

Total Pelvic Exenteration for Huge Rectal Gastrointestinal Stromal Tumour with Concurrent Urinary and Intestinal Reconstruction

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Abstract

A 67-year-old female presented to our hospital with a 12-month history of abdominal pain. Preoperative pelvic magnetic resonance imaging revealed a large pelvic mass arising from the anterior wall of the rectum invading the bladder and uterus, with suspected lymph node metastases. To eradicate the tumor, total pelvic exenteration was carried out. An end-to-end colorectal anastomosis was performed. A Y-shaped uretero-ureteral anastomosis coupled with ureterovesical reimplantation was used for urinary reconstruction. Pathology showed the lesion was a high-risk gastrointestinal stromal tumor (GIST). The patient was treated with imatinib for 6 months following the operation, without tumor recurrence. In conclusion, total pelvic exenteration for huge rectal GISTs with concurrent urinary and intestinal reconstruction is technically feasible. However, whenever a preoperative biopsy and pathological diagnosis is available, neoadjuvant imatinib should be prescribed for large rectal GISTs, as it significantly decreases both tumour size and mitotic activity, thereby permitting less radical and sphincter-preserving surgery.

Keywords: Pelvic exenteration, gastrointestinal stromal tumour, urinary reconstruction, intestinal reconstruction

Introduction

Gastrointestinal stromal tumours (GISTs) represent the most common mesenchymal soft tissue tumours of the gastrointestinal tract (1). Approximately 5% of GISTs originate from the rectum, and often require radical surgery to achieve a complete resection (2). Despite the use of neoadjuvant imatinib for rectal GISTs significantly decreases the tumour size and permits less radical sphincter-preserving surgery (3), accurate preoperative pathological diagnosis of GISTs was not always possible due to the unusual imaging manifestations (4). Here we present a case report of total pelvic exenteration for a large rectal GIST with a concurrent urinary and intestinal reconstruction. The written informed consent was obtained from the patient.

Case Presentation

A 67-year-old female presented to our hospital with the symptom of abdominal pain for 12 months. Preoperative pelvic magnetic resonance imaging revealed a large pelvic mass arising (12.7 cm at its largest diameter) from the anterior wall of rectum invading the bladder and uterus with suspectable lymph node metastases in the mesorectum

(Figure 1). After a multidisciplinary team discussion, a computed tomography (CT) or endoscopic ultrasound (EUS) guided biopsy was recommended. However, the patient refused the advice of biopsy and insisted on getting surgery. To eradicate the tumor, total pelvic exenteration was carried out after obtaining written informed consent.

The patient was placed in the lithotomy Trendelenburg position. The invaded ileum was first resected. The sigmoid colon was then mobilized. The inferior mesenteric artery and vein were ligated and divided. Then, the ovarian vessels were ligated and cut. The ureters were identified. The internal iliac artery was divided at the distal location (after branching off the superior gluteal artery). The internal iliac veins were transected with linear staplers. The para-vesical spaces were dissected. The lateral pelvic lymph nodes were removed with the obturator nerves preserved. The left ureter was transected near the bladder. The right ureter was transected at the level of sacral promontory because the distal part seemed to be invaded by the tumour. A partial cystectomy was performed because the bladder neck was not infiltrated by the tumour. Total hysterectomy was performed. The vaginal stump was closed with a 2-0 monofilament,

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absorbable suture. The rectum was mobilized and transected at 5 cm distal to the tumour with a linear stapler. Then the specimen was removed (Figure 2a). The distal margin of rectal resection was 5 cm from the anal verge. A straight end-to-end colorectal anastomosis was performed with a circular stapler. Then, a cystoplasty was performed, and a Y-shaped uretero-ureteral anastomosis coupled with ureterovesical reimplantation was used for urinary reconstruction (Figure 2b). A modified Lich-Gregoir reimplantation technique was applied for the ureterovesical anastomosis (5). The postoperative

bladder capacity was about 180 mL. A greater omental pedicle was prepared and transplanted into the pelvic cavity. Drainage tubes were placed appropriately. The pelvic floor was reconstructed with a biologic mesh (Figure 2c) (6). The operative time was 420 minutes. The estimated blood loss was 600 mL. The specimen was completely intact with a wide resection margin (Figure 2d). The postoperative course was uneventful and the patient was discharged 12 days after surgery. Pathology report showed the lesion was a high-risk GIST, with mitotic rate of 15/50 high power field, involving rectal wall, uterine, bladder,

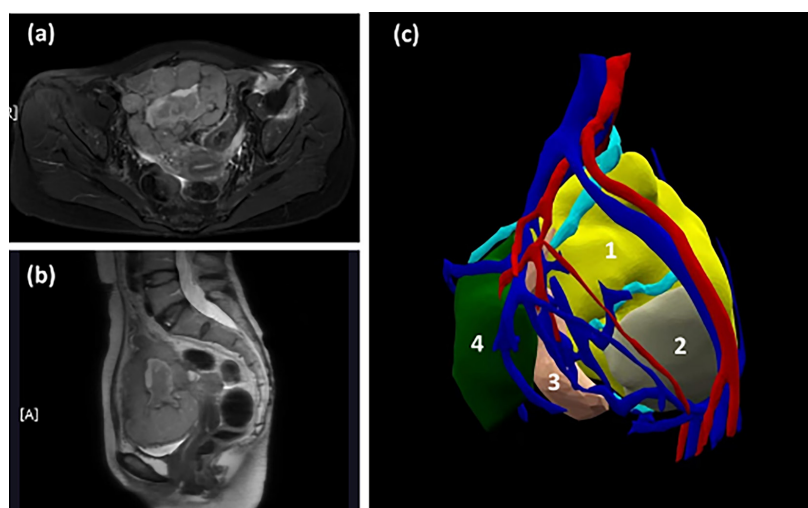


Figure 1. (a) Transverse section of MRI; (b) Sagittal section of MRI; (c) 3D reconstruction of MRI
MRI: Magnetic resonance imaging, 3D: Three-dimensional

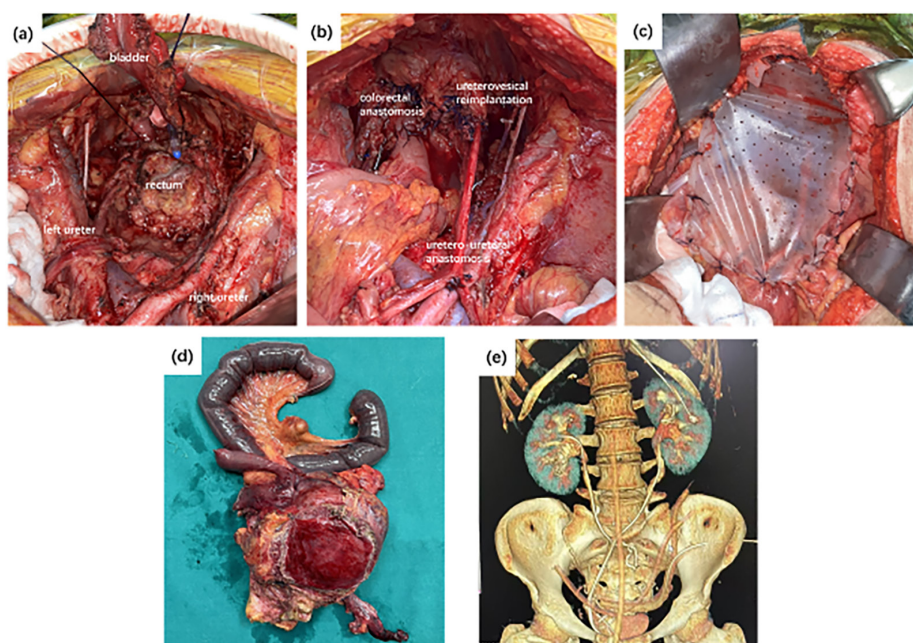


Figure 2. (a) Intraoperative photograph after removing the specimen and before reconstruction; (b) A Y-shaped uretero-ureteral anastomosis coupled with ureterovesical reimplantation for urinary reconstruction and an end-to-end colorectal anastomosis for intestinal reconstruction; (c) Pelvic reconstruction with a biological mesh; (d) Resected specimen; (e) Postoperative CTU showed the reconstructed urinary tract

CTU: Computed tomography urography

ureters and ileum, with no (0/43) lymph node involvement. A postoperative CT urography was performed one month after surgery, showing no urinary fistula or stricture (Figure 2e). Double J catheters were inserted intraoperatively and removed 3 months after surgery. No hydronephrosis occurred after their withdrawal. The patient was treated with imatinib for 6 months following the operation without tumor recurrence.

Discussion

Here we report a case of large rectal GIST treated with total pelvic exenteration. We believed this top-level case had an educational value for both urologists and colorectal surgeons.

Firstly, a multidisciplinary approach was particularly important in the case of rectal GIST. It should be noted that the imaging manifestations of GISTs were quite variable, so the surgeons would not necessarily be aware of the disease preoperatively (4). Nevertheless, it was preferable to obtain a preoperative diagnosis to exclude differential diagnoses including prostate cancer, leiomyosarcoma, germ cell tumours, lymphoma, neurogenic tumours, and desmoid tumours, as these pathologies may require different treatment strategies (7-9). A CT or EUS, guided biopsy should be considered before the surgery whenever possible. The benefits of neoadjuvant imatinib treatment in patients with high-risk GISTs arising from the rectum have been proven, including tumour down-sizing, reduction in mitotic activity, and a reduced risk of recurrence (3,10). The European Society for Medical Oncology (ESMO) guidelines recommended considering neoadjuvant imatinib treatment if R0 tumour resection is not feasible, and if resection can be achieved by less mutilating surgery, or if the operation can be made safer, for instance by decreased blood loss and risk of tumour rupture (11).

Secondly, if neoadjuvant treatment with imatinib was planned, it was essential to confirm the diagnosis of GISTs by histological and molecular tests, as there may be a small number of tumours with platelet-derived growth factor receptor alpha mutations or a wild-type that does not respond well to imatinib (11). Preoperative treatment should be continued until best response, which typically occurred after 6-9 months (9). After surgery, patients at high risk of recurrence or distant relapse should receive 3 years of adjuvant imatinib, if their tumour was not likely to be resistant to therapy (11).

Lastly, surgery should be considered for all rectal GIST cases, regardless of size. Surgery for GISTs must achieve a complete removal (full layer) of the tumor-bearing rectal wall because GISTs originate from the muscularis propria and not the mucosa. Surgical strategy needed to be tailored to the anatomic site, the size of the tumour, and the relation to the adjacent

organs and sphincter complex. The approach should be carefully considered, as rectal GISTs may be accessed and resected via transabdominal, transanal, pararectal approaches, and minimally invasive approaches (12,13). When possible, an organ-preserving approach should be considered. However, with large rectal GISTs as in this case, it was sometimes difficult to preserve the adjacent organs despite neoadjuvant imatinib therapy, and extended operations such as total pelvic exenteration should be selected, if necessary, to achieve R0 resection (13). Besides performing a pelvic exenteration, we carried out a complex urinary reconstruction and an end-to-end intestinal reconstruction. The advantage was that it led to a good quality of life. However, complex reconstruction was technically challenging and might need more re-interventions (14). Our center performs more than 200 pelvic exenteration surgeries annually. The use of the greater omentum pedicle and a biological mesh was our standard approach for pelvic reconstruction. In our experience, the combined use of greater omental pedicle and biological mesh promoted neovascularization, hemostasis, tissue healing, and regeneration, which led to faster and more effective recovery (6).

Conclusion

Total pelvic exenteration for large rectal GISTs with concurrent urinary and intestinal reconstruction is technically feasible. However, whenever preoperative biopsy and pathological diagnosis is available, neoadjuvant imatinib should be prescribed for large rectal GISTs, as it significantly decreases both tumour size and mitotic activity, which may permit less radical and sphincter-preserving surgery.

Ethics

Informed Consent: The written informed consent was obtained from the patient.

Footnotes

Authorship Contributions

Surgical and Medical Practices: H.Z., Y.L., Concept: H.Z., Design: H.Z., Data Collection or Processing: H.Z., Y.L., P.Z., Analysis or Interpretation: H.Z., Literature Search: H.Z., Y.L., P.Z., Writing: H.Z., Y.L., P.Z.

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