Diverticulum of the rectum due to a rectal leiomyosarcoma

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Summary: A 78 year old woman with a rectal leiomyosarcoma is presented. The case is of interest because of very unusual radiological and operative features of a large rectal diverticulum. As a result of the difficulty in making the correct diagnosis pre- or intraoperatively, a simple, but possibly suboptimal, resection was performed. Although the patient has done well, the long-term outlook is uncertain. The problems of optimum management and prediction of outcome in this uncommon condition are discussed.

Introduction

Rectal leiomyosarcomas are rare. A patient with a rectal leiomyosarcoma, presenting late and with very unusual radiological and intraoperative features of a large rectal diverticulum is described, and the problems of optimum management and prediction of outcome are discussed.

Case report

A 78 year old woman presented with bright red rectal bleeding. She confessed to having had episodes of rectal blood loss for 3 months and she had noticed a sensation of incomplete bowel evacuation. The volume of blood loss had increased dramatically for 4 days prior to hospitalization and she was opening her bowels approximately 6 times per day, passing unaltered blood on each occasion.

On examination, she was thin with atrial fibrillation and breathless with signs of severe obstructive airways disease. Blood pressure was 130/80 mmHg. Temperature was elevated at 38.5°C. Abdominal examination revealed guarding and a fullness in the left lower quadrant. There was no distension and bowel sounds were normal. Digital examination of the rectum confirmed fresh bleeding and a firm mass in the pelvis causing extrinsic compression on the rectum. There was no obvious mucosal abnormality on sigmoidoscopy.

Further investigation yielded normal haemato-

logical and biochemical indices. Ultrasonography demonstrated the pelvic mass which was interpreted as a bulky uterus containing fibroids. Barium enema examination showed a large cavity communicating with the mid part of rectum. There was no other colonic abnormality. The cavity contained gas and solid material, and was interpreted as an abscess cavity or a large, solitary rectal diverticulum (see Figure 1).

At laparotomy, the large, cystic lesion was found occupying most of the pelvis and lower abdomen. It was attached to and apparently arising from the upper rectum by only a narrow 'stalk'. It was excised with a thin cuff of rectum around the base of the 'stalk' leaving most of the rectum undisturbed (a 'diverticulectomy').

Pathological examination of the 9 cm diameter pararectal lump confirmed its cystic nature. It was filled with necrotic material. Microscopy showed the necrotic cavity to be within the muscle layer of the bowel wall. It had a well defined, densely fibrotic pseudocapsule and was lined by an anaplastic malignant tumour with both sarcomatous and epithelioid elements. Immunohistochemical studies demonstrated vimentin and desmin, therefore suggesting an epithelioid type leiomyosarcoma arising in the muscle layer of the rectum. There was no evidence of malignancy in the 'stalk' of the diverticulum or in the cuff of rectum.

The patient made an uneventful recovery from surgery and she remains well at 9 months follow-up. She has no evidence of disease recurrence either clinically, or radiologically with barium enema and computed tomographic (CT) scan studies.

Figure 1 Barium enema showing large diverticulum of rectum containing gas and solid material.

Discussion

Rectal leiomyosarcomas are rare. Smooth muscle tumours constitute less than 1% of all gastrointestinal neoplasms.¹ The majority, 50–80%, of these affect the stomach and small bowel, with only 7–12% occurring in the large bowel.^{1–4} The remainder affect the oesophagus. Of the colorectal smooth muscle tumours, over two thirds occur in the rectum,^{1–3} and about two thirds are malignant.^{2–4} Stated another way, leiomyosarcomas form less than 0.1% of colorectal malignancies.⁵

Rectal leiomyosarcomas tend to occur in the fifth and sixth decades of life,^{5,6} roughly a decade before the peak incidence for rectal adenocarcinomas. Rectal leiomyosarcomas have also been described in children and infants⁷ though these are very rare. There is a male sex preponderance reported in most series, except for the equal sex distribution reported by Evans.³

The usual presentation of rectal leiomyosarcoma is with rectal bleeding and alteration in bowel habit, as in this case. Less common features include intestinal obstruction, intussusception and perforation. It appears that infants are more likely than adults to present with these complications.⁷

Rectal leiomyosarcomas can usually be demonstrated on endoscopy or with barium contrast studies. However, these modalities may fail to detect the tumour because leiomyosarcomas tend to grow extraluminally, and thus CT scanning may be required to demonstrate the extent of the tumour.⁸

To our knowledge, this is the first case in which a rectal leiomyosarcoma had the radiological features of a rectal diverticulum. It can be postulated that the large tumour outgrew its blood supply and underwent central necrosis. The rectal wall

between the bowel lumen and the central cavity of the tumour then broke down allowing direct communication between the two spaces, and explaining the barium enema appearance of a large rectal diverticulum. Alternatively, the tumour may have arisen within the wall of a diverticulum – this explanation is less likely as there was no diverticular disease elsewhere and diverticula contain little muscle.

The operative appearance was also unusual, especially as the correct diagnosis was not suspected preoperatively. As a result, an operation was performed which did not adhere strictly to 'conventional' oncological principles of wide excision of the tumour along with its field of lymphatic drainage. Instead, a relatively simple procedure involving excision of the tumour with its 'stalk' and a thin cuff of rectum was performed. Most authors recommend wide resection of leiomyosarcomas whenever technically feasible. For example, Akwari *et al.*, in a large and detailed series from the Mayo Clinic of 108 small and large bowel leiomyosarcomas, found that prognosis was related, not surprisingly, to resectability and histological grade of tumour.⁵ Only 48% of cases could be submitted to potentially 'curative' resections and these had a 50% 5-year survival rate. Prognosis was not related to site of the primary tumour. These findings are broadly in keeping with other studies.^{2,3,6}

As well as offering the only hope of cure, wide surgical resection is advocated for other reasons: (1) It is difficult or impossible to differentiate between benign and malignant smooth muscle tumours pre- or intraoperatively.¹ (2) Resection affords worthwhile palliation with better quality, though not prolonged, survival.^{1–3} (3) Therapeutic or adjuvant chemotherapy and radiotherapy have nothing to offer in the treatment of gastrointestinal leiomyosarcomas.^{5,6}

Our patient had a rare rectal leiomyosarcoma 'locally' excised not in accordance with 'conventional' oncological principles, because of the difficulty in establishing an exact diagnosis preoperatively. It is therefore not possible to be certain that the disease has been eradicated and to give an accurate prognosis. It is encouraging that the 'stalk' of the diverticulum and the cuff of rectum was microscopically free of malignancy. The patient has no evidence of residual, recurrent or metastatic disease after 9 month's follow-up but, clearly, long term review will be necessary to establish whether or not this form of limited surgery is appropriate for rectal leiomyosarcoma presenting in this unusual way as a rectal diverticulum.

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