Case Report

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Duodenal Perivascular epithelioid cell tumor with liver dysfunction: a case report and literature review

Keyi Du*

Department of medicine, Zhejiang University

***Correspondence:**
Keyi Du,
Department of medicine, Zhejiang University, China.
Email: 3180106364@zju.edu.cn

Authors' contributions:
(I) Conception and design: Linping Cao;
(II) Provision of study materials or patients: Linping Cao;
(III) Collection and assembly of data: Keyi Du, Jiawei Hong;
(IV) Data analysis and interpretation: Keyi Du, Jiawei Hong;
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1. Abstract

Perivascular epithelioid cell tumor (PEComa) is a rare solid tumor, and the gastrointestinal tract is the most common pathogenic site of PEComa. Here we reported a duodenal PEComa patient with bile duct obstruction and liver dysfunction who received laparoscopic pancreaticoduodenectomy after amelioration of hepatic function with medicines therapy. Our report proved that duodenal PEComa may cause biliary obstruction and hepatic dysfunction.

2. Keywords:
Perivascular epithelioid cell tumor; Gastrointestinal tract; Duodenum; Liver dysfunction.

3. Introduction

Perivascular epithelioid cell tumor (PEComa) is a rare solid tumor, which express myomelanin markers[1-3]. PEComa is most commonly found on the gastrointestinal tract (GI), while duodenal PEComa is relatively rare. Here we report a duodenal PEComa case with bile duct obstruction and liver dysfunction. We reviewed the clinical features and treatment of gastrointestinal tract PEComa, especially duodenal PEComa, and concluded that surgical intervention should be actively performed for gastrointestinal tract PEComa with digestive tract symptoms, hemorrhage, adjacent organs influence or neuroendocrine dysfunction.

4. Case report

A 30-year-old man presented to our clinic institution with a 1-month history of liver dysfunction. The patient's liver function test showed that ALT (alanine transaminase) was 699 U/L and AST (aspartate aminotransferase) was 277 U/L. While the patient's tumor markers were normal. Further abdomen enhances computerized tomography (CT) demonstrated a uniform mass in the duodenal papilla area about 32 * 24 mm in size, which could be enhanced in arterial phase. The MRCP (magnetic resonance cholangiopancreatography) also showed a long T2 signal tumor in the duodenal papilla area with "Beak like" stenosis at the lower end of common bile duct and gallbladder enlargement (Figure 1.A-C). The patient underwent laparoscopic pancreaticoduodenectomy after amelioration of hepatic function with medicines therapy (Figure 1.D) with multidisciplinary diagnosis and treatment (MDT). Within two weeks after surgery, the patient's liver function returned to normal.

Figure 1: the pre-operative imaging and surgical specimen of a 30-year-old man with Duodenal Perivascular epithelioid cell tumor and liver dysfunction. Contrast-enhanced CT shows the arterial phase imaging(A) and the vein phase imaging(B) of the patient's duodenal perivascular epithelioid cell tumor (red arrow); (C) the MRCP imaging shows the patient's biliary tract dilatations due to the external compression on the lower end of common bile duct; (D) laparoscopic pancreaticoduodenectomy's surgical specimen.

The intraoperative frozen section offered a diagnosis as vascular eosinophilic tumor. The postoperative pathology finally offered the diagnosis as PEComa. Immunohistochemistry showed that SDHB (+), Ki-67 (+, 15%), Desmin (smooth muscle +), CD34 (vascular +), SMA (+), , HMB45 (+), β -catenin (plasma +), Melan A (+), , Calponin(CP) (smooth muscle +), CD3 (+), CD20 (+), CD30 (+), CD21 (+), Bcl-2 (+), Bcl-6 (+), CD10 (+),PD-1 (+), IgG (small +), IgG4 (+), (some immunohistochemistry results is showed in Figure 2). Five lymph nodes were examined at the same time, and no tumor metastasis was observed. And no recurrence or other complications were observed in the follow-up for more than 17 months at the time of writing.

Figure 2: Postoperative pathologic imaging of patient's duodenal perivascular epithelioid cell tumor specimen (A) The HE staining of

tumor; (B) the HMB45 staining; (C) the Melan-A staining; (D) the SMA staining. (x100)

5. Discussion

PEComa is a rare solid tumor with unique morphology and expression of myomelanocyte markers[1]. And it was also defined as a family of mesenchymal neoplasm[4]. Bonetti et al first proposed the term perivascular epithelioid cells (PEC), describing some perivascular epithelioid cells with melanocyte markers immune reactivity in 1992[5]. World Health Organization defined PEComa as " mesenchymal tumors composed of histologically and immunohistochemically distinctive perivascular epithelioid cell " in 2002[1]. Duodenal PEComa ranked fourth among the most common gastrointestinal PEComa, followed by Colon, mesentery and Stomach PEComa[1,6]. PEComa has a malignant potential. WHO's statement on the malignancy PEComa: PEComas displaying any combination of infiltrative growth, marked hypercellularity, nuclear enlargement and hyperchromasia, high mitotic activity, atypical mitotic figures, and coagulative necrosis should be regarded as malignant[1]. Folpe and colleagues suggested that malignancy could be predicted by satisfying two or more of the following features: a >5-cm tumor, infiltrative growth, high nuclear grade and cellularity, the presence of >1 mitosis in 50 HPF, tumor necrosis, and vascular invasion[7]. PEComas presented non-specific clinical signs. The main clinical manifestations are abdominal pain, black feces, rectal bleeding, obstruction, weight loss, anemia, some even asymptomatic. The most common is abdominal pain (35%), which can be caused by compression, impaction and bleeding[1]. In addition, patient might have hormonal changes due to PEComa's potential neuroendocrine functions. In our case, duodenal PEComa can lead to biliary obstruction and liver function injury which was reported for the first time. Imaging detection (CT, MRI (magnetic resonance imaging)) is of great significance for the diagnosis of PEComa. PEComa is obviously weakened during the CT venous phase, and the density of the delayed phase is lower than that of surrounding tissues[8]. On MRI imaging, the mass is bright on T1 weighted images and dark on fat-suppression images[1]. In our case, the tumor compressed the lower end of the common bile duct, resulting in significant dilatation of the upper bile duct, which was obvious in MRCP.

Table 1: The Characteristics of duodenal PEComa cases

Authors	Year	Characteristic	Follow-up time	Malignancy
Leon Toye et al	2002	Duodenal angiomyolipoma	n.d.	n.d.
Mhanna T et al	2005	Child, with a history of viral neuroblastoma	2 years	No
Michelle De Padua et al	2007	Duodenal angiomyolipoma	n.d.	n.d.
Chen Z et al	2015	With a history of periumbilical pain, dizziness, palpitation and severe black stool	4 months	No
Wang F et al	2015	Misdiagnosis of endoscopic biopsy	6 months	No
Current case	2022	With a history of liver dysfunction	1 year	No

Generally, central ulcer, hemorrhage and necrosis are also frequently detected in PEComa under the microscope, which is mainly composed of epithelioid cells (70%) and spindle cells[9]. And pathologically, GI PEComa is often well bounded without capsule formation, and sometimes has unclear boundaries or invasive growth. Immunohistochemical results showed that HMB45, Melania, S-100 and SMA were positive, CGA, syn, CK, CD117, CD10 and CD34 were negative[8]. According to a systematic review of PEComa, each GI PEComas case expressed at least one melanocyte marker. HMB-45 was considered to be the most sensitive marker. 44 (96%) of 46 tumors were positive, followed by Melan-A, 22 (65%) of 34 tumors and 5 (55%) of 9 tumors. 21 (64%) of 33 lesions were positive for smooth muscle actin. In our case, HMB45, SMA, CD10, CD34 and Melan-A immunohistochemistry markers were positive. Surgical resection is the first choice for the treatment of PEComa. Whether to combine chemotherapy or immunotherapy should be selected according to the pathological results. Sun Ju Park and colleagues reported a 7-year-old child who received interferon-α immunotherapy for 1 year after colon PEComa operation[10]. Due to the low incidence rate of, basically all case reports, whether it is need for combined chemotherapy or immunotherapy still need further large sample study. At present, there is no report of malignant transformation of duodenal PEComa, and the prognosis is good, but it needs the support of long-term follow-up data. The only five duodenal PEComa cases reported in PubMed literature searching. The characteristic for four cases of duodenal PEComa (including our case) was listed in Table 1[11-15].

6. Conclusion

Positive surgical resection is recommended in case of liver function injury, cholestasis, gastrointestinal bleeding and other symptoms. And the possibility of PEComa should be considered for patients with lower common bile duct space occupying and abnormal liver function. Surgical resection is appropriate for this kind of disease.

7. Abbreviations

ALP, Alkaline phosphatase; ALT, Alanine transaminase; AML, Angiomyolipoma; AST, Aspartate aminotransferase; CCST, Clear-cell "sugar" tumor; CT, Computerized tomography; ERCP, Endoscopic Retrograde Cholangiopancreatography; GGT, Gamma-glutamyl transpeptidase; GI, Gastrointestinal tract; HPF, High power field; LAM, Lymphangileiomyomatosis; MDT, Multidisciplinary diagnosis

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and treatment; MRCP, Magnetic resonance cholangiopancreatography; MRI, Magnetic resonance imaging; PEC, perivascular epithelioid cells; PEComa, Perivascular epithelioid cell tumor; VEGF, Vascular endothelial growth factor.

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9. Competing interests

All authors have completed the ICMJE uniform disclosure form. The authors have no conflicts of interest to declare.

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