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Case Report

Perivascular epithelioid cell tumor in the duodenum: challenge in differential diagnosis

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Abstract: Defined as a family of scarce mesenchymal neoplasm which distinctively co-express melanocytic markers and muscle markers, perivascular epithelioid cell tumors (PEComas) have been reported almost everybody site. Perivascular epithelioid cell tumors-not otherwise specified (PEComas-NOS) arising in the gastrointestinal (GI) tract are still restricted into sporadic case reports. Herein we present a case of GI PEComas-NOS which occurs in the duodenum of a 27-year-old male. Our initial diagnosis tended to gastrointestinal stromal tumor or smooth muscle tumor till the correct diagnosis of perivascular epithelioid cell tumor (PEComa) was established by postoperative pathological examination. We also make a literature review of GI PEComas-NOS and highlight the challenge it brings to the differential diagnosis.

Keywords: Perivascular epithelioid cell tumor, duodenum, differential diagnosis

Introduction

Perivascular epithelioid cell tumors (PEComas) contain a family of rare neoplasms that derive from mesenchyma. The term of perivascular epithelioid cell (PEC) was firstly proposed by Bonetti et al who used to describe some kind of cells characterized by the presence of epitheliod type and immunoreactive with melanocyte markers as well as a perivascular distribution [1]. Lately, the World Health Organization defined the perivascular epithelioid cell neoplasms (PEComas) as "mesenchymal tumors composed of histologically and immunohistochemically distinctive perivascular epithelioid cells" [2]. The family of PEComas comprise of angiomyolipoma (AML), clear-cell "sugar" tumor (CCST), lymphangioleiomyomatosis (LAM), clear-cell myomelanocytic tumor of the falciform ligament/ligamentum and rare clear cell tumors of other anatomical sites [3, 4]. The later subgroup is classified as termed perivascular epithelioid cell tumors-not otherwise specified (PEComas-NOS) which has been reported almost everybody site. The gastrointestinal (GI) tract ranks second of occurrence in PEComas-NOS with a proportion of 20% to 25% [5]. For the scarcity of such a distinctive disease, the pathogenesis, biologic behavior, natural history and prognosis of GI PEComas-NOS are out of clarity. Here we present a case report of duodenal perivascular epithelioid cell tumor (PEComa) as well as a review of the literature, hoping to have further exploration of GI PEComas-NOS.

Case report

Clinical history and presentation

Along with periumbilical pain, dizziness and palpitation, the symptom of melena became so severe and frequent that the patient, 27-year-old, came to consult and was admitted to our department. Medical history showed that the patient had melena and was hospitalized 2 years ago. The initial diagnosis was gastrointestinal stromal tumor (GIST) of duodenum based on the finds of endoscopic biopsy and contrast-enhanced computed tomography (CE-CT) examinations. After receiving symptomatic treatment, the patient decided to auto-discharge by himself when melena was in remission. On this occasion, physical examinations were largely

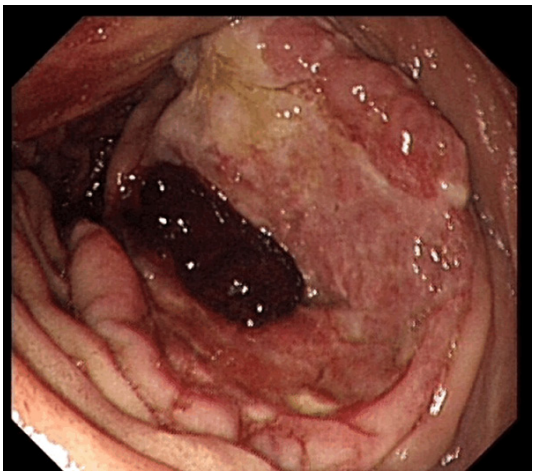

Figure 1. Finding of endoscopy. Enoscopy showed an oval mass surrounding the bowel tract in the junction between the descending and horizontal part of duodenum with ulcerative surface.

unremarkable. Laboratory investigations were largely within the normal ranges except hemoglobin reading of 5.7 g/dl and hematocrit of 18.5%.

Endoscopy and CE-CT representation

Endoscopy including biopsy and CE-CT were carried out. As for the findings on endoscopy, there were nothing abnormal detected in esophagus and stomach apart from duodenum, which displayed an oval mass surrounding the bowel tract. With the lesion Located on the junction between the descending and horizontal part and covered with blood clot, its surface is ulcerative and rugged (**Figure 1**). The biopsy tended to smooth muscle tumor but GIST. Plain CT scan revealed that there's a 68×57×82 mm well-demarcated mass in the bowel wall of the junction between the descending and horizontal part of the duodenum. The mass appeared as a rough oval with homogeneous density. CE-CT scan showed heterogeneous enhancement in that duodenal lesion and indicated that the mass bulged into the lumen and out of the duodenal wall. The tumor had definite margins surrounded by fat and mucosal defect was obvious partly. No obvious swelling lymph node was detected in peritoneal or retroperitoneal space. Identified with endoscopic finding, the scope of mass increased as compared with former CE-CT scan performed two years ago (**Figure 2**).

Treatment

Owing to the duration of dark stool and inefficiency of symptomatic treatment, we decided to perform abdominal laparotomy. During the surgery we found a mass, whose surface was covered by hematoma, in the duodenum of the junction between the descending and horizontal part, and duodenectomy was performed finally.

Pathologic findings

Grossly, the tumor was a partially encapsulated lesion in the duodenal wall of descending part measuring about 6×7×8 cm. Located in the submucosa and muscular layer, the tumor was soft and tan-white. The serosa was intact. Mass of hematoma was adhering to surface of the lesion whose appearance showed ulcerative and rugged.

Histopathological examination with hematoxylin-eosin staining was applied. Immunohistochemical studies were performed with antibodies including HMB45, Melan-A, Actin, Desmin, CD177, CD34, DOG-1 and PDGFRa. Microscopically, the lesion was found involving the submucosal and muscular layer with clear outline. The tumor was composed of sheet arranged epithelioid cells and bundles of spindle-shaped cells with clear-to-eosinophilic granular cytoplasm. The shape of nuclei was round or ovoid with prominent nucleoli. Multinucleated giant cells like osteoclast were seen in some areas so were focal necrosis (**Figure 3**). Mitotic figure was inconspicuous (1/50HPF) and lymphovascular invasion was undetected. Immunohistochemically, cells of the neoplasm were positive for HMB45, Melan-A, Actin and Desmin, while the markers including CD177 (**Figure 4**), CD34, DOG-1 and PDGFRa were negative. Evaluation of ki-67 was about 2%.

Finally, the diagnosis of PEComa was established. The patient discharged uneventfully and recovered well after a 4-month follow-up.

Discussion

Defined as a family of mesenchymal neoplasm which distinctively co-expresses melanocytic markers and muscle markers, PEComas still have unclear origin [6]. Suspicions such as

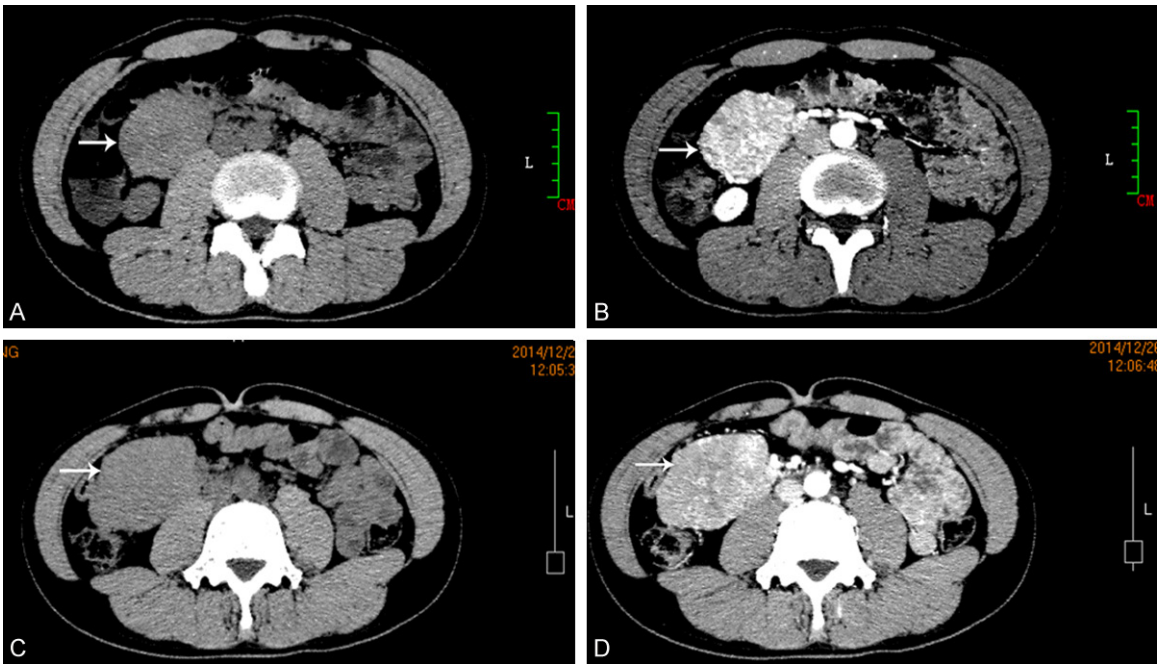

Figure 2. CT images. CE-CT scan revealed that there's a 68×57×82 mm well-demarcated mass in the bowel wall of duodenum. Arrows mark the lesion, image in the year of 2012 (A and B), image in the year of 2014 (C and D).

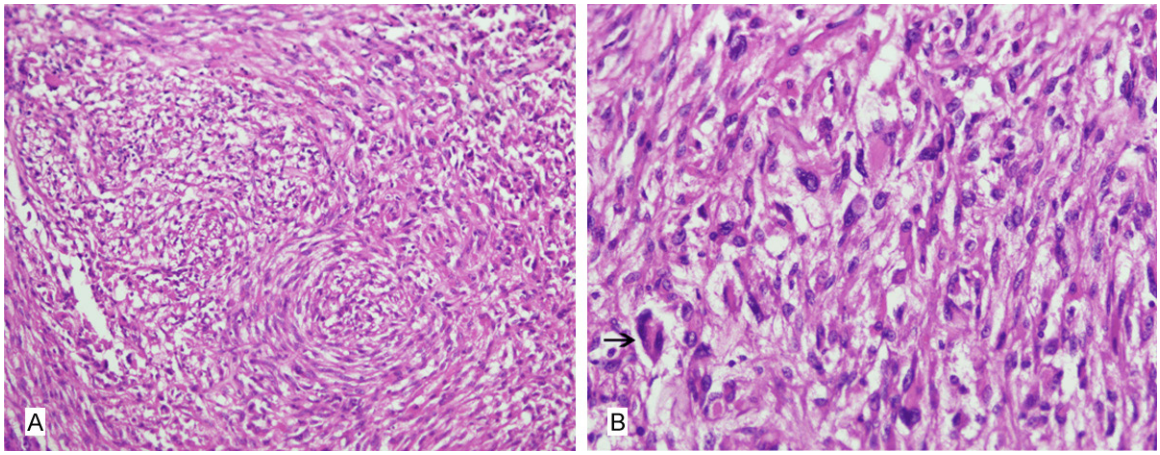

Figure 3. Photomicrograph from postoperative specimen using hematoxylin-eosin staining. In some areas, the tumor displayed sheet arranged epithelioid cells and bundles of spindle-shaped cells (A). Multinucleated giant cells like osteoclast is seen in some area, marked by arrow (B).

undifferentiated neural crest cells, a possible molecular alteration from a myoblastic smooth muscle origin or evolution from a pericytic origin were put forward [7]. Being a kind of enigmatical entity, there is no known natural cellular counterpart to PEC as well as a precursor lesion for PEComas [4, 6, 8, 9].

We focused our attention upon GI PEComas-NOS and excluded classic AML, LAM and CCST in our literature review. Ranking the 2nd of occurrence in PEComas-NOS, the understanding of GI PEComas-NOS is still confined to sporadic case reports [5]. To the best of our knowledge, there are only 33 sporadic cases (including this report) of GI PEComas-NOS recorded in the English medical literature (**Table 1**) [5, 7, 10-34]. The most frequent site of lesion is colon (N=13) [20-29], followed by small intestine (N=8) [5, 15-19], rectum (N=5) [10-14],

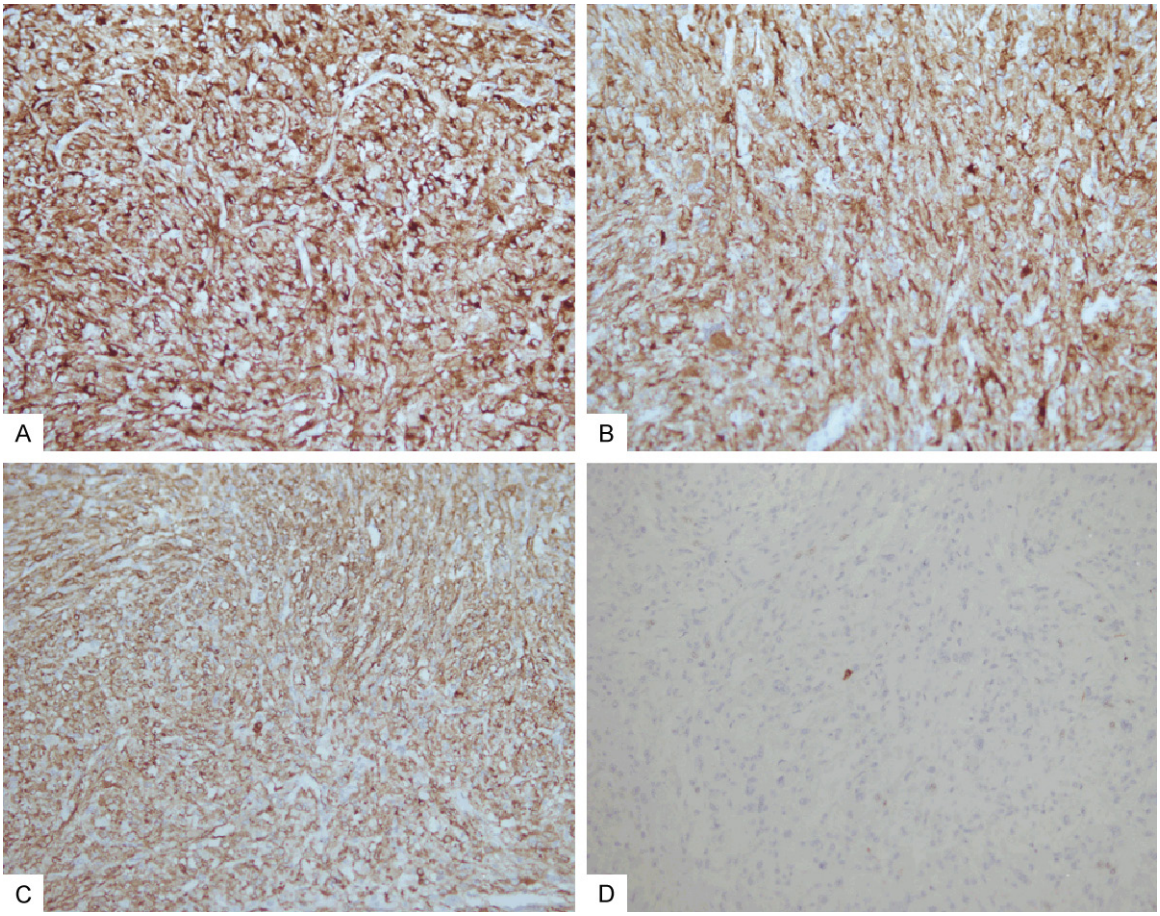

Figure 4. Photomicrograph from postoperative specimen using Immunohistochemical staining. The tumor cells were positive for HMB45 (A), Melan-A (B), and Actin (C) and negative for CD117 (D).

and stomach (N=2) [7, 30]. Elaboration to the location of small intestine, it is composed of ileum (N=4) [5, 15, 16], duodenum (N=3) [18, 19], and jejunum (N=1) [17]. Interestingly, the sporadic 3 case reports of GI PEComas-NOS in the duodenum are limited to male [18, 19]. Age of onset ranges from 5.5 to 75. With the ratio 1:2 approximately, there are 21 female and 12 male in the cases, supporting the opinion that female preponderate in PEComas.

There is a strong association between AML, LAM, CCST and tuberous sclerosis complex (TSC), a genetic disease caused by heterozygous mutations in the TSC1 (9q34) or TSC2 (16p13.3) genes, while the connection between PEComas-NOS and TSC is much less clear [4]. To date, no case of GI PEComas-NOS has been reported to associate with TSC. Corresponding with the former reports, we confirm the present case doesn't show TSC after careful examination.

PEComas-NOS bring differential diagnosis challenge to the pathologists and physicians. The differential diagnosis of GI PEComas-NOS includes GIST, smooth muscle tumor, metastatic malignant melanoma, metastatic renal cell carcinoma and paraganglioma. The most important differential diagnosis should be GIST. Morphologically, GIST and PEComas can similarly show spindle cell and/or epithelial cell appearance. The tumor cells of GIST, especially epithelioid GIST, have an eosinophilic or clear cytoplasm, lacking of perivascular concentric proliferation and typical granular cytoplasmic features. Immunohistochemically, melanocytic markers help distinguish PEComas-NOS from GIST. Sometimes PEComas-NOS expressing CD177 can mimic GIST, if cases were diagnosed as GIST in which CD177 positive is less than 50% cells, the detection of melanocytic markers is essential [4, 6]. Smooth muscle tumors should also be realized as easily confusing diagnosis. Sharing common shape of

Table 1. Review of reported cases of GI PEComas-NOS

Case/ Year	Author	Sex/Age (year)	Location	Size (cm)	IFB	Mitoses (/50 HPF)	Ki67 (%)	Tumor necrosis	LVI	Treatment	AC	Follow-up and result
1/2003	Yanai et al [17]	F/32	Jejunum	7.5	-	ND	ND	+	-	Partial resection of jejunum	-	Pelvic wall recurrence (13 mos) and ovary recurrence (25 mos)
2/2004	Birkhaeuser et al [31]	F/35	Cecum	3.5	-	0	ND	-	ND	Right hemicolectomy	-	NR (3 mos)
3/2004	Genevay et al [14]	F/35	Rectum	ND	ND	up to 5	ND	< 1	ND	ND	ND	ND
4/2004	Genevay et al [14]	F/36	Cecum	3.5	ND	≤ 5	< 1	-	ND	Hemicolectomy	ND	ND
5/2005	Evert et al [13]	F/56	Rectum	8.0	ND	286	25	+	ND	ND	ND	ND
6/2005	Mhanna et al [18]	M/12	Duodenum	3.5	+	low	< 2	-	ND	Pylorus-preserving pancreaticoduodenectomy	-	NR (24 mos)
7/2006	Agaimy et al [16]	F/63	Terminaln ileum	6.0	ND	13	60-70	+	ND	Partial ileectomy	-	Abdominopelvic area recurrence (14 mos)
8/2006	Yamamoto et al [29]	F/43	Descending colon	3.5	+	2	2.9	+	+	Partial colectomy	-	Peritoneal dissemination (20 mos) and DOT (38 mos)
9/2007	Baek et al [28]	F/16	Transverse colon	2.0	ND	0	ND	-	ND	Partial transverse colectomy	-	NR (24 mos)
10/2008	Pisharody et al [26]	M/11	Sigmoid colon	1.2	+	occasional	ND	+	+	Partial resection of sigmoid colon	-	NR (5 mos)
11/2008	Righi et al [25]	M/11	Descending/Sigmoid colon	3.5	+	infrequent	5-10	infrequent	ND	Segmental resection of the large bowel	-	ND
12/2008	Narayanaswamy et al [19]	M/34	Duodenum	3.5	+	sparse	ND	-	-	Duodenectomy	-	NR (18 mos)
13/2008	Cho et al [27]	F/16	Transverse colon	1.8	ND	ND	ND	-	-	Resection of the tumor	-	NR (41 mos)
14/2009	Tanaka et al [24]	F/14	Sigmoid colon	6.4	-	rare	ND	ND	ND	Subtotally excised of the tumor	ND	ND
15/2009	Qu et al [32]	F/43	Ileocecal junction	1.5	-	≤ 3	ND	-	-	Right hemicolectomy	-	NR (25 mos)
16/2009	Ryan et al [12]	F/15	Rectum	3.7	+	≤ 2	5-10	+	-	Resection of rectum	+	NR (9 mos)
17/2010	Freeman et al [23]	F/17	Sigmoid colon	6.0	+	low	ND	ND	ND	Sigmoid resection	-	NR (180 mos)
18/2010	Park et al [21]	M/7	Ascending colon	4.0	+	low	low	-	-	Right hemicolectomy	+	NR (26 mos)
19/2010	Mitteldorf et al [30]	F/71	Stomach	3.0	-	1	ND	-	-	Partial gastrectomy	-	ND
20/2010	Gross et al [22]	M/5.5	Ascending colon	5.0	+	ND	ND	ND	ND	Resection of theascending colon	-	NR (24 mos)
21/2010	Shi et al [20]	M/36	Descending colon	4.8	-	low	ND	ND	ND	Segmental resection of the bowel	-	NR (32 mos)
22/2010	Shi et al [20]	F/38	Ascending colon	6.0	+	low	ND	-	ND	Segmental resection of the bowel	-	NR (8 mos)
23/2010	Shi et al [20]	M/42	Sigmoid colon	4.5	-	1-2	ND	-	ND	Segmental resection of the bowel	-	NR (15 mos)
24/2010	Shi et al [20]	F/45	Ascending colon	3.5	-	low	ND	-	ND	Segmental resection of the bowel	-	NR (36 mos)
25/2012	Waters et al [7]	M/42	Stomach	10	ND	ND	ND	ND	ND	Distal gastrectomy	-	DOT (3 mos)
26/2012	Unluoglu et al [15]	M/36	Ileum	2.0	ND	1-2	3-4	minimal	ND	partial ileectomy, appendectomy	-	NR (10 mos)
27/2012	Fassan et al [33]	M/75	Esophagus	8.0	ND	3	ND	-	ND	Percutaneous transperitoneal jejunostomy	-	Liver metastasis and DOT (3 mos)
28/2013	Im et al [11]	M/17	Rectum	3.0	-	< 1	5	-	-	Partial rectectomy	-	NR (10 mos)
29/2014	Kanazawa et al [10]	F/55	Rectum	2.5	ND	0	15	-	-	Partial rectectomy	-	NR (15 mos)
30/2014	Pizzi et al [34]	F/8	Peritoneum	8.0	ND	< 2	ND	-	ND	Complete resection	-	NR (8 mos)
31/2015	Lu et al [5]	F/29	Terminaln ileum	13.5	+	3-5	5	+	-	Resection of partial ileum	+	NR (28 mos)
32/2015	Lu et al [5]	F/41	Terminaln ileum	2.2	-	0	< 1	-	-	Partial ileectomy and colectomy	-	NR (39 mos)
33/2015	The present case	F/27	Duodenum	8.2	-	1	2	+	-	Duodenectomy	-	NR (4 mos)

IFB: infiltrative border, LVI: lymphovascular invasion, AC: adjuvant chemotherapy, F: female, M: male, ND: no record, NR: no recurrence, DOT: died of tumor.

epithelium and spindle, Smooth muscle tumors express diffuse cytoplasm eosinophilia, perinuclear vacuoles, and "cigar-shaped" nuclei but PEComas-NOS not. Smooth muscle tumors typically don't show the delicate capillary network seen in many PEComas-NOS. Melanocytic markers, immunohistochemically, are useful tools for differential diagnosis. In dubious cases, a series of immunostains containing HMB45, Melan A, smooth muscle actin, and desmin is needful [4]. Metastatic malignant melanoma (MM) is the most common metastatic tumor of GI tract [35]. While MM and PEComas-NOS can show similar morphology, MM is usually S100 and c-kit positive. Furthermore, MM always has a clinical history of primary tumor of skin [12]. The chromophobe cell type of metastatic renal cell carcinoma (RCC), which expresses epithelioid figures and abundant angiogenesis, should also be recognized as an important differential diagnosis. But the positive for CK, CD117, CD10, RCC antigen and the specificity of CD10, RCC could help distinguish RCC from PEComas-NOS [20]. Paraganglioma whose morphology is similar as that of PEComas-NOS, shows tumor cells of abundant cytoplasm surrounded by vessels and fibrous stroma. But the positive for NSE, CgA, SY and the negative for HMB45 and Melan A could lead to the correct diagnosis of paraganglioma [20].

The diagnosis of GI PEComas-NOS is always made after excision, however, 3 cases were diagnosed before surgery [7, 18, 32]. How to pick the correct diagnosis before surgery is a challenge. Another challenge is that this entity shows a variable spectrum from benign to malignant, which makes a dilemma in management. Given the scarcity of such distinctive tumor, clear criteria for malignant PEComas are yet to be established. Folpe et al concluded that tumor size > 8 cm, mitotic activity greater than 1/50 HPF, and necrosis were strongly associated with recurrence and/or metastasis, proposing that it should be classified as "benign", "uncertain malignant potential" and "malignant" when the tumor with one, two and more than two that features [3]. After studying 35 cases of GI PEComas, Doyle et al came to a conclusion that behavior of metastases were significantly associated with marked atypia, diffuse pleomorphism, and mitoses ≥ 2/10 HPF [9].

The optimal management of GI PEComas-NOS has not been well established. Complete excision is the mostly common measure at present and most cases are treated with laparotomy. There were some cases which tried to resect the neoplasm under endoscopy but failed [26, 28]. Other strategies such as chemotherapy and immunotherapy have been considered or implemented in some malignant cases, but the benefit of such measures is uncertain which calls for larger numbers and longer follow-up cases being discussed.

In conclusion, it should be aware that GI PEComas-NOS is an entity in clinical work, GI PEComas-NOS should be regarded as an important differential diagnosis for undetermined intra-abdominal mass. Surgery plays an important role in its treatment. Because GI PEComas-NOS can behave in a malignant manner, close surveillance with imaging and endoscope is required. Further understanding of this neoplasm may depend on more standard case reports and histopathological studies.

Disclosure of conflict of interest

None.

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