



Desmoid fibromatosis involving the ureter: A rare presentation with intraoperative challenges

John Lahoud^{a,*}, Voon Fong^a, Saissan Rajendran^b, Vincent Tse^a

^a Department of Urology, Concord Hospital, Sydney, Australia

^b Department of Vascular Surgery, Concord Hospital, Sydney, Australia

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ABSTRACT

Following failed retrograde and antegrade ureteric stenting, a 35-year-old male patient underwent an elective boari flap for marked proximal hydronephrosis due to a periureteric mass in the right iliac fossa. Intraoperative vascular surgical assistance was required for control of arterial bleeding due to friable vessel wall. Histopathology demonstrated desmoid fibromatosis.

1. Introduction

Desmoid fibromatosis is a rare, locally invasive, highly recurrent, and benign neoplasm, most frequent in patients in their second or third decade of life.¹ There are very few reports that describe ureteric invasion by a desmoid tumour and the perioperative experiences of institutions. Furthermore, there is no established optimal management of this disease in general.² This report describes the case of desmoid fibromatosis in a young male patient that required surgical exploration and reconstruction at our institution. Furthermore, we detail the intraoperative challenges associated with local invasion of desmoid tumour on the ureter and surrounding vasculature.

2. Case presentation

A 35-year-old man born in India with hypercholesterolaemia (on low dose statin) and previously treated helicobacter pylori, presented with a five-day history of right lower quadrant abdominal pain that radiated to his lower back on the same side. This was only associated with nausea and vomiting, on a background of a year history of urinary frequency with no constitutional symptoms. On initial blood tests, the patient had normal renal function and inflammatory markers were not raised. Computed tomography (CT) of the abdomen and pelvis demonstrated marked proximal right hydronephrosis with the right ureter passing directly through a peripherally enhancing, septated mass in the right iliac fossa.

An intraoperative retrograde pyelogram demonstrated multiple filling defects at the right iliac fossa with failed ureteric stent insertion

(Fig. 1). Diagnostic laparoscopy demonstrated serous fluid in the pelvis and right upper quadrant, and an irregular firm, soft tissue mass in the right paracolic gutter that did not vermiculate. A biopsy demonstrated non-specific fibrosis and mild acute inflammation and the serous fluid showed acute inflammatory infiltrate with no malignant cells. The following day, the patient had a right-sided nephrostomy tube inserted under interventional radiology with failed antegrade stenting. Microbiology results assessing tuberculosis, hepatitis, urine, and parasite screens were all negative.

Two weeks following initial insertion of the percutaneous nephrostomy, an antegrade pyelogram demonstrated a zone of complete disruption of the middle third of the right ureter at the level of the first segment of the sacrum with an intervening urinoma. A follow up CT scan demonstrated resolution of the hydronephrosis with the nephrostomy tube. The patient also underwent a colonoscopy two months following initial presentation that only demonstrated hyperplastic polyps on the splenic flexure and rectum.

Following discussion of risks and benefits, the patient underwent an elective laparotomy with ureteric reimplantation into a boari flap. Intraoperatively, a mass was seen in the right iliac fossa encasing the ileocaecum, part of ileum, ureter, and common iliac bifurcation, eroding the anterior wall of the artery (Fig. 2). To save as much ureter as possible to enable an extended boari flap without tension, we saw it best not to divide the ureter too far proximal from the mass. The surrounding mesentery was inflamed but not resected. Inadvertent injury of the internal iliac artery occurred, with intraoperative vascular surgical assistance required. Internal iliac artery ligation and reconstruction was performed utilising PTFE from healthy common iliac artery to healthy

* Corresponding author. Hospital Road, Concord, Sydney, NSW, 2139, Australia.

E-mail address: john.lahoud@health.nsw.gov.au (J. Lahoud).

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Fig. 1. Pre-operative retrograde pyelogram (colour should be used for this image).

external iliac artery. Once vascular control was established, a long boari flap up to the level of L5 was created.

Post-operatively the patient developed bladder spasms following extensive mobilisation. This was managed by oxybutynin and resolved after seven to 10 days, especially after removal of the Foley catheter. A cystogram at day seven post-operatively demonstrated no urinary leakage, and the urethral catheter and percutaneous nephrostomy tube were removed (Fig. 3).

3. Discussion

There have been very few reports that have described desmoid tumours involving the ureter causing severe hydronephrosis. Yoon et al., in 2014 described a case of spontaneous ureteric rupture due to a locally invasive desmoid tumour leading to rupture of the ureteric wall and urinoma, managed by surgical exploration and reconstruction following failure of conservative management.³ More recently, Ono et al., in 2018 described a case of retroperitoneal desmoid-type fibromatosis that involved the ureter in gynaecological surgery.¹

Whilst no specific aetiology has been identified, the development

and growth of desmoid tumours may correlate with genetic predisposition, with associations found in patients with Familial Adenomatous Polyposis and Gardner's syndrome, hormone factors, and surgical trauma.³ Given local recurrence following excision, resection with clear margins may provide but not guarantee a low risk of recurrence.⁴

The optimal conduit for vascular reconstructions in the setting of retroperitoneal cancer surgery remains to be defined. Traditional vascular principles dictate the use of autologous conduit to reduce the risk of graft infection in an operative field that may be contaminated. The decision to utilise a prosthetic conduit was the need to perform reconstruction immediately, in an operation that involved blood loss and a long operative time. In a published series of arterial reconstructions in pelvic exenteration surgery, where 11 of 20 interposition grafts were performed using PTFE, only one post-operative graft infection resulting in arterial anastomotic breakdown was identified.⁵ Given the histopathology of the mass was also yet to be determined, the other benefit of the use of PTFE is its reduced target vessel thrombosis rate compared to autologous grafts in a field that may require post-operative radiotherapy.

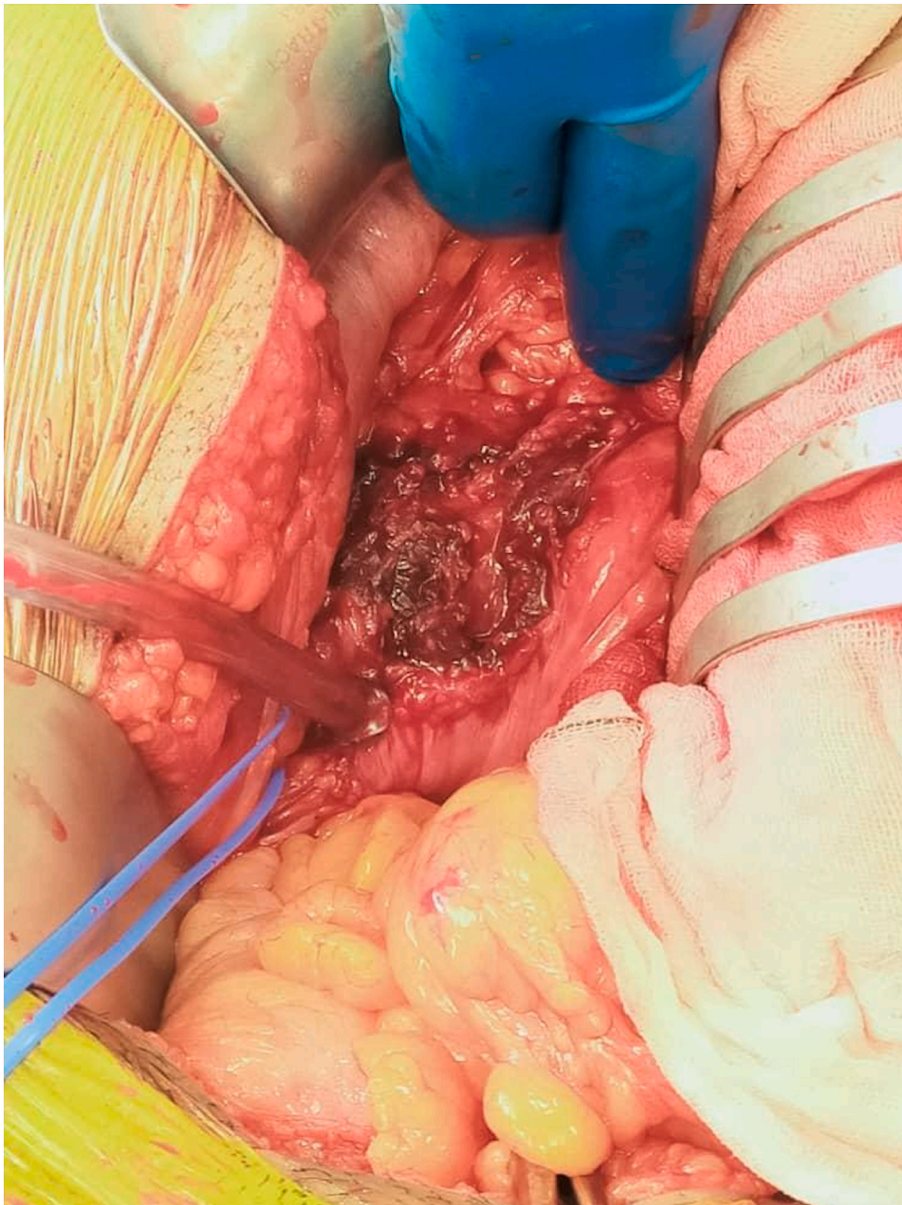


Fig. 2. Intra-operative image of desmoid tumour (*colour should be used for this image*). (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

4. Conclusion

We have described our approach in the perioperative management of a patient who presented with desmoid fibromatosis. It is important to anticipate the need for vascular surgical assistance intraoperatively given fragility of vasculature. The use of a prosthetic conduit such as PTFE has benefits in the context of reconstruction surgery and unknown pathology as mentioned.

Consent

Patient consent was obtained and documented. Patient is aware that personal information remained confidential. Patient is aware that written case report is intended for publication in peer-reviewed literature, and that intra-operative and radiographic images have been used.

Declaration of competing interest

The authors have no conflict of interests.



Fig. 3. Post-operative cystogram (colour should be used for this image).

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