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Sarcomatoid carcinoma of the jejunum with gastric metastases: A case report and review of the literature

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ABSTRACT

INTRODUCTION: Sarcomatoid carcinoma (SCA) of jejunum is an extremely rare condition. To our knowledge, only 17 cases have been reported in the literature.
PRESENTATION: We introduced an additional case of the sarcomatoid carcinoma of jejunum in a 62-year-old Chinese male who presented with epigastric pain for 3 weeks. Multiple tumors originated in the jejunum and metastases to mesentery lymph nodes and distal stomach were found during the laparotomy. The patient underwent palliative resection of the tumors. He died 11 days after the operation.
DISCUSSION: Sarcomatoid carcinoma (SCA) of jejunum is an extremely poor prognostic tumor in human being. The diagnosis of SCA was based on pathological observations and immunohistochemical staining. There is no official treatment guideline for SCA, but wide excision including the tumor is the main goal of treatment.
CONCLUSION: This is the first case of sarcomatoid carcinoma of jejunum with gastric metastases being reported and also the shortest survival period after the operation. Surgery is the cornerstones of treatment but the ideal means is still unknown due to the short survival and inadequate reports.

1. Introduction

Sarcomatoid carcinoma (SCA) of jejunum is an extremely rare biphasic tumor characterized by a combination of malignant epithelial and mesenchymal cells [1–3]. Carcinosarcoma, pleomorphic carcinoma and anaplastic giant-cell carcinoma have been previously used to describe this kind of tumor. Nowadays, SCA became the most accepted term used in diagnostic surgical reports [2,3]. To our knowledge, only 17 cases of primary SCA of the jejunum have been reported in the literature to date [4]. The tumor is known to be more malignant than other small intestine cancers [5], and patients with this disease have a significantly worse prognosis due to the tumor's metastatic nature and aggressive clinical course. We report an additional case (18th case) of primary SCA of the jejunum in a 60-year-old Chinese male. He received laparotomy and palliative resection of the tumors. However, he died 11 days after the operation.

2. Case presentation

A 60-year-old Chinese male with no medical morbidity in the past presented with epigastric pain for three weeks. He was a heavy smoker with two packs a day, quitted for two months; and alcohol drinking with two bottles every day for several decades, quitted for several months. Abdominal fullness, general malaise and poor digestion took place in the meantime. He visited several local clinics vainly and the symptoms got worse. Our upper gastrointestinal endoscopy displayed a protruding nodular mass in a 2 cm deep dirty based gastric ulcer at lesser curvature side of low body of stomach. Malignancy or gastrointestinal stromal tumor (GIST) was suspected and biopsy was performed. Moreover, colonoscopy showed a 1.5 cm pedunculated polyp at 40 cm from anal verge and polypectomy was executed. Laboratory data including blood routine, biochemistry and tumor markers were within normal limits except mild anemia (hemoglobin (Hb) 10.4 g/dL) and hypoalbuminemia (2.4 g/dL).

Blood loss was noticed after one week of his admission (Hb: 7.8 g/dL). Chest X-ray showed a right upper lobe soft tissue opacity and further computed tomography detected a $5.3 \times 5 \times 6.7$ cm lung cancer over right upper lobe with adrenal metastases, cT2bN0M1b; mucosal wall thickening at lesser curvature of lower body of stomach; ileus of bowel loops; a $5.72 \times 7.54 \times 7.03$ cm well defined heterogeneous cystic mass with peripheral contrast enhancement in the left adrenal gland; and multiple gall stones. The pathology reports of stomach biopsy and colon polypectomy showed adenocarcinoma and tubulovillous adenoma respectively. Gastric cancer and lung cancer were impressed. Surgical treatment for abdomen was first considered in priority before treatment for lung condition.

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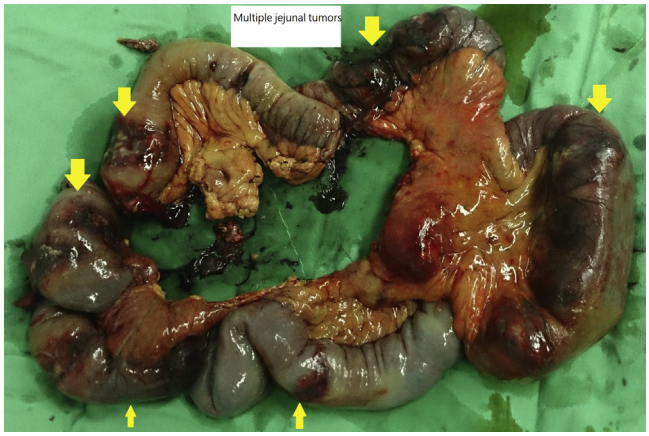

Fig. 1. Multiple tumor masses in the jejunum (yellow arrows).

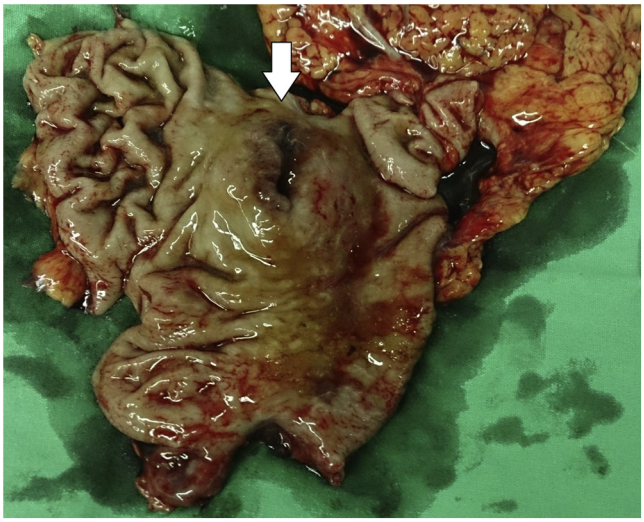

Fig. 2. 3 cm Ulcerative tumor in the lesser curvature side of distal stomach at angularis (white arrow).

The operation findings during the exploratory laparotomy included: a 3 cm ulcerative tumor in the lesser curvature side of distal stomach at angularis (Fig. 1); multiple tumor masses in the jejunum (Fig. 2), lymph nodes enlargement in the jejunal mesentery and at the 4th portion of duodenal mesentery, and multiple gall stones in the gall bladder. We performed hemigastrectomy, excision of a long segment of jejunum, gastrojejunostomy and open cholecystectomy. The initial post-operative course was smooth. However, gastroparesis occurred and parenteral nutrition support was given. Sudden onset of apnea and cardiac arrest occurred and the patient had no response to our resuscitation effort. Finally the patient died 11 days after the operation.

The pathology report showed poorly differentiated sarcomatoid carcinoma of jejunum (5 in submucosa and one infiltrated to mesointestine, largest 5 cm in circumference), one of the lymph node contained tumor cells, and metastatic to stomach. The main tumors in jejunum invaded to the visceral peritoneum with discontinuous extramural extension of tumor deposits, present of lymphovascular invasion and macroscopic tumor perforation. The immunohistochemistry showed CK(+), vimentin(+), P63(−), S100(−), CD117(−), CD34(−), SMA(−), CD30(−), EMA(−), Bc12(−), CD21(−), CD1a(−), CK7(−), CD20(−), WT-1(−) and calretinin(−) (Fig. 3). Our final diagnosis was jejunual sarcomatoid carcinoma with stomach, jejunum, mesenteric lymph nodes metastasis, pT(m)3N1M1, stage IV, and right upper lobe lung cancer with adrenal metastases, cT2bN0M1b.

3. Discussion

Adenocarcinoma, neuroendocrine tumors, sarcomas and lymphomas are the four most common malignant tumors arising in the small intestine [6]. Primary SCA of small bowel is rare and most often it occurred in ileum, followed by jejunum and duodenum [2]. To our knowledge, there were 17 cases of jejunal SCA reported in the literature to date (Table 1) but the real incidence rate may be underestimated. These tumors normally affect middle-aged to elderly patients with a mean age at presentation of 57 years (range, 35–85 years). Reid-Nicholson et al. reported that SCA of the small intestine is more prevalent in male patients, with a male:female ratio of 1.5:1.0 [2], which is different from the recent report from Zhang B et al. showing no significant difference with a male:female ratio of 0.89:1.0 [4]. Most SCAs of the jejunum are typically single tumors, and only 2 patients exhibited multifocal primary tumors [7]. Most of the reported cases presented with metastases (13/18) and most cases died rapidly after the diagnosis. Our case is an additional (the 3rd case) multifocal tumor of SCA in jejunum, and also the first case with stomach metastases. The late presentation and diagnosis can explain the poor prognosis of our patient and we felt distressed with his ultrashort survival since he died 11 days after surgery, which is the shortest one presented in the literature. However, we guess there is a portion of patients died before any definite diagnosis can be made because of its rapid progression and metastases, which leads to under presentation of this kind of cancer.

The risk factors of SCA are unknown, but in certain publications, a correlation with long-standing regional enteritis has been referenced [8,9]. Zhang et al. [4] listed the complete clinical and pathological data (Table 2) in his report earlier. Anemia was the

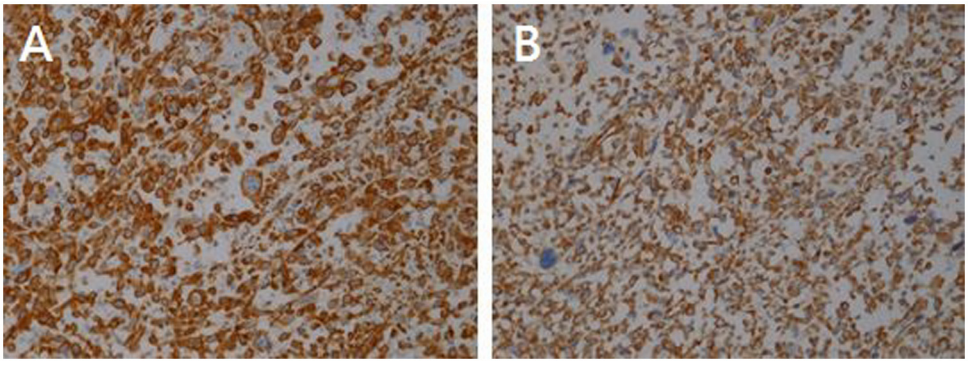

Fig. 3. Positive immunohistological stain for (A) CK and (B) vimentin.

Table 1
Review of published cases of sarcomatoid carcinoma (SCA) of the jejunum. (Modified from Zhang et al. [4]).

Source year	Age, years /gender	Diagnostic term	Size, cm	Morphology	Immunohistochemistry				Presence of metastases	Death after diagnosis, mo
					Vimentin	CK	EMA	CEA		
Robbey-Cafferty et al. [5]	38/F	Anasplastic and SCA	16	Endophytic	N/A	+	N/A	+	Yes	8
	48/F	Anasplastic and SCA	6	Endophytic	N/A	+	N/A	+	Yes	29
	65/M	Anasplastic and SCA	5	Endophytic	N/A	+	N/A	+	Yes	5
	35/F	Anasplastic and SCA	7.5	Endophytic	N/A	-	N/A	-	Yes	36
Bak and Teglbjaery [7]	52/F	Pleomorphic CA	8	N/A	+	+	+	N/A	Yes	7
	56/M	Pleomorphic CA	8	N/A	+	+	+	N/A	Yes	8
Lam et al. [10]	76/F	SCA	8	N/A	+	+	+	N/A	No	2
Tsukadaira et al. [11]	56/M	SCA	9.2	Endophytic	+	+	+	-	Yes	3
Reid-Nicholson et al. [2]	55/M	SCA	7.5	Polypoid	+	+	+	-	Yes	11
Moriwaki and Sugiyama [12]	55/M	SCA	N/A	N/A	N/A	+	+	N/A	N/A	9.4
Yucel et al. [13]	51/F	SCA	8	Polypoid	+	+	N/A	N/A	Yes	1.9
Pata et al. [14]	85/F	SCA	3	Polypoid	N/A	+	N/A	N/A	No	3
Han et al. [15] 2013	70/F	SCA	N/A	N/A	+++	+	-	N/A	No	N/A
Kang et al. [16]	78/M	SCA	5	Endophytic	+++	+	N/A	N/A	N/A	N/A
Alfonso Puentes et al. [17]	56/F	SCA	10	N/A	+	+	+	N/A	Yes	6
Lee et al. [3] 2015	67/M	SCA	10	N/A	+++	+	N/A	-	Yes	1.4
Zhang et al. [4]	62/M	SCA	12	Endophytic	+++	+	+	-	Yes	1
Present case, 2016	60/M	SCA	3.6	Endophytic	+	+	-	N/A	Yes	0.36

F, female; M, male; CK, cytokeratin; EMA, epithelial membrane antigen; CEA, carcinoembryonic antigen; mo, month; N/A, not applicable; CA, carcinoma.

Table 2
Summary of clinicopathological characteristics of patients with sarcomatoid carcinoma (SCA) of the jejunum. Modified from Zhang et al. [4].

Characteristics	No. of patients (%)
Clinical presentation (n = 14)	
Anemia	8 (57.1)
Abdominal pain	6 (42.9)
Abdominal mass	4 (28.6)
Gastrointestinal bleeding	4 (28.6)
Weight loss	4 (28.6)
Fatigue	4 (28.6)
Small intestinal obstruction	2 (14.3)
Shortness of breath on effort	2 (14.3)
Palpitations	2 (14.3)
Subphrenic free air	2 (14.3)
Vomiting	2 (14.3)
Fever	1 (7.1)
Diarrhea	1 (7.1)
Dyspepsia	1 (7.1)
Anorexia	1 (7.1)
Greatest dimension of tumor, cm (n = 12)	
<5	1 (8.3)
5–10	9 (75.0)
>10	2 (16.7)
Depth of invasion (n = 12)	
Muscularis propria	1 (8.3)
Subserosal layer	2 (16.7)
Full thickness of the wall	9 (75.0)
Regional lymph nodes (n = 10)	
Metastasis	9 (90.0)
No metastasis	1 (10.0)
Distant metastasis (n = 12)	
Lung	6 (50.0)
Liver	3 (25.0)
Para-aortic lymph nodes	2 (16.7)
Cervical lymph nodes	2 (16.7)
Axillary lymph nodes	1 (8.3)
Pelvic bone	1 (8.3)
Brain	1 (8.3)
Lymphovascular or perineural invasion (n = 11)	
Yes	6 (54.5)
No	5 (45.5)
Extension into adjacent organs (n = 6)	
Small bowel loops	3 (50.0)
Colon	2 (33.3)
Mesentery	2 (33.3)
Omentum	2 (33.3)
Ovary	1 (16.7)

most common symptom, followed by abdominal pain, palpable abdominal mass, gastrointestinal bleeding, weight loss and fatigue. Two patients presented with cancerous bleeding that required emergency laparotomy [11], and another 2 patients experienced tumor perforation with subphrenic free air and acute peritonitis [13,15]. Additionally, shortness of breath on effort, palpitations, vomiting, fever, nausea, diarrhea, dyspepsia and anorexia may also be the initial symptoms of patients with primary SCA of the jejunum. The remarkably non-specific clinical manifestations of jejunal SCA make early diagnosis difficult.

The diagnosis of SCA was based on pathological observations. They are typically endophytic or polypoid masses with central ulceration. Most tumors involve the entire wall of the jejunum with regional lymph node metastasis, and directly invaded neighboring structures. Moreover, prominent lymphovascular or perineural invasion was found in 54.5% of the patients. The most common metastatic locations are the lung, distant lymph nodes and liver, while the brain and pelvic bones may also be involved. All the 18 cases reported (including our patient) succumbed to the disease, and more than 80% had metastatic or recurrent disease at death, indicating the aggressive and metastatic nature of this tumor. The overall survival time after diagnosis is only 8.25 months (range, 0–36 months, including the present case).

Immunohistochemical staining also contributes to the diagnosis. SCA may exhibit a monophasic or biphasic pattern [5]. A predominance of the mesenchymal-like component, with minimal to absent epithelioid areas, characterizes the typical monophasic pattern, whereas biphasic tumors display a mixture of epithelial-like and mesenchymal-like cells. In the majority of SCA, epithelial- and mesenchymal-like components exhibit positivity for EMA. In addition, ~90% of intestinal sarcomatoid carcinomas are positive for vimentin. Certain cases may also exhibit focal positivity for neuroendocrine and neuron-specific markers [2]. Han et al. reported a suggestion of relatively favorable prognosis with the presentation of low positivity for Ki-67 in the tumor [15]. Our immunohistochemical staining results revealed positive expression of vimentin and CK, indicating that the sarcomatous component displayed epithelial and mesenchymal differentiation and SCA was impressed.

There is no official treatment guideline for SCA, but wide excision including the tumor is the main goal of treatment. There are cases in which adjuvant chemotherapy using 5-FU and/or cisplatin and radiotherapy were administered, but no report identified

improvements in survival. Recent clinical trials using monoclonal antibodies targeting immune checkpoint programmed death protein I (PD-1) and its ligand, PD-L1, reported promising antitumor activity in several malignancies. Upregulated PD-L1 expression on tumor cells is considered to mediate immune evasion via activation of the PD-1/PD-L1 pathway and suppression of effector immune responses [18]. B Zhang et al. discovered a distinct expression of the PD-L1 protein in primary SCA of the jejunum [4], indicating the potential of PD-L1–targeted immunotherapy for the treatment of this type of cancer.

4. Conclusion

Sarcomatoid carcinoma of jejunum is an aggressive neoplasm with metastatic nature and extremely poor prognosis. Surgery is the cornerstones of treatment but the ideal means is still unknown due to the short survival and inadequate reports. Early detection may improve the prognosis.

Conflicts of interest

None.

Funding

None.

Ethical approval

None.

Consent

Written informed consent was obtained from the patient's family for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Ching-Ming Kwok is the only author in this case report.

Guarantor

The author is the only person for the responsibility of this report

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