Abdominotransanal resection of a strangulated rectal carcinosarcoma

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Accepted 8 August 2021

SUMMARY

This is a case of a 75-year-old man who presented with a 7-month history of a reducible rectal mass. The patient came to the emergency department with a prolapsed necrotic bowel involving a strangulated segment with the rectal mass. He underwent an abdominotransanal resection through a combined abdominal and perineal approach. His postoperative course was unremarkable. Histopathological and immunohistochemical studies showed a rectal carcinosarcoma. Because of a state-mandated lockdown due to the COVID-19 pandemic, the patient failed to follow-up. He was later seen to have metastatic progression. Owing to the poor functional status of the patient, the shared decision of the multidisciplinary team, the patient and his family was to manage him with palliative intent.

BACKGROUND

Carcinosarcoma is a rare type of mixed mesodermal malignancy consisting of both of an epithelial and a mesenchymal component.[1] When encountered, it usually affects the head and neck region. Very rarely, these tumours may arise from the gastrointestinal tract—with an even lower reported incidence in the more distal segments. Prior to this report, only eight reported cases have fulfilled the criteria for rectal carcinosarcoma. This report documents a case of a 75-year-old man who underwent surgical resection for a strangulated rectal carcinosarcoma.

CASE PRESENTATION

A 75-year-old man came to the emergency department with a prolapsed rectal mass. He had no known comorbidities and had no previous surgery. He presented with a 7-month history of a reducible rectal mass with associated colicky abdominal pain, weight loss, hematochezia and incontinence. Prior to this consult, he was seen by a private physician, and was managed as a case of haemorrhoidal disease without the benefit of a rectal examination or a colonoscopy. Due to the persistence of symptoms, he consulted with another physician who was able to detect the presence of a fungating rectal mass. No biopsy of the mass was performed.

INVESTIGATION

A contrast-enhanced CT scan showed a multilobulated, heterogeneously-enhancing intraluminal mass with internal calcifications in the rectal area measuring 20.8×7.8×9.4 cm that partially herniates through the anal verge. No enlarged lymph nodes and no liver lesions were noted (figure 1).

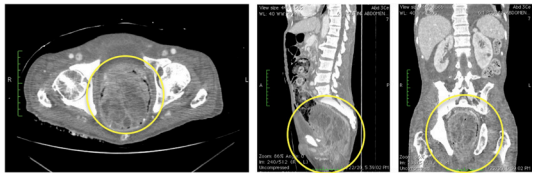

Figure 1 Representative cuts of the abdominal CT scan with triple contrast enhancement showing the multilobulated, heterogeneously-enhancing intraluminal mass with internal calcifications in the rectal area intimately related to the prostate. No liver lesions were noted. Note that this imaging was performed prior to the incarceration of the rectal mass.

Persistent prolapse of the rectal mass prompted consult at our institution. The patient was haemodynamically stable with unremarkable abdominal findings. Rectal examination revealed a 15×10 cm necrotic, foul-smelling prolapsed mass (figure 2).

TREATMENT

The patient was prepared for emergent surgery, for which an abdominotransanal resection (ATAR) with no neorectal construction, using a combined abdominal and perineal approach, was performed. The abdominal phase was started with a midline infraumbilical laparotomy incision that extended to the symphysis pubis. This allowed better exposure for the dissection. A lateral-to-medial mobilisation of the sigmoid colon from the peritoneal reflection, and high ligation of inferior mesenteric

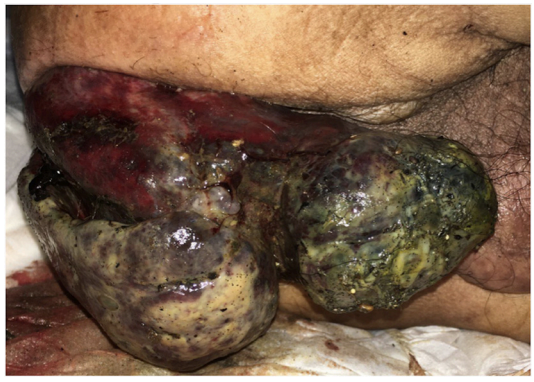

Figure 2 The prolapsed rectal tumour with evidence of necrosis. The involved bowel segment is noted to be oedematous and dusky.

To cite: Manlubatan SIT, Lopez MPJ, Maglangit SACA, *et al*. *BMJ Case Rep* 2021;**14**:e244501. doi:10.1136/bcr-2021-244501

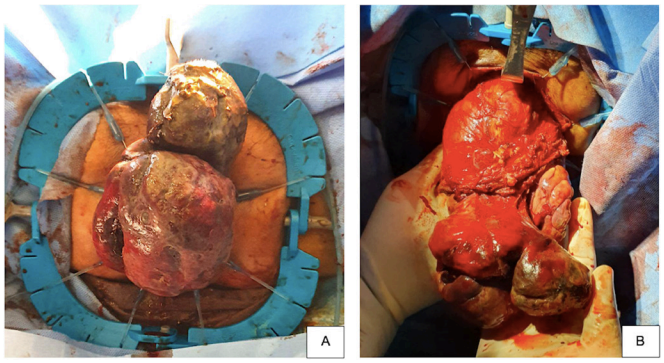

Figure 3 (A) Intraoperative photo showing the involved bowel segment and tumour prior to beginning the perineal phase of the operation. (B) The specimen was delivered through the perineum.

artery and vein were performed, followed by total mesorectal excision (TME). TME involves the removal of the entire rectum, including the surrounding lymphovascular tissues, using sharp dissection. The perineal phase was performed starting with a circumferential incision at the mucocutaneous junction of the anal opening. Total intersphincteric resection then followed, which was performed by circumferentially dissecting between the external anal sphincter and the internal anal sphincter. This resulted in the inclusion of the internal anal sphincter in the resection of the tumour, while preserving the external anal sphincter. The mass was delivered through the perineal incision, and a permanent stoma was created.

Intraoperatively, an irreducible fungating rectal mass was seen prolapsing distally beyond the anal verge, measuring 23.5 cm in greatest diameter (figures 3 and 4). There were areas of necrosis and foul-smelling discharge. Further exploration showed enlarged mesenteric nodes along the inferior mesenteric vessels, dilated proximal bowel loops and serous pelvic ascites. No signs of distant spread were noted. The postoperative course was uneventful.

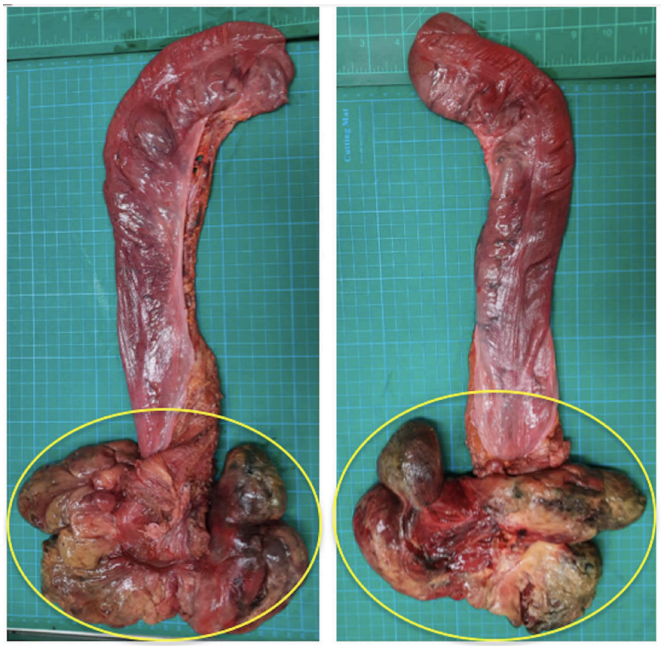

Figure 4 The specimen composed of the distal sigmoid, rectum and the prolapsed tumour (yellow circle).

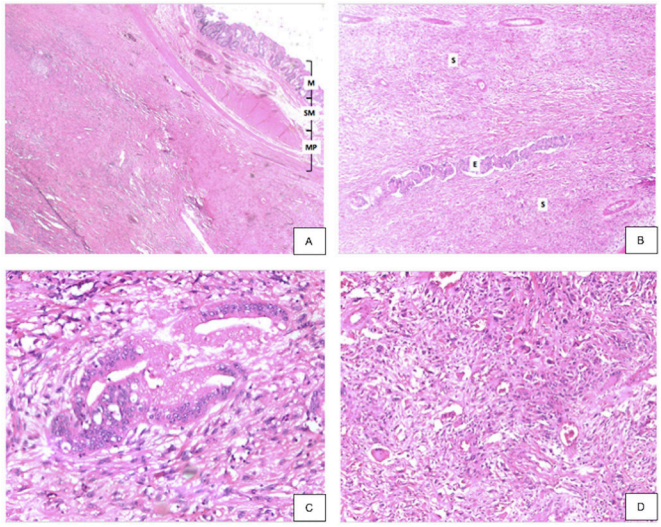

Figure 5 Histopathological features of the mass showing high-grade malignancy. (A) The mass is located at the level of the MP (H&E, 20×). It is predominantly composed of spindle cells. (B) The tumour is composed of a biphasic population of epithelial elements overwhelmed by a malignant epithelial elements (H&E, 100×). (C) High power magnification shows malignant glands dispersed and overwhelmed by the malignant stromal component (H&E, 400×). (D) High power magnification of the stromal component reveals a high grade undifferentiated sarcomatous component with marked nuclear atypia and presence of bizarre multinucleated giant cells (H&E, 200×). E, epithelial carcinoma component; M, mucosa; MP, muscularis propria; S, sarcomatous stromal component; SM, submucosa.

Histopathology revealed a high-grade malignancy for which the primary consideration was a carcinosarcoma, involving the submucosa and perirectal fat, with associated lymphovascular space invasion (figure 5). All 19 pericolic and pararectal lymph nodes; the proximal, distal and radial margins of resection; and the peritoneal fluid collected were negative for malignant cells. Immunohistochemical (IHC) testing confirmed the diagnosis of rectal carcinosarcoma (ie, cytokeratin (CK) AE1/AE3 positive in the epithelial cells, and vimentin positive in the mesenchymal cells) (figure 6).

OUTCOME AND FOLLOW-UP

Due to the delay in adjuvant treatment brought about by the COVID-19 pandemic, the patient developed metastatic spread to the lungs, retroperitoneal nodes and bones. Owing to the poor functional status of the patient, the shared decision of the multidisciplinary team (MDT), the patient and his family was to

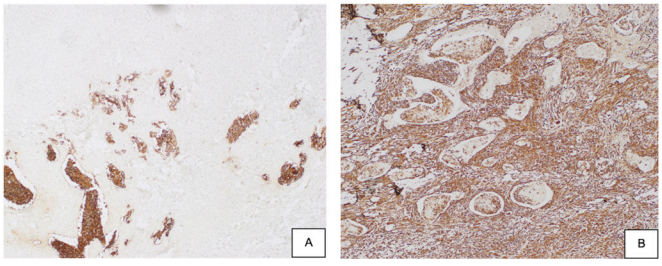

Figure 6 (A) Immunohistochemical studies showed reactivity to cytokeratin in the epithelial cells, but not in the mesenchymal cells (cytokeratin AE1/AE3, 40×). (B) Vimentin is expressed by both the sarcomatous and carcinomatous component (vimentin 40×).

Table 1 Summary of the documented cases of rectal carcinosarcoma

Study	Age/sex	Chief complaint	Onset of symptoms	Distance from anal verge	Size of the tumour	Metastasis	Surgery	Lymph node involvement	Neoadjuvant or adjuvant treatment	Immunohistochemical studies	Recurrence	Survival
Roncaroli *et al*[13]	71/F	Abdominal pain	2 months	2 cm	8 cm	N	Anorectal resection with colostomy	Y	Preoperative RT	Epithelial: cytokeratin and chromogranin Mesenchymal: vimentin, desmin and actin	Pelvis after 2 months	6 months
Takeyoshi *et al*[14]	82/M	Rectal bleeding	Not reported	2 cm	Not reported	N	LAR	N	N	Epithelial: CEA Mesenchymal: vimentin	Skin after 1 month	6 months
Ishida *et al*[6]	80/F	Abdominal pain	Not reported	Not reported	18 cm	N	Hartmann procedure en bloc ileal resection-anastomosis	Y	N	Epithelial: cytokeratin and CEA Mesenchymal: desmin, myoglobin, HHF35 and S-100	Pelvis after 5 months	6 months
Ozturk *et al*[5]	65/F	Rectal bleeding	3 months	2.2 cm	2 cm	N	APR	N	N	Epithelial: cytokeratin Mesenchymal: vimentin and S-100	N	5 years
Tsekouras *et al*[2]	60/M	Constipation	Few weeks	7 cm	7 cm	N	APR	Y	Postoperative chemoRT	Epithelial: CAM, MNF and p53 Mesenchymal: vimentin and actin	Lungs, liver, peritoneum, pelvis and inguinal node after 4 months	6 months
Jeong *et al*[12]	13/F	Abdominal pain	3 weeks	Not reported	13 cm	N	LAR	Y	Postoperative chemotherapy	Epithelial: cytokeratin, epithelial membrane antigen and CEA Mesenchymal: vimentin	–	–
Kolodziejczak *et al*[15]	83/M	Rectal bleeding	Not reported	Not reported	5.3 cm	N	R2 resection of tumour with sigmoid loop colostomy	N	N	Epithelial: cytokeratin Mesenchymal: vimentin and SMA	R2 resection only	5 weeks
Sudlow *et al*[10]	80/F	Rectal bleeding	4 weeks	4.2 cm	Not reported	N	Laparoscopic-assisted APR	Y	Preoperative RT	Epithelial: cytokeratin and CEA Mesenchymal: vimentin, SMA and desmin	Pelvis, lungs after 16 months	25 months
Manlubatan *et al* (2021)	75/M	Rectal mass	7 months	At the verge	23.5 cm	N	Abdominotransanal resection	Y	N	Epithelial: cytokeratin Mesenchymal: vimentin	Lung, nodes and bone after 9 months	–

APR, Abdominoperineal resection; CEA, carcinoembryonic antigen; chemoRT, chemoradiotherapy; LAR, Low anterior resection; N, no; RT, radiotherapy; SMA, smooth muscle actin; Y, yes.

render palliative care. He underwent radiotherapy to the pelvic bones and lumbar vertebrae, as a means of controlling pain.

DISCUSSION

First described by Virchow in 1864, carcinosarcoma is a rare mixed mesodermal malignancy that consists of both glandular epithelial (carcinoma) and mesenchymal or connective tissue (sarcoma) components.[1 2] In literature, the most common sites of occurrence are the head and neck regions and the female urogenital system.[3] When seen in the gastrointestinal tract, it is most commonly seen in the oesophagus, and very rarely in the colorectum.[4] It does appear that the more distal its location is in the gastrointestinal tract, the less frequently is it encountered.[5] Among published case reports prior to 2021, a total of eight cases of rectal carcinosarcoma were described (table 1).

In a review of these cases, carcinosarcoma within the rectum was found mostly among the older population (mean age 67). Only one paediatric case has been reported to this date.

The malignant behaviour of carcinosarcoma is similar to adenocarcinoma with its tendency to metastasise to lymph nodes and distant sites.[6 7] The sites of metastasis identified among the previous studies include the liver, lymph nodes, peritoneum, lungs and skin. Note that in the previous case reports, none of the patients had distant metastases on detection of the primary tumour. Similarly, chest CT scan performed for this patient did not show any pulmonary metastasis.

Clinical signs and symptoms of colorectal carcinosarcoma are related to obstruction and bleeding depending on the localisation, similar to other colorectal malignancies.[5 8 9] These signs and symptoms manifest late, and approximately 50% of patients with colorectal carcinosarcoma present with obstructive symptoms.[9]

Prior to consultation at our institution, this patient was seen months ago by a primary care physician, and was managed as a case of haemorrhoidal disease due to the aforementioned signs and symptoms without the benefit of a rectal examination and colonoscopy. For any patient presenting with abdominal or colorectal symptoms, proper physical examination is essential; supported by appropriate diagnostic modalities, as indicated, such as colonoscopy. Screening colonoscopy should be started as early as 50 years of age for the general population.

Histopathological evaluation of the tumour is crucial in establishing the diagnosis. This shows a biphasic population composed of both malignant stromal and epithelial nature. The mesenchymal stromal component is composed of high grade pleiomorphic spindle to epithelioid cells with high grade nuclear atypia and bizarre giant cells. There are no heterologous components identified. Malignant glands with nuclear atypia are found randomly dispersed into the sarcomatous component. There is no transitional area appreciated between the two populations.

Immunohistochemistry remains the gold standard for diagnosing carcinosarcoma. The two key features are differential staining of the distinct components (ie, positive IHC staining for both CKs and vimentin), and the lack of staining for epithelial markers in the sarcomatous component.[2 10] The carcinomatous component commonly shows reactivity to CK20 and carcinoembryonic antigen, whereas the sarcomatous component stains positively for vimentin, desmin, and smooth muscle actin.[10] The biphasic component mentioned was further emphasised by the IHC studies, with CK AE1/AE3 highlighting only the carcinomatous cell population. Further IHC studies with mesenchymal markers, including H-caldesmon, DOG1, S100, SALL4 and CD31 do not impart a specific lineage differentiation for the sarcomatous component (figure 7). With the following features, a diagnosis of carcinosarcoma was rendered.

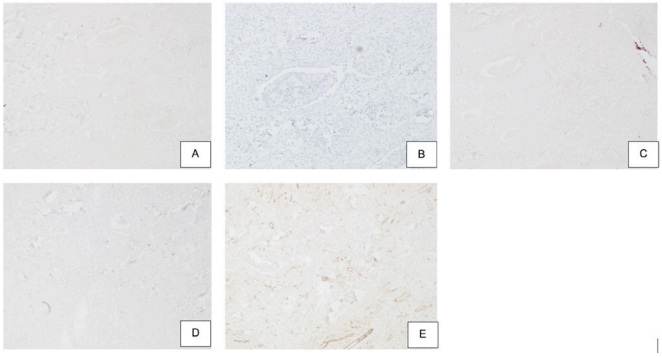

Figure 7 Further immunohistochemical evaluation shows no specific lineage differentiation for the sarcomatous component of the tumour. (A) Caldesmon, (B) DOG1, (C) S100, (D) SALL4, and (E) CD31.

The nomenclature for this diagnosis has been constantly changing, with 'carcinoma with sarcomatoid component' being the closest term in the latest edition of the WHO Classification of Tumours.[11] However, with the prominence of the sarcomatous component in this tumour, it is the pathologist's decision to retain the title of 'carcinosarcoma' as the more appropriate diagnosis.

No treatment guidelines have been established due to the rarity of this condition, but case reports have suggested that it should be treated in a similar way to the more common rectal adenocarcinoma.[12] However, long-term benefits of neoadjuvant and adjuvant chemoradiotherapy have not yet been proven.[10] Of the eight reported rectal carcinosarcoma cases, two underwent neoadjuvant radiotherapy, one of which had recurrence, the other one had none. Two other patients underwent adjuvant treatment, one of which had documented recurrence.

For this patient, since there were signs and symptoms of bowel necrosis, he was brought to the operating room for an emergency procedure. An ATAR with no neorectal construction was performed.

Learning points

- Rectal carcinosarcoma is a rare entity that consists of both epithelial (carcinoma) and mesenchymal (sarcoma) components. Immunohistochemical study is the gold standard to diagnose this disease.
- Meticulous history-taking and thorough physical examination are imperative to avoid misdiagnosis of this rare and aggressive tumour.
- For the general population, screening colonoscopy should be started as early as 50 years of age. For any patient presenting with gastrointestinal symptoms, endoscopy should be performed, as indicated, to allow for early detection and timely management of any pathology.
- No standardised treatment guidelines have been established, but reports have suggested that it should be managed in a similar way to a rectal adenocarcinoma. Further investigation is needed to effectively manage this disease.
- Due to the aggressive nature of this disease, strict follow-up and close monitoring is highly encouraged. Likewise, a multidisciplinary team approach is important to render the best care for the patient.

This technique has better outcomes in terms of healing and function compared with an abdominoperineal resection, where both the external and internal anal sphincters are removed. Had this been detected and diagnosed earlier, neoadjuvant therapy could have been offered as part of an investigational treatment option to decrease the size of the tumour prior to surgery. The collaborative efforts of a MDT, including colorectal surgeons, medical oncologists and radiation oncologists, is vital in planning the best strategy for this kind of patients.

In the limited case reports available, survival of patients with rectal carcinosarcoma ranged from 5 weeks to 5 years. Prognostic factors have not been clearly identified, but some of the indicators that were used include the size of the mass, lymphatic or vascular invasion, stage and the histology of the carcinomatous component, whether low, mid or high grade.[5 12] Due to the aggressive nature of this disease, strict follow-up and close monitoring is recommended.[10 12]

Contributors SITM is the primary author of the report, and assisted during the operation. MPJL performed the operation on the patient, and is one of the coauthors. SACAM and GMO are the coauthors of the report.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent for publication Parental/guardian consent obtained.

Provenance and peer review Not commissioned; externally peer-reviewed.

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