CASE REPORT

Ureteric obstruction secondary to retroperitoneal fibrosis leading to acute kidney injury

Harald Bjorndalen,¹ Robert A Hastings²

¹Department of Surgery, South Warwickshire NHS Foundation Trust, Warwick, Warwickshire, UK

²Department of Medicine, University Hospitals Coventry and Warwickshire NHS Trust, Coventry, Warwickshire, UK

Correspondence to

Dr Harald Bjorndalen, h.bjorndalen@doctors.org.uk

SUMMARY

A 65-year-old man presenting with apparent acute coronary syndrome was found to have an acute kidney injury. A CT scan of the abdomen revealed bilateral hydronephrosis secondary to retroperitoneal fibrosis, and an unruptured infrarenal abdominal aortic aneurysm. Medical management with corticosteroids relieved the ureteric obstruction and renal function started to normalise within 72 h.

BACKGROUND

Retroperitoneal fibrosis is a rare condition in which most of the cases are idiopathic but can be caused by other inflammatory conditions. It has been linked with abdominal aortic aneurysms (AAA).^{1 2} In this case, retroperitoneal fibrosis caused obstruction of the ureters leading to hydronephrosis and subsequent acute kidney injury (AKI). The combination of AAA and retroperitoneal fibrosis is a rare cause of AKI, but it may be easily treatable and has good outcomes.³

CASE PRESENTATION

A 65-year-old Caucasian man with a 75-pack-year smoking history presented with acute central chest pain associated with severe nausea and vomiting. The pain lasted for 20 min and went away after the administration of a sublingual glyceryl trinitrate spray. The patient's medical history included an AAA, hypertension, subarachnoid haemorrhage 40 years ago, duodenal ulcer 20 years ago and a hiatus hernia. Regular medications included omeprazole, metoclopramide and co-codamol.

On examination, his heart rate was 90 beats/min, blood pressure 165/90 mm Hg, respiratory rate 16 breaths/min and oxygen saturations 95% on air.

There were no cardiac abnormalities on examination, but auscultation revealed crepitations in the left lower lobe of the lungs. On palpation of the abdomen, there was a pulsatile expansive mass at the level of, and below, the umbilicus.

INVESTIGATIONS

Laboratory tests revealed potassium 8.6 mmol/l, urea 33.2 mmol/l, and creatine 821 µmol/l. Troponin T was mildly elevated at 0.02, but did not increase over the next 12 h. D-dimer was raised at 1.7 mg/l.

ECG showed borderline ST-elevation in leads V1, V2, V3 and V4.

There were no abnormalities noted on the chest x-ray.

A noncontrast CTscan of the abdomen and pelvis was performed due to the patient's poor renal function. The scan revealed bilateral hydronephrosis

secondary to retroperitoneal fibrosis and a 5.2 cm unruptured infrarenal AAA. A renal cyst was also noted in the left kidney (figures 1 and 2).

These findings suggested that retroperitoneal fibrosis around the ureters was the cause of the bilateral hydronephrosis and subsequent AKI.

DIFFERENTIAL DIAGNOSIS

A 65-year-old gentleman presenting with central crushing chest pain, nausea and vomiting made acute coronary syndrome a likely diagnosis. This was initially high on the list of differentials when reviewing the initial ECG. Ruptured AAA is another necessary differential in this patient with a known AAA. The patient was haemodynamically stable, making this diagnosis less likely. Laboratory tests showing hyperkalaemia and AKI could have precipitated an acute cardiovascular event.

TREATMENT

1. Immediate medical management of hyperkalaemia;
 - ▶ myocardial protection with calcium gluconate,
 - ▶ Repeated insulin and dextrose infusions,
 - ▶ Salbutamol nebulisers,
 - ▶ Potassium sequestration with calcium resonium.
2. Urinary catheterisation;
3. Stop nephrotoxic drugs;
4. Twice daily blood tests of renal function;
5. Intravenous rehydration;
6. Corticosteroids for resolution of ureteric obstruction: 60 mg prednisolone orally daily for 1 week, then 40 mg daily for the next week, then 30 mg daily until the next renal outpatient review 2 weeks after discharge.

Figure 1 CT abdomen showing bilateral hydronephrosis.

To cite: Bjorndalen H, Hastings RA. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2012-008093

Figure 2 CT abdomen showing dilated abdominal aorta and surrounding fibrosis (arrows).

OUTCOME AND FOLLOW-UP

Initially, renal function did not recover, but after 72 h, serum creatine was significantly improving, decreasing from 895 to 540. The patient improved steadily while on prednisolone for 10 days. He then self-discharged against the doctors' advice. When he attended an outpatients' review 3 weeks later, renal function had improved further (creatinine 140). The patient has since been lost to follow-up.

DISCUSSION

Retroperitoneal fibrosis is a rare condition with an incidence of 1–2 in 200 000.⁴ Most of the cases are idiopathic but can be caused by inflammation and retroperitoneal fibrosis has been linked with AAA.² Autoimmune processes are suspected to be involved as some patients have autoantibodies and histological findings similar to vasculitis.^{2–4} A response to the leakage of inflammatory cells from the atheromatous plaque of the AAA is considered to be the underlying cause of the associated fibrosis of retroperitoneal structures.^{1–2} Other causes of retroperitoneal fibrosis include malignancy and drugs.

Presentation is usually with nonspecific symptoms such as back or flank pain, abdominal pain, vomiting, leg oedema or symptoms or signs suggestive of other conditions occurring as a result of retroperitoneal fibrosis, such as urinary tract obstruction.^{4–5}

There are no specific laboratory tests for retroperitoneal fibrosis. C reactive protein may be elevated or normal, and it is therefore not useful to exclude retroperitoneal fibrosis or to monitor it as a marker of response to therapy.²

Imaging is the primary diagnostic tool for both AAA and retroperitoneal fibrosis. Ultrasound may be used to look for AAA and ureteric obstruction but has limited application in the diagnosis of retroperitoneal fibrosis. Contrast-enhanced CT or MRI are the imaging modalities of choice, enabling the identification of all structures enveloped by fibrosis and the extent of fibrosis (see figures 1 and 2).² Repeat CT or MRI is invaluable in monitoring for recurrence, and this is essential in patients treated medically as significant recurrence has been reported.^{3–5}

Ureteric obstruction can be medically managed with steroids, as in this case, or invasively with a stent or open ureterolysis. Prednisolone prevents the inflammatory process and inhibits fibrosis. Treatment may be required for up to 1 year.^{3–5} Steroid-sparing agents such as methotrexate, azathioprine or cyclophosphamide may also be used in conjunction with corticosteroids.^{2–5–6} Immunosuppressive therapy may also be of value combined with more invasive treatment options such as

ureteral stenting and open ureterolysis.^{2–4} Open ureterolysis is effective, but laparoscopic ureterolysis has been shown to have lower complication rates and may be more suitable in certain populations, such as the elderly.^{7–9}

Our case showed that although there was initially a poor response to steroids, it is worth pursuing as the outcome was positive and it is noninvasive, not carrying the associated risks of invasive procedures.

A similar case is described by Li Wai Suen *et al*.¹⁰ Here, an 85-year-old Caucasian man presented to the emergency department with a 6 month history of nonspecific symptoms and was found to have acute renal failure and bilateral hydronephrosis secondary to retroperitoneal fibrosis. He was receiving treatment with bilateral ureteric double-J stents. He consequently had adequate diuresis with demonstrable improvement in renal function.¹⁰

Causes of AKI are best categorised as Pre-renal, Intrarenal and Postrenal.

Postrenal causes include:

- ▶ Renal/ureteric calculi,
- ▶ Retroperitoneal fibrosis,
- ▶ Urethral stricture,
- ▶ Bladder cancer,
- ▶ Prostate hypertrophy.

Epidemiology of AKI¹¹;

- ▶ Acute tubular necrosis 45%,
- ▶ Prerenal 21%,
- ▶ Obstruction 10%,
- ▶ Acute on chronic 13%,
- ▶ Glomerulonephritis 3%,
- ▶ Acute interstitial nephritis 2%,
- ▶ Vasculitis 1.5%,
- ▶ Vascular 1%,
- ▶ Other 3.5%.

Learning points

- ▶ Retroperitoneal fibrosis is a rare disease with many possible causes and complications.
- ▶ Retroperitoneal fibrosis should be considered in patients with long-standing abdominal aortic aneurysms when presenting with acute kidney injury (AKI).
- ▶ Consider persisting with steroids even when the initial response is poor as it is less invasive than other treatment options.
- ▶ Principles of management in AKI:
 - Improve renal perfusion.
 - Aim for euvolemia.
 - Investigate obstruction.
 - Treat specific underlying causes.
 - Review drugs.
 - Treat life-threatening complications.
- ▶ Seek early advice from a nephrologist in cases of AKI due to retroperitoneal fibrosis.
- ▶ Follow-up over 1 year with CT or MRI to monitor for recurrence.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Rasmussen TE, Hallett JW Jr. Inflammatory aortic aneurysms: a clinical review with new perspectives in pathogenesis. *Ann Surg* 1997;225:155–64.
- 2 Warnatz K, Keskin AG, Uhl M, *et al.* Immunosuppressive treatment of chronic periaortitis: a retrospective study of 20 patients with chronic periaortitis and a review of the literature. *Ann Rheum Dis* 2005;64:828–33.
- 3 Van Bommel EFH, Siemes C, Hak LE, *et al.* Long-term renal and patient outcome in idiopathic retroperitoneal fibrosis treated with prednisone. *Am J Kidney Dis* 2007;49:615–25.
- 4 Burkhardt SS, Fehr A, Brandt AS, *et al.* Retroperitoneal fibrosis. *Aktuelle Urol* 2007;38:221–31.
- 5 Swartz RD. Idiopathic retroperitoneal fibrosis: a review of the pathogenesis and approaches to treatment. *Am J Kidney Dis* 2009;54:546–53.
- 6 Marcolongo R, Tavolini IM, Laveder F, *et al.* Immunosuppressive therapy for idiopathic retroperitoneal fibrosis: a retrospective analysis of 26 cases. *Am J Med* 2004;116:194–7.
- 7 Stifelman MD, Shah O, Mufarrij P, *et al.* Minimally invasive management of retroperitoneal fibrosis. *Urol* 2008;71:201–4.
- 8 Srinivasan AK, Richstone L, Permpongkosol S, *et al.* Comparison of laparoscopic with open approach for ureterolysis in patients with retroperitoneal fibrosis. *J Urol* 2008;179:1875–8.
- 9 Tobias-Machado M, Correa WF, Korkes F, *et al.* Transmesocolic ureteral intraperitonealization: a new approach for laparoscopic treatment of retroperitoneal fibrosis. *J Laparoendosc Adv Surg Tech A* 2011;21:341–4.
- 10 Suen CFDLW, Aw HC, Eapen R, *et al.* Case report of a patient with bilateral hydronephrosis secondary to retroperitoneal fibrosis. *Internet J Uro* 2011;8. http://www.ispub.com/journal/the_internet_journal_of_urology/volume_8_number_2_31/article/case-report-of-a-patient-with-bilateral-hydronephrosis-secondary-to-retroperitoneal-fibrosis.html?utm_source=feedburner&utm_medium=feed&utm_campaign=Feed%3A+InternetScientificPublications+%28Internet+Scientific+Publications+-+RSS%29 (accessed 20 Sep 2012).
- 11 Liaño F, Pascual J. Madrid ARF study group: epidemiology of acute renal failure: a prospective, multicenter, community-based study. *Kidney Int* 1996;50:811–18.

Copyright 2013 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.

BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- Submit as many cases as you like
- Enjoy fast sympathetic peer review and rapid publication of accepted articles
- Access all the published articles
- Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow