Spontaneous ureteric rupture secondary to an invasive desmoid tumour

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INTRODUCTION: Spontaneous ureteric rupture is a rare entity that presents as an extravasation of urine from the ureter without previous surgery, ureteric manipulation and external trauma of the ureter. We report the case of a desmoid tumour presenting as spontaneous ureteric rupture which was managed in our institution.

PRESENTATION OF CASE: A 28 years old healthy male presented with a four day history of generalised abdominal pain secondary to spontaneous right ureteric rupture. Patient was initially managed via insertion of nephrostomy tube and antibiotics. After unsuccessful attempts of retrograde and antegrade ureteric stent insertion, patient was subsequently managed via elective surgical intervention. The excised specimen revealed desmoid tumour as cause of the ureteric rupture.

DISCUSSION: Desmoid tumours are rare benign tumours arising from fascial or musculoaponeurotic structures that do not metastasise, but tend to invade locally. It is often initially managed medically prior to undertaking a definitive surgical intervention. To our knowledge this is the first reported case of ureteric perforation secondary to a desmoid tumour of the mesentery.

CONCLUSION: Spontaneous ureteric rupture of the ureter is often misdiagnosed as other conditions. History taking and examination can be unreliable, hence a high level of suspicion and further investigations should be utilised. Once the diagnosis is made, treatment can be individualised based on aetiology.

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1. Introduction

Spontaneous ureteric rupture is a rare entity that presents as an extravasation of urine from the ureter without previous surgery, ureteric manipulation and external trauma of the ureter. Historically, there has been a small number of cases reported in the literature usually associated with urolithiasis. Other causes include pelvic cancer, retroperitoneal fibrosis, fluid overload, urinary retention and pregnancy. We report the case of a desmoid tumour presenting as spontaneous ureteric rupture which was managed in our institution. To our knowledge this is the first reported case of ureteric perforation secondary to a desmoid tumour of the mesentery.

2. Presentation of case

A 28 years old healthy male with no previous medical history presented with a four day history of generalised abdominal pain. He had no urinary or systemic symptoms. In particular he was afebrile and haemodynamically stable. On examination, there was non-specific generalised abdominal tenderness. Auxiliary blood tests revealed normal renal function and mildly raised liver function tests. Urinalysis, abdominal and chest X-rays were unremarkable. He was discharged from the emergency department with the view to have further outpatient investigation.

The abdominal ultrasound performed 2 weeks later revealed a right sided hydronephrosis. A CT with delayed urogram views confirmed the hydronephrosis and dilated ureter to the level of iliac vessels, where a 5 cm urinoma was apparent (Fig. 1). There was right ureteric disruption at the level of the pelvic brim without associated calculus or mass. Free pelvic fluid with extravasation of contrast was also noted. Patient was referred to urological surgical team who managed patient with intravenous antibiotics and percutaneous nephrostomy tube. The following day patient underwent retrograde study which confirmed the extravasation of contrast at the level of pelvic brim. Retrograde attempts to insert a ureteric stent were unsuccessful (Fig. 2).

Antegrade urogram via percutaneous nephrostomy two weeks later again confirmed contrast extravasation at the same location and two separate attempts of antegrade stenting were both unsuccessful.

After discussion of current status and risks, the patient underwent elective laparotomy with view of ureteric reimplantation.
Intraoperative findings revealed a mesenteric mass involving the ureter, terminal ileum and caecum. The intra-operative frozen section showed dense fibrous tissue and chronic inflammation. Due to poor quality of tissue and suspicious nature of the mesenteric lesion, an ileocolic resection was performed with functional end to end enterocolic anastomosis with the assistance of general surgical colleagues. Ureter was divided above the fibrous lesion and implanted to bladder via a Boari flap and psoas hitch. Patient had an uneventful post-operative recovery. Post-operative cystogram showed no leak and the ureteric stent was removed at six weeks post operatively (Fig. 3). Follow up MR abdomen showed no further mesenteric mass lesions intra-abdominally.

The final histopathology confirmed the diagnosis of mesenteric desmoid-type fibromatosis. The macroscopic and microscopic examination revealed a 55 mm × 45 mm × 40 mm irregular mass centred in the mesentery which invaded and destroyed the muscle layer in the small bowel wall and extended into the submucosa (Fig. 4). The ureter was entrapped and completely encircled by the mass which appeared to focally penetrate and destroy the ureteric muscle. Immunohistochemical staining results of the mass is shown in Table 1.

Patient was well at 6 months post-surgery on follow up. Outpatient colonoscopy and geneticist referral showed no abnormalities. The patient was found to not have any familial disorders such as Familial Adenomatous Polyposis (FAP) or Gardner’s syndrome.
3. Discussion

Spontaneous ureteric rupture presents as a sudden onset of severe abdominal and flank pain, often with nausea and vomiting. There can be associated urinary symptoms such as dysuria, urinary frequency and haematuria. Peritoneal irritation secondary to urine often leads to misdiagnoses with other causes of abdominal pain such as gastroenteritis, appendicitis or diverticulitis. Our patient who was young and clinically stable, was initially discharged from the emergency department with the presumed diagnosis of gastroenteritis for further outpatient investigations.

Ureteric rupture is attributed to an increased ureteric pressure due to obstruction of the collecting system or an ureteric tear during the ureteric stone passage.2 There has also been a report of spontaneous ureteric rupture secondary to intra-abdominal tumours such as mucinous ileocaecal tumour.10 To our knowledge, this is the first case of locally invasive desmoid tumour disrupting the ureteric wall, leading to rupture and urinoma.

The initial management of ureteric rupture is usually conservative. This includes insertion of a percutaneous nephrostomy tube to assist with retrograde insertion of stent. Simultaneous antegrade and retrograde urograms can be performed prior to insertion of ureteric stent to further characterise the rupture. In our case, when retrograde stenting was unsuccessful, patient was managed with period of free drainage via nephrostomy tube and antibiotics to wait for the urinoma to decrease in size before we attempted antegrade stenting. Finally surgical exploration and reconstruction was required when conservative management was unsuccessful.

Desmoid tumours are rare benign tumours arising from fascial or musculoaponeurotic structures that do not metastasise, but tend to invade locally.11 These connective tissue hyperplasias exhibit fibroblastic proliferation and account for 0.03% of all neoplasms.11 They tend to recur locally following excision. Recurrence is seen at a mean interval of 5 years in 78% of the patients after excision with curative intent.12

Surgical trauma, genetic predisposition and hormonal factors are considered to correlate with the development and growth of desmoid tumours.13 They have been associated with Familial Adenomatous Polyposis and Gardner’s syndrome.11 Intra-abdominal desmoids can remain asymptomatic for a considerable period until they enlarge and infiltrate into adjacent structures, causing compression or obstruction of other abdominal viscera such as intestine, blood vessels, or ureter. Symptoms can also arise from direct neural involvement. Mesenteric desmoid tumours can cause small bowel obstruction or ischaemia, hydronephrosis or form fistulae.13

Desmoid tumours cannot be distinguished from similar soft tissue tumours due to a similar attenuation to muscle on contrast-enhanced CT scans. This makes the accurate diagnosis on initial CT scan difficult. In our case, the diagnosis was only made intraoperatively.

Once the diagnosis of desmoid fibromatosis is established, first line treatment of mesenteric desmoid tumours is a non-steroidal anti-inflammatory drugs (NSAIDs) in combination with tamoxifen.13 The rationale for use of NSAIDs and tamoxifen is based on inhibition of prostaglandin synthesis and inhibition of ornithine decarboxylase induction, which restricts proliferation and growth of the tumour via enhancing immune responses of the tumours suppressor T-cells.14 In addition, tamoxifen is involved in blocking oestrogen receptors that often exists in desmoid tumours and mediating an inhibition of TGF-beta in fibroblasts resulting in the inhibition of collagen synthesis and lowered proliferation of fibroblasts.15 The daily dosage of sulindac (an indomethacin analogue with prolonged effect) is 200–400 mg and tamoxifen is 20–40 mg.13

Surgical excision remains the principle therapeutic management. However the influence of involved margins on local recurrence remained controversial. Aggressive resection with clear margins is a crucial factor in a successful outcome but clear margins do not guarantee a low risk of recurrence.11 Role of radiotherapy and its efficacy is disputed.11

4. Conclusion

Spontaneous rupture of the ureter is often misdiagnosed as other conditions. History taking and examination can be unreliable, hence a high level of suspicion and further investigations including an ultrasound and/or CT scanning should be utilised. Once the diagnosis is made, treatment can be individualised based on aetiology.

Conflict of interest

None.

Funding

None.

Ethical approval

Discussed with Ethics Committee Board at Westmead Hospital (Sydney, Australia). It was recommended that publication of a case study is sufficient with informed consent from the patient.
Author contributions

Dr. Peter Yoon executed the data collection, literature review and he is the main author of writing the paper. Dr. Nariman Ahmadi, Dr. Stephen Strahan and Dr. Audrey Wang studied the concept and design, wrote the paper and performed the final review.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Key learning points

- Spontaneous rupture of the ureter is rare and can be often misdiagnosed.
- Early diagnosis can be facilitated by high suspicion and appropriate ancillary investigations such as ultrasound and CT scans.
- Initial management of ureteric rupture is usually conservative however operative management can be feasible when carefully planned.

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