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Disclosure of conflict of interest

None.

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