CASE REPORT

Primary sarcomatoid carcinoma of the small intestine: very rare and aggressive tumour

Peter Abotaga Andrawes,^{1,2} Masood Shariff,¹ Qing Chang,³ Roman Grinberg¹

¹Department of Surgery, Staten Island University Hospital, Northwell Health System, Hofstra School of Medicine, Staten Island, New York, USA

²Department of Minimally Invasive Surgery, Staten Island University Hospital, Northwell Health System, Hofstra School of Medicine, Staten Island, New York, USA

³Department of Pathology, Staten Island University Hospital, Northwell Health System, Hofstra School of Medicine, Staten Island, New York, USA

Correspondence to

Dr Peter Abotaga Andrawes, pabotaga@gmail.com

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SUMMARY

Sarcomatoid carcinoma of the small intestine is a very rare and aggressive variant of small intestinal cancers with poor prognosis. The tumour primarily affects middle-aged and older patients with a mean age of 57 years at the time of presentation. We report a woman aged 58 years without any relevant medical history who presented with small intestinal obstruction. She underwent radiologic and endoscopy investigation with persistent features of small bowel obstruction. The patient was found to have a small bowel tumour causing the obstruction and underwent surgical excision of the tumour. Pathology revealed malignant neoplasm with sarcomatoid and epithelioid features involving the terminal ileum. The use of immunohistochemical markers helps in wide range of differential diagnoses. Surgical resection is still considered the best and first-line therapy with poor response to chemotherapy and radiotherapy.

CASE PRESENTATION

A woman aged 58 years with medical history of hypertension and history of hysterectomy presented to the emergency department with diffuse abdominal pain, nausea and vomiting. The symptoms had been persistent for 1 week. Her last bowel movement was on the day of presentation but denied any flatus.

On physical examination, the abdomen was distended, lower abdominal tenderness and positive bowel sounds. CT scan showed dilated loops of the small intestine measuring up to 3.3 cm with trace interloop ascites with a transition point in the right lower quadrant. Laboratory work showed white cell count of 19 000 with unremarkable basic metabolic panel. The patient was admitted and was treated non-operatively with presumed diagnosis of adhesive small bowel obstruction, given her history of hysterectomy. The patient's CT scan of the abdomen and pelvis showed air within the colon which was compatible with incomplete obstruction. Within 24 hours of non-operative management and GI decompression, the patient improved clinically. On physical examination, the abdomen was soft, non-distended and non-tender. Her WCCs was 8. The patient had bowel movement, passed flatus, was started on diet and was discharged on HD#6.

BACKGROUND

Primary small intestinal carcinoma is rare with incidence of 0.5–0.8 per 100 000 population per year.^{1–2} Sarcomatoid carcinoma of the small intestine is a very rare and aggressive variant of small intestinal carcinoma with a poor prognosis.^{1–9} This subtype of carcinoma contains mesenchymal-like spindle cells and carcinomatoid features. We report a case of woman aged 58 years with sarcomatoid tumour at the terminal ileum causing small intestinal obstruction. We also reviewed all the other reported cases with sarcomatoid carcinoma of the small intestine, their characteristics and the presence or absence of metastasis.

INVESTIGATIONS

The patient underwent an outpatient follow-up and continued to have mild lower abdominal pain. The patient underwent gastrointestinal (GI) series which showed irregularity of the small bowel inferior to the caecum with findings suggestive of small intestinal wall thickening (figure 1A,B). Magnetic resonance (MR) enterography showed circumferential

Figure 1 Small bowel series showing: irregularity of the small bowel inferior to the caecum (blue arrow).

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wall thickening with luminal narrowing of 4.3 cm segment of the distal ileum, with marked wall enhancement, and surrounding mesenteric stranding suspicious for neoplasm (figure 2A–D). Colonoscopy was performed, the terminal ileum was intubated, no mass was visualised, but it appeared to be narrowed and hence biopsies were performed which showed normal small bowel mucosa and focal increased eosinophils in the lamina propria.

Metastatic work up: The patient underwent CT scan of the abdomen and pelvis when she was admitted, which showed no evidence of metastatic disease. Chest X-ray showed no evidence of cardiopulmonary disease or metastatic disease.

DIFFERENTIAL DIAGNOSIS

Adenocarcinoma, neuroendocrine tumours, sarcomas and lymphomas are the four most common malignancies of the small intestine and the highest in differential diagnosis.⁸ The use of immunohistochemical markers helps in a wide range of differential diagnoses which include epithelioid angiosarcoma, leiomyosarcoma, epithelioid malignant peripheral nerve sheath tumour (MPNST), gastrointestinal stromal tumours (GIST), lymphoma and melanoma.

TREATMENT

The patient underwent robotic-assisted right hemicolectomy with resection of the terminal ileum and ileocolic anastomosis. Her diet was resumed on postoperative day (POD) 1 and she was discharged to home on POD 6.

OUTCOME AND FOLLOW-UP

Pathology revealed a malignant neoplasm with sarcomatoid and epithelioid features involving the terminal ileum. The ileal mucosa overlying the tumour was not dysplastic and showed no evidence of polypoid precursor. Markers of epithelial origin, including Pankeratin and CK7, were diffusely positive in the neoplastic cells. Tumour size in greatest dimensions was 3×3×2 cm. The final diagnosis was primary sarcomatoid carcinoma of the terminal ileum. The tumour had invaded through the muscularis propria into the subserosal adipose tissue (pT3) and lymphovascular invasion was also noted. Metastatic tumour presented in 1 out of 8 lymph nodes (pN1). Immunohistochemical stains showed positive pancytokeratin, Vimentin, Ki67 and ck7. Surgical margins were free of tumour (figure 3A–D).

Figure 2 MR enterography showing circumferential wall thickening and luminal narrowing at the distal ileum: (A) for coronal view and (B) for transverse view. (C, D) (yellow arrow) Enlarged, centrally necrotic right lateral caval lymph node measures 2.7×2.8×3.7 cm.

Figure 3 Microscopically, the tumour was composed of spindle cells forming interlacing fascia (A) and sheets of epithelioid cells with oval vesicular nuclei, prominent nucleoli and pale eosinophilic to clear cytoplasm (B). The tumour cells are positive for vimentin (C) and pancytokeratin (D).

At 6 months follow-up, the patient is doing well. She was started on chemotherapy with no evidence of metastasis to date.

DISCUSSION

Sarcomatoid carcinoma is extremely rare type of small intestinal carcinoma which displays carcinomatous and sarcomatous features.⁹ The tumour was first described by Dikman and Toker in 1973 and named it enteroblastoma.⁶ This tumour can occur in various anatomical sites, including lungs, thyroid gland, salivary glands, breast, urinary bladder, prostate, skin and digestive system.¹⁻⁸ GI sarcomatoid tumours are rare and they occur more frequently in the stomach, gallbladder and oesophagus the most common locations in the GI tract.¹ According to the literature, small intestinal sarcomatoid carcinomas arise most frequently in the ileum followed by the jejunum and less commonly in the duodenum.⁷⁻⁸ However, after reviewing most of sarcomatoid carcinomas (table 1), we found that the most common location is the jejunum followed by the ileum and only one case of duodenal sarcomatoid carcinoma was reported.¹⁰⁻¹⁹

The tumour primarily affects middle-aged and older patients with a mean age of 57 years at presentation.⁷ Patients usually have different, non-specific symptoms including: abdominal pain, intestinal obstruction, as in our case presentation, palpable abdominal mass, nausea, vomiting, weight loss, GI bleeding and anaemia.³⁻⁷ Zhang *et al* noted that anaemia and abdominal pain have been the two most common presentations.⁸ The most

common site of metastasis is lung followed by lymph nodes, liver, brain and pelvis.

Most cases of small intestine sarcomatoid carcinoma showed polypoid or fungating mass with or without ulceration and frequently have necrotic and haemorrhagic features.¹⁻⁴ The average size is about 7.5 cm with a range of 3–16 cm.⁸ The use of immunohistochemical markers helps in a wide range of differential diagnoses which include epithelioid angiosarcoma, leiomyosarcoma, epithelioid MPNST, GIST, lymphoma and melanoma.³ Lymphovascular invasions are found in 55% of the cases, similar to our case.⁸

Evaluation of primary small bowel tumours is not easy. CT scan findings were described in some cases as symmetric or asymmetric wall thickening with fold enhancement; however, in our case, the CT scan failed to demonstrate the lesions probably due to its small dimensions.¹ Small bowel series has been also used to help with the diagnosis. GI series showed irregularity of the small bowel inferior to the caecum that suggested further work up of the finding. In other case reports, GI series showed intraluminal polypoid mass. Video capsule endoscopy and double-balloon enteroscopy have shown superiority compared with small bowel follow through and CT scan in identifying all small bowel pathologies, especially if an intervention needed, like a biopsy.^{2,5}

Surgical resection is still considered first line of therapy with a poor response to chemotherapy and radiotherapy. After

Table 1 Review of published cases of sarcomatoid carcinoma of the small bowel

Age, years/Sex	Site	Size, cm	Morphology	Symptoms	Metastasis	Immunohistochemistry		Ref no.
						CK	Vimentin	
35/F	Jejunum	7.5	Endophytic	NA	Yes	—	NA	10
38/F	Jejunum	16	Endophytic	NA	Yes	+	NA	10
44/M	Ileum	5	Polypoid and necrotic	Lower GI bleed	Yes	NA	NA	6
45/M	Ileum	14	NA	NA	No	+	+	11
48/F	Jejunum	6	Endophytic	NA	Yes	+	NA	10
51/F	Jejunum	8	Polypoid	Anaemia/abdominal pain	Yes	+	+	12
52/F	Jejunum	8	Nodular	Anaemia/abdominal pain	Yes	+	+	13
53/M	Ileum	13	Polypoid and necrotic tumour	Coffee ground emesis/abdominal pain	Yes	+	+	14
54/F	Ileum	4.5	Endophytic	NA	No	—	NA	10
55/M	Jejunum	7.5	Polypoid necrotic	Lower GI bleeding	Yes	+	+	4
55/M	Jejunum	NA	Ulcerating mass	Anaemia	Yes	+	NA	15
56/F	Jejunum	10	Ulcerating/necrotic	Coffee ground emesis	Yes	+	+	5
56/M	Jejunum	8	Nodular	Weight loss/abdominal pain	Yes	+	+	12
56/M	Ileum	9.2	Fungating ulcer	Anaemia/supraclavicular lymph nodes	Yes	+	+	16
57/M	Ileum	6	NA	NA	No	+	+	11
59/M	Ileum	10	NA	NA	No	NA	NA	4
60/M	Ileum	5	Nodular	Abdominal pain	Yes	+	NA	7
62/M	Ileum	15	Ulcerofungating	Abdominal pain	No	+	+	3
62/M	Jejunum	12	Endophytic	Abdominal pain	Yes	+	+	8
63/M	Ileum	3	NA	NA	No	+	+	11
65/F	Jejunum	5	Endophytic	NA	Yes	+	NA	10
65/M	Ileum	5	NA	NA	Yes	—	NA	10
67/M	Jejunum	10	Polypoid	Vomiting/weight loss	Yes	+	+	3
68/F	Ileum	6	NA	NA	No	NA	NA	17
69/M	Jejunum	6	Polypoid	Melena	No	+	+	2
70/F	Jejunum	NA	Polypoid	Abdominal pain	No	+	+	9
75/M	Ileum	5	NA	NA	No	+	+	17
76/F	Jejunum	8	NA	NA	No	+	+	18
77/M	Duodenum	3	NA	NA	Yes	+	+	17
78/M	Jejunum	5	Exophytic	Abdominal pain	NA	+	+	1
85/F	Jejunum	3	Polypoid	Shortness of breath	No	+	NA	19
58/F	Ileum	3	Polypoid	Intestinal obstruction	NO	+	+	Current Case

CK, cytokeratin; GI, gastrointestinal; NA, not available.

reviewing the literature, 70% of the patients died within 2 months to 3-year follow-up.⁷

Learning points

- ▶ Sarcomatoid carcinoma of the small intestine is rare and aggressive tumour with poor prognosis.
- ▶ Sarcomatoid carcinoma should be considered as one of the differential diagnoses when considering a small intestine tumour as a diagnosis.
- ▶ Wide spectrum of *immunohistochemistry* stains help distinguish different types of small intestinal tumour.
- ▶ Most cases of small intestine sarcomatoid carcinoma were found in the jejunum.
- ▶ Surgery remains the mainstay of treatment since chemotherapy and radiotherapy have not shown improvement in survival.

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