CASE REPORT

Xanthogranulomatous cystitis with malacoplakia, leading to spontaneous intraperitoneal perforation of the urinary bladder in a 9-year-old girl

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Accepted 27 July 2015

SUMMARY

A 9-year-old girl presented with sudden onset continuous abdomen pain, oliguria and constipation for 2 days. Ultrasound of the abdomen and voiding cystourethrography revealed intraperitoneal perforation of the urinary bladder. Cystoscopy showed two perforations at the dome of the bladder. Open surgical repair of the perforations was performed and biopsy was taken from their edges. Histopathological examination of biopsied material revealed malacoplakia with xanthogranulomatous cystitis. The patient was doing well at 15 months follow-up. Xanthogranulomatous cystitis with malacoplakia leading to spontaneous bladder perforation has not been reported earlier in the English literature.

BACKGROUND

This is a rare cause of a spontaneous intraperitoneal urinary bladder (UB) perforation in a 9-year-old girl due to xanthogranulomatous cystitis (XC) and malacoplakia; the case required the collaboration of a paediatrician with a urologist and pathologist to make the diagnosis and for management.

CASE PRESENTATION

A 9-year-old girl presented with sudden onset of a continuous dull aching pain of the abdomen, with abdominal distension, constipation, oliguria and elevated serum creatinine (159.12 µmol/L). On examination, she was found to have mild ascites, with diffuse tenderness in the abdomen. She was provisionally diagnosed as subacute intestinal obstruction with suspicion of bowel perforation, and was initially managed conservatively along with per urethral catheter placement. She responded well to treatment initially, but following catheter removal, she again developed symptoms. She was referred to our institute, where she was investigated further.

INVESTIGATIONS

An ultrasound of the abdomen was performed, revealing the presence of a Foley's catheter balloon outside the UB (figure 1A).

Voiding cystourethrography was performed, showing extravascular contrast into the peritoneal cavity from the UB up to the dome of the diaphragm (figure 1B).

A preoperative cystoscopy was performed, revealing two perforations at the dome of the bladder (figure 2A, B). Cystoscopy was beneficial to rule out bladder diverticulae and other associated

congenital anomalies, as the patient was 9 years old. Cystoscopy also helped to correctly diagnose the number of perforations.

DIFFERENTIAL DIAGNOSIS

Spontaneous UB rupture has an insidious presentation, and often results in delayed diagnosis and management because of vague signs and symptoms.¹

Rupture of bladder may be present with lower abdominal pain, inability to void and ascites.²

XC may present with non-specific urinary complications, abdominal pain or umbilical discharge.³ Microscopic examination shows multinucleated giant cells, lipid-laden macrophages (xanthoma cells) and cholesterol crystals.⁴

UB malacoplakia may present as haematuria, acute or chronic renal failure, and signs of urinary tract infection such as hesitancy, dysuria and frequency.

Case reports of these entities can be found in the literature, but their association with bladder perforation has not been reported.

TREATMENT

Open surgical repair of the bladder was performed and biopsy was taken from the edge of a perforation. The margins of the perforations were freshened-up by excising the edges. The bladder perforations were closed using absorbable polyglactin 3–0 sutures. The excised margins were sent for histopathological examination.

OUTCOME AND FOLLOW-UP

We administered a low dose suppressive antibiotic (cefixime) and ascorbic acid. The patient was doing

Figure 1 (A) Ultrasound of the bladder shows the presence of a Foley's catheter balloon outside the urinary bladder. (B) Voiding cystourethrogram shows extravascular contrast into the peritoneal cavity from the urinary bladder up to the dome of the diaphragm.

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To cite: Sharma K, Singh V, Gupta S, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2015-210786

Figure 2 Cystoscopy shows two perforations at the dome of the bladder (A and B).

well at 15-month follow-up. As the patient has continued to be asymptomatic, no further invasive investigation has been carried out to document malacoplakia persistence or disappearance.

Histopathological examination of the biopsied material revealed accumulation of histiocytes with granular eosinophilic cytoplasm and foamy macrophages in the lamina propria, with a few histiocytes showing intracytoplasmic basophilic inclusions. These findings are suggestive of XC with malacoplakia (figure 3A, B).

DISCUSSION

Spontaneous UB perforation is rare and, when encountered, is a diagnostic challenge.⁵ Around 60–85% of all bladder perforations result from blunt abdominal trauma and the majority of these are extraperitoneal.

Sudden onset of abdominal discomfort, ascites, ileus, haematuria and oliguria with elevated renal parameters should prompt the diagnosis. This can occur in cases of weakened bladder wall, such as in radiation injury, bladder tumours, ageing, trauma, urinary tract infections, catheter obstruction and in long-term catheterisation.⁶ Intraperitoneal perforation of the UB presents with abdominal pain and tenderness due to peritonitis. However, its diagnosis may be difficult, as the symptoms are vague and non-specific.² Differentiation of extraperitoneal from intraperitoneal perforation is essential for the management of bladder injuries.

CT cystography (CTC) and/or retrograde cystography (RGC) are the standard imaging tools for the diagnosis of bladder perforation.⁷ As RGC confirmed intraperitoneal perforation, CTC was not performed; moreover, CTC should be avoided in girls, for reasons to do with future fertility. Extraperitoneal rupture is treated conservatively while intraperitoneal UB rupture requires immediate surgical treatment because it can lead to peritonitis, which is less likely to heal with only catheter drainage.⁸ Rupture

Figure 3 Histopathological examination shows accumulation of histiocytes with granular eosinophilic cytoplasm and foamy macrophages in the lamina propria. A few histiocytes showing intracytoplasmic basophilic inclusions suggestive of xanthogranulomatous cystitis with malacoplakia (A) H&E $\times 5$, (B) H&E $\times 10$.

of the UB should always be considered in the differential diagnosis of patients presenting with free fluid in the abdomen/peritonitis, decreased urine output and haematuria, and in patients with increased levels of urea/creatinine detected in the serum and/or peritoneal fluid aspirate.

The aetiology of XC is also unclear. XC is an extremely rare, chronic inflammatory disorder found throughout the world, and approximately 30 cases have been reported since the first report in 1932 by Wassiljew.^{9–10} XC could possibly be a chronic inflammatory process caused by an immunological defect of the macrophage,¹¹ chronic infection with gram negative or anaerobic bacteria such as encountered in urinary tract infections or in foreign material, such as retained suture material, and abnormal lipid metabolism.^{12–13}

Malacoplakia is an inflammatory condition due to abnormal macrophage response to *Escherichia coli* and other bacterial urinary infection, and has a similar clinical picture and similar histological change as in XC. Most patients of malacoplakia are aged older than 50 years and are male. Malacoplakia may be characterised by basophilic lamellar inclusion bodies (Michaelis Guttman bodies) and large aggregates of monocytes (Von Hanseman bodies).¹³ In the urinary tract, females are more commonly affected (4:1).¹⁴

The disease is more common in immunocompromised individuals. Though the exact pathogenesis is not known, it is considered to be the result of chronic bacterial infections in patients with chronic debility or immunosuppression. The pathogenesis of malacoplakia mainly involves defective macrophage killing of bacteria, which results in an accumulation of bacterial degradation products and a granulomatous reaction, which clinically manifests as the formation of a papule, a plaque or ulceration.¹⁵ Usually, malacoplakia is a self-limiting benign condition associated with a good prognosis.¹⁶

The initial treatment of malacoplakia and XC consists of prompt treatment of the urinary infection and surgery at the affected site.

To the best of our knowledge, this will be the first case reported in the English literature on XC coexisting with malacoplakia leading to spontaneous perforation of the bladder. This report describes an extremely rare case of spontaneous bladder perforation in a young girl due to XC and malacoplakia.

Learning points

- ▶ Although lesions of malacoplakia are innocuous in most patients, if present in the urinary bladder, they should be followed-up and regularly monitored to prevent possible perforation.
- ▶ Perforation of the urinary bladder should always be considered in the differential diagnosis of patients presenting with free fluid in the abdomen/peritonitis, decreased urine output and haematuria, and in patients with increased levels of urea/creatinine detected in the serum and/or peritoneal fluid aspirate.
- ▶ Owing to vague symptoms, spontaneous bladder perforation may only be diagnosed after considerable delay, substantially increasing mortality. Therefore, early and accurate diagnosis with imaging techniques is imperative.
- ▶ The urologist should be aware of possible intraperitoneal bladder perforation, as emergency exploratory laparotomy and surgical repair of perforation is required in almost all cases.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- Galbraith JG, Butler JS, McGreal GT. Opioid toxicity as a cause of spontaneous urinary bladder rupture. *Am J Emerg Med* 2011;29:239.
- Ketata S, Boulaire JL, Al-Ahdab N, et al. Spontaneous intraperitoneal perforation of the bladder: a late complication of radiation therapy for prostate cancer. *Clin Genitourin Cancer* 2007;5:287–90.
- Ekici S, Dogan EI, Ruacan S, et al. Xanthogranulomatous cystitis: a challenging imitator of bladder cancer. *ScientificWorldJournal* 2010;10:1169–73.
- Cozzutto C, Carbone A. The xanthogranulomatous process. Xanthogranulomatous inflammation. *Pathol Res Pract* 1988;183:395–402.
- Cabezali BD, Guerrero RF, López VF, et al. Spontaneous bladder perforation in an infant neurogenic bladder: laparoscopic management. *Case Rep Urol* 2013;2013:986362.
- Saleem MA, Mahmoud AM, Gopinath BR. Spontaneous urinary bladder rupture: a rare differential for lower abdominal pain in a female patient. *Singapore Med J* 2009;50:410–11.
- Quagliano PV, Delair SM, Malhotra AK. Diagnosis of blunt bladder injury: a prospective comparative study of computed tomography cystography and conventional retrograde cystography. *J Trauma* 2006;61:410–21.
- Aghbari SA, Harthy AA, Ahmed M, et al. Laparoscopic repair of traumatic intraperitoneal bladder rupture. *Sultan Qaboos Univ Med J* 2011;11:515–18.
- Wassilijew AI. Über Erkrankungen des urachus. *Z Urol Chir* 1932;35:199–212.
- Yang KS, Kim YH, Seong YK, et al. Xanthogranulomatous cystitis arising from the posterior wall of the bladder. *Korean J Urol* 2011;52:868–9.
- Bates AW, Fegan AW, Baithun SI. Xanthogranulomatous cystitis associated with malignant neoplasms of the bladder. *Histopathology* 1998;33:212–15.
- Walther M, Glenn JF, Vellios F. Xanthogranulomatous cystitis. *J Urol* 1985;134:745–6.
- Fornari A, Dambros M, Teloken C, et al. A case of xanthogranulomatous cystitis. *Int Urogynaecol J Pelvic Floor Dysfunct* 2007;18:1233–5.
- Yousef GM. Malacoplakia outside the urinary tract. *Arch Pathol Lab Med* 2007;131:297–300.
- Curran FT. Malakoplakia of the bladder. *Br J Urol* 1987;59:559–63.
- Patnayak R, Kumaraswamy MR, Subramanian S, et al. An unusual case of bilateral hydronephrosis caused by uretero-vesico malacoplakia in a young male: a case report and review of the literature. *Cases J* 2009;2:7527.

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