Micropapillary carcinoma of the bladder presented with spontaneous intraperitoneal bladder rupture

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Abstract

Spontaneous bladder perforation is a rare presenting feature of bladder malignancy. We describe an unusual case of a patient, admitted to emergency, with diffuse abdominal pain due to spontaneous bladder rupture in association with a micropapillary carcinoma. A diagnosis of an intraperitoneal bladder perforation was made during an emergency operation. Aspects of etiology, clinical presentation, diagnosis and management are described. Although cases of spontaneous carcinomatous bladder rupture are associated with high morbidity and mortality, prompt identification and treatment can lead to favourable outcomes.

Introduction

The spontaneous intraperitoneal rupture of the urinary bladder is extremely rare and life-threatening. The reported cases of spontaneous bladder rupture have been associated with very different conditions, such as ongoing chronic diseases of the bladder wall (squamous or transitional cell carcinoma,[1] tuberculosis, cystitis, radiation necrosis, stones[2]), acquired or congenital bladder diverticula,[3] alcohol intoxication,[4] normal vaginal delivery,[5] enterocystoplasty, pelvic radiotherapy,[6] anatomical outflow obstruction, indwelling catheters and neurogenic bladder.[7] The reported mortality rate associated with complications from bladder rupture has been estimated at 50%, but has declined in recent years due to better management of its serious complications, like hyperkalemia, renal failure and sepsis.[8]

Micropapillary carcinoma (MPC) of the bladder is the last defined bladder carcinoma.[9] It is considered a rare variant of urothelial carcinoma with aggressive behaviour and accounts for less than 1% of bladder tumours.[10] According to the published reports, its prognosis is poor and mainly affects men between 50 and 90 years.[11] The patient with spontaneous rupture of the bladder presented here had a perforation which occurred through an area of MPC. This was very unusual and, to our knowledge, there is no similar report in the literature.

Case report

A 79-year-old male was admitted with a 6-hour history of sudden-onset generalized abdominal pain. He had complained of hematuria and dysuria for many years, but this had been considerably more troublesome just before admission. He denied any abdominal trauma or alcohol intake and has no documented accompanying illness. On physical examination, the patient was afebrile, with moderate to severe lower abdominal pain, suprapubic and diffuse abdominal tenderness, moderate distention and rigidity and left inguinal scrotum hernia. He also had subtle signs of circulatory failure (blood pressure 105/65 mmHg, heart rate 120 beats per minute). His white cell count was 31.5×10^9/L, serum sodium 134 mmol/L, potassium 5.2 mmol/L and creatinine 290 umol/L. His level of urea (20.1 mmol/L) showed moderate acute renal failure. Plain abdominal X-ray showed only distension of the stomach with a level. An urgent abdominal ultrasound examination showed multiple intraperitoneal fluid collections and full urinary bladder, hidden episode of acute urinary retention, with calcified filling defects of about 30 mm on the right side and dome (Fig. 1). Upon catheterization, 1.5 L of purulent urine was drained from the bladder. There was significant pyuria and bacteriuria on urinalysis. No cross-sectional imaging was done.

A provisional diagnosis of an acute abdomen was made and an open laparotomy carried out. At laparotomy, the peritoneal cavity was found to be filled with about 300 mm of foul smelling urine. The dome and posterior bladder wall was thinned out in areas of about 5 cm with suspicion of a neoplastic transformation, and a 3 mm perforation was identified from which there was leaking bloodstained urine (Fig. 2). The area of bladder necrosis was partly excised and

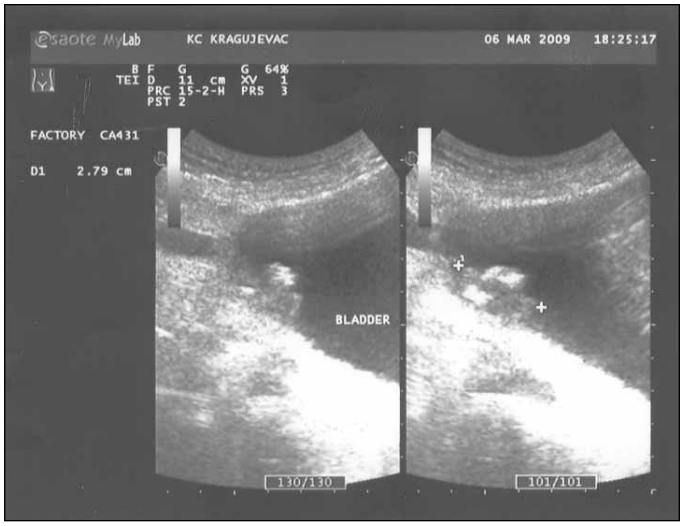

Fig. 1. Ultrasound of the pelvis shows distended bladder with calcified filling defects of about 28 mm on the dome.

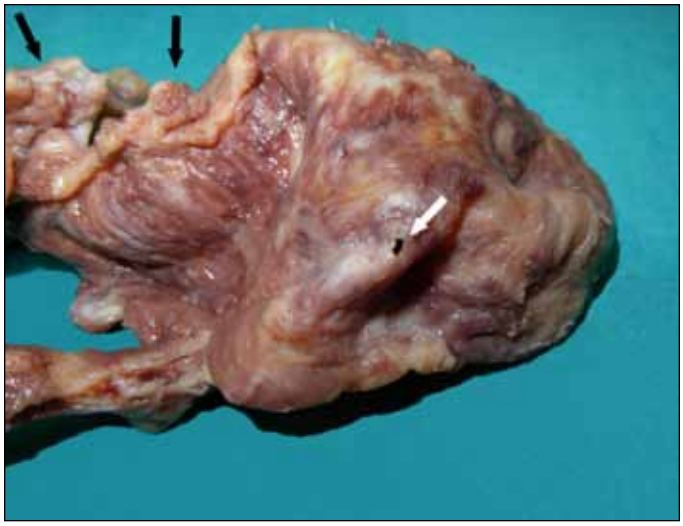

Fig. 2. Macroscopic appearance of the resected urinary bladder wall: tissue sample of irregular shape, size 110 × 65 mm, with perforation of the wall (3 mm in diameter, white arrow) and with discrete proliferation of the tumour (black arrows).

the defects were closed primarily in two layers. The bladder was drained via urethral catheters. A histological examination of the specimen around the fistula revealed MPC of the bladder with stromal invasion (Fig. 3, panels A and B). Postoperatively, computed tomography (CT) scan revealed dextroposition of the bladder, reduced bladder capacity, irregular thickness of the bladder wall (up to 19 mm), the most callous in the area of the vault, with severe bladder trabeculation or diverticula and with no lymph node metastases in the pelvic cavity. Transrectal ultrasonography of the prostate revealed an enlarged benign prostatic hyperplasia (66 cm^3) with normal homogeneous, low-level echogenicity. Due to the patient's severe clinical course, his supportive and antibiotic therapy was prolonged. The patient was discharged with a long-term indwelling urethral catheter; after two months, he showed significant improvement in renal function (anticipated for his age) and he was referred to the team of oncologists. They decided to administer postoperative palliative radiotherapy only, considering the patient's age, general health status and prognosis. Six months after radiotherapy, a surveillance CT scan revealed a similar picture, reduced bladder capacity with bladder muscle thickening, the most pronounced in the area of the vault, no clear sign of tumor recurrence, and no lymph node metastases in the pelvic cavity (Fig. 4). He remained well for months, but died 12 months after the operation. According to the death certificate, completed by the patient's primary physician, there was no clinical evidence of disease recurrence.

Discussion

Spontaneous carcinomatous bladder rupture is very rare and most reported cases occur in men.[12] The true incidence of the condition is not known and to date 19 cases have been reported.[13] Cases of rupture through an area of squamous cell carcinoma,[14,15] sarcomatoid carcinoma[16,17] and transitional cell carcinoma[1] have been reported. There is only one reported case of childhood rhabdomyosarcoma causing spontaneous bladder rupture 12 years after the successful initial treatment.[18] To our knowledge, this is the first case of spontaneous intraperitoneal rupture of the urinary bladder due to a micropapillary carcinoma of the bladder. We estimate that the mortality rate with spontaneous bladder rupture is between 47% and 80%,[12,14] depending on the clinician's awareness of the disease. Our patient survived acute illness and his subsequent death was not due to the disease.

In our patient, both the tumour site and the rupture were at the dome of the bladder. Perforations at the lateral and posterior walls, as well as the base of the bladder, were sporadically reported, but the dome or fundus is the weakest

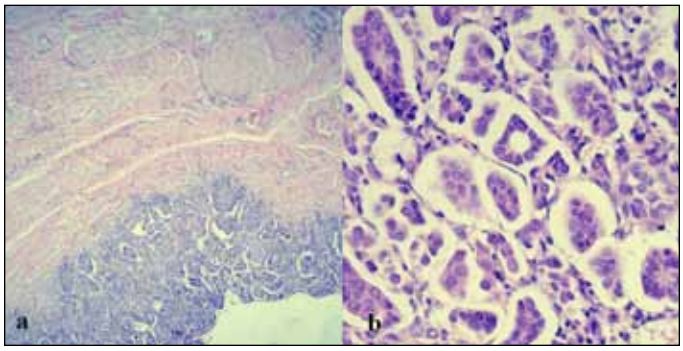

Fig. 3. Invasive micropapillary carcinoma: a) microscopic view of the bladder wall with tumour localized in the superficial part and extended to muscle (hematoxylin–eosin stain, original magnification, × 40); b) cytological features of invasive micropapillary carcinoma: tumour cells showing moderate amounts of cytoplasm, vesicular nuclei and markedly uneven coarse chromatin (hematoxylin–eosin stain, original magnification, × 400).

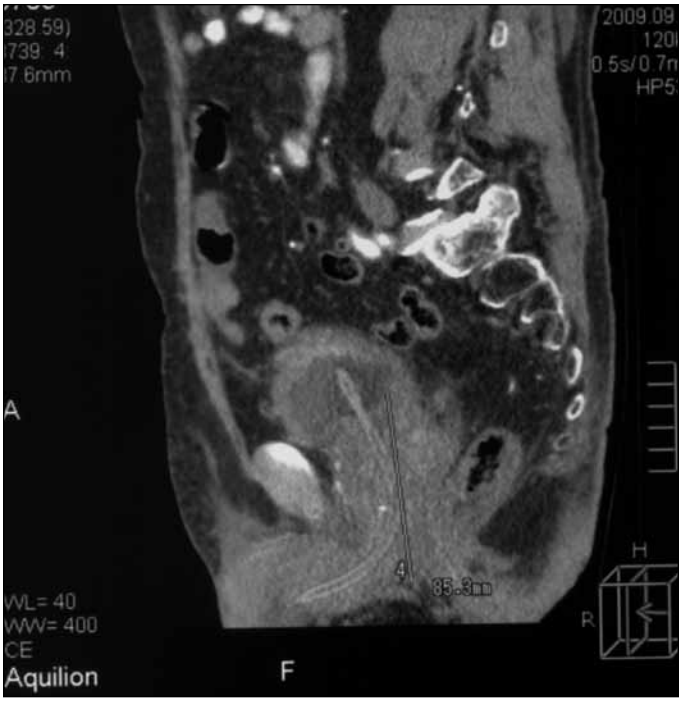

Fig. 4. A computed tomography scan of the bladder shows reduced bladder capacity with bladder muscle thickening, the most pronounced in the area of the vault, no clear sign of tumour recurrence, enlarged benign prostatic hyperplasia, and also without lymph node metastases in the pelvic cavity.

area of the bladder and therefore virtually all spontaneous bladder rupture cases occur there.[4] Increasing intraluminal pressures,[3] due to prostatic hypertrophy, was also considered a predisposing factor.[19] Therefore, the factors contributing to the bladder rupture in this case are urinary retention, the tumour itself and the likely minimal increase in intraabdominal pressure and underlying bladder wall disease. The contribution of each factor in our patient is difficult to surmise.

Retrograde cystography or CT cystogram is the procedure of choice to diagnose a ruptured bladder. Once the diagnosis is established, surgical intervention is indicated for intraperitoneal rupture, whereas extra-peritoneal rupture usually can be managed conservatively.[20] In the early stages after bladder rupture, symptoms can still be minimal, vague and diffuse. Failure to demonstrate perforation should not preclude immediate surgical intervention if such treatment is clinically indicated.

MPC of the bladder is considered a rare, distinct variant of urothelial carcinoma and occurs mostly in elderly men.[10] There are about 100 cases of MPC in the literature.[11] The incidence is about 0.7% to 2.2% among bladder tumours,[10,21] and the most common presenting symptom is hematuria. No currently defined imaging techniques can reliably diagnose some types of deeply invasive urothelial carcinoma of urinary bladder, in particular its micropapillary variant. Histologically, the micropapillary pattern is almost always associated with conventional transitional cell carcinoma. Overlapping features of immunohistochemical profiles do not allow discrimination between different variants of urothelial carcinoma.

This type of tumour is aggressive and has limited response to treatment. The current most generally favoured treatment for all patients with MPC is immediate radical cystectomy.[11] The patient in this case finally received palliative radiotherapy, although radiotherapy in itself usually is not effective for MPC,[11] particularly in asymptomatic patients. However, in the state of spontaneous carcinomatous bladder rupture, the patient's condition usually does not allow an appropriate radical procedure. Furthermore, most patients have high stage tumours and are not suitable for radical surgery. In addition, gross features of MPC are highly variable and range from a strikingly ulcerated mass having an obviously malignant appearance upon gross examination to where it is barely visible in the form of granular mucosa with low suspicion of a neoplastic transformation.[22] These features make radical surgery a less likely option, as was the case with our patient. Finally, in some cases, there is complete response with radiation therapy in lymph node metastasis of micropapillary bladder tumour.[23]

Extenuating circumstances in our case (increased bladder capacity) make bladder preservation and the avoidance of radical surgery necessary, as we have achieved good immediate oncological outcome in this critically affected patient. Patients with MPC who received surgical treatment had a median disease-free survival of 11 months and relapsed after a median follow-up period of 13.5 to 62 months.[24] Our patient lived shorter than the reported median relapse time, which further suggested that his death was unrelated to the malignant disease. As of now, there are only 3 reported patients who had radical surgery due to carcinomatous bladder rupture and one of them died 2 months after surgery.[13,14]

Conclusion

We encountered a case of spontaneous rupture of the bladder associated with MPC. Extenuating circumstances in our case enabled us to perform a wide partial cystectomy around the perforation site and tumour and repair of the bladder rupture. The procedure enabled satisfactory immediate and probably delayed oncological outcome in this critically affected patient.

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