Urology Case Reports 17 (2018) 36–38

Contents lists available at ScienceDirect

Urology Case Reports

journal homepage: www.elsevier.com/locate/eucr

Inflammation and infection

Unilateral renal forniceal rupture - A rare presentation of retroperitoneal fibrosis

Anthony Adimonye*, Iram Parwaiz, Arthur Cotton

Department of Urology, Cheltenham General Hospital, London, UK

ARTICLE INFO

Article history:
Received 4 October 2017
Received in revised form
18 October 2017
Accepted 17 November 2017
Available online 28 December 2017

Keywords:
Retroperitoneal fibrosis
Ureteric obstruction
Urinoma
Declarations

1. Introduction

Retroperitoneal fibrosis (RPF) is a rare inflammatory process associated with the development of extensive fibrosis around the abdominal aorta and surrounding retroperitoneal structures. The symptoms of RPF are often insidious and non-specific resulting in diagnostic delay with ensuing complications related to the progressive fibrotic deposition. A common complication of RPF is ureteric obstruction leading to hydronephrosis and varying degrees of renal impairment.[1,2] However, spontaneous renal forniceal rupture due to RPF is exceedingly rare and to our knowledge has not been previously described.

An unusual case of RPF causing left ureteric obstruction with spontaneous left renal forniceal rupture and its management is discussed in light of the current literature.

2. Case report

A 51 year-old lady presented with a four-month history of intermittent left flank pain with a subsequent history of sudden-onset worsening central abdominal pain lasting for one week. Her past medical history and drug history were unremarkable. Physical examination was normal, except for a pyrexia of 38 °C. Urinalysis was normal although her blood results revealed raised inflammatory markers and renal impairment (eGFR 74 → 42); hence she was commenced on broad-spectrum IV antibiotics.

Due to the non-specific nature of her symptoms an abdominal computed tomography (CT) scan was organised which revealed extensive retroperitoneal para-aortic infiltration of soft tissue with left ureteric obstruction and renal forniceal rupture resulting in an urinoma (Fig. 1).

She subsequently underwent an emergency cystoscopy and left retrograde ureteric stent insertion resulting in an improvement of her pain and renal function. The nature of this soft tissue was further investigated via haematological and histological means. Tumour (CA 19-9, CA-125 and alpha-fetoprotein) and immunological (IgA, IgM and IgG 1–4) markers were all normal. CT biopsy was inconclusive, revealing a core of fibrous connective tissue with prominent plasma cells, however no distinct pathognomonic features of RPF. Despite a normal immunological blood profile and an inconclusive biopsy, the radiological findings pointed to a diagnosis of benign idiopathic RPF.

Dual therapy with prednisolone and methotrexate were commenced. A follow-up CT scan and dimercaptosuccinic acid (DMSA) scan performed 2 and 6 months post initial presentation respectively, showed significant resolution of the periaortic and retroperitoneal fibrotic infiltration, good residual left kidney function with no evidence of scarring (Fig. 2).

3. Discussion

RPF, also known as chronic periaortitis, is an uncommon condition with an incidence of 1.38 cases per 100,000 people,[2] with men affected twice as commonly as women with a mean age of diagnosis between 50 and 60 years.[1,3] Though malignancy, surgery, infections, radiotherapy and drugs such as b-blockers, are known secondary causes, the primary cause of RPF is idiopathic.[1]

* Corresponding author. Department of Urology, Cheltenham General Hospital, Sandford Road, Cheltenham, GL53 7AN
, UK.
E-mail addresses: anthony247@doctors.org.uk (A. Adimonye), iram.parwaiz@nhs.net (I. Parwaiz), arthur.cotton1@nhs.net (A. Cotton).

https://doi.org/10.1016/j.eucr.2017.11.014

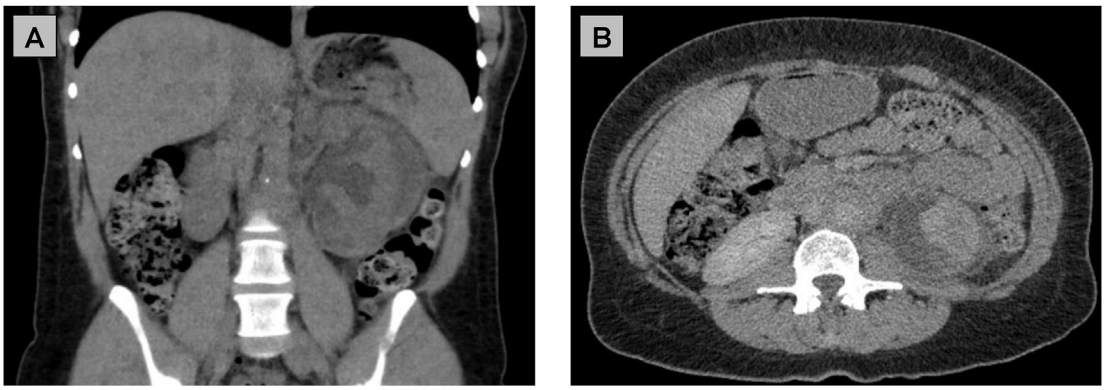

Fig. 1. CT Scan. (A) Coronal view and (B) Axial view at the level of L1 depicting the extensive retroperitoneal para-aortic infiltration of soft tissue, left renal forniceal rupture and resulting perinephric urinoma.

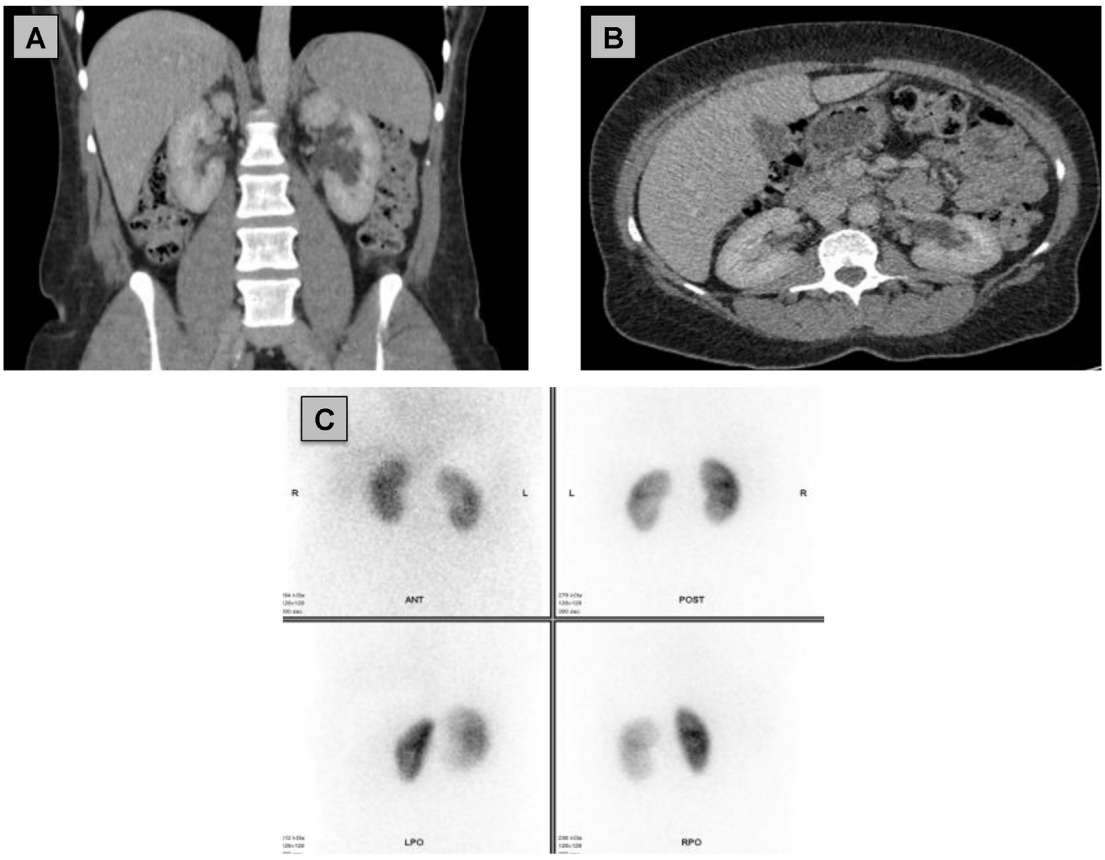

Fig. 2. CT and DMSA Scan. (A) Coronal and (B) Axial (at the level of L1) CT views shows complete resolution of the perinephric urinoma and significant resolution of retroperitoneal para-aortic soft tissue infiltration. (C) DMSA Scan revealed no evidence of renal scarring on either kidney with uptake favouring the right kidney at 57%.

Recent reports suggest approximately half of all cases of idiopathic RPF can be included in the spectrum of immunoglobulin G4-related disease (IgG4-RD).[3] One study showed IgG4-related RPF had a significantly higher recurrence rate compared to IgG4-unrelated RPF thus the need to distinguish between the two.[4]

The diagnosis of RPF can be challenging. The symptoms of RPF are often insidious, with patients presenting with abdominal pain and constitutional symptoms such as fatigue and weight loss. These non-specific symptoms often lead to diagnostic delay resulting in complications related to the progressive fibrotic deposition. This includes ureteric obstruction, the commonest complication, which occurs in 47–100% of patients, causing hydronephrosis, which is bilateral in over 50% of cases.[1,2] Rarely, does renal forniceal rupture occur due to ureteric obstruction from idiopathic RPF and to our knowledge we are the first to describe this. Gershman et al.[5] who on review of 108 cases found that renal forniceal rupture was most commonly caused by ureteric obstruction by ureteric calculi (80 cases; 74%), malignancy extrinsic ureteric compression (9 cases; 8%) and rarely by benign compression (two cases - gravid uterus and pelvic abscess; 2%). However, they did not describe RPF as a possible benign cause of renal forniceal rupture. Other late complications of RPF include bowel obstruction and spinal cord compression.

Ureteric obstruction is managed conservatively where possible. However, temporary ureteric stenting or insertion of percutaneous nephrostomies while treating the cause systemically is generally successful.

In our case, we were able to achieve a good outcome despite a left renal forniceal rupture secondary to ureteric compression from RPF, with left ureteric stenting and dual therapy with prednisolone and methotrexate. Significant fibrotic resolution was noted with 2 months of medical therapy. However, due to the risk of recurrence, she will need a long follow-up period.

4. Conclusion

RPF is rare occurrence and this report depicts an unusual and interesting case of the management of a left renal forniceal rupture secondary to ureteric compression from RPF with a favourable outcome.

Ethics approval and consent to participate

'Not applicable'.

Consent for publication

Consent obtained from patient.

Conflicts of interest

The authors declare that they have no competing interests.

Funding

'Not applicable'.

Acknowledgments

None.

Appendix A. Supplementary data

Supplementary data related to this article can be found at https://doi.org/10.1016/j.eucr.2017.11.014.

References

1. Kermani TA, Crowson CS, Achenbach SJ, Luthra HS. Idiopathic retroperitoneal fibrosis: a retrospective review of clinical presentation, treatment and outcomes. *Mayo Clin Proc*. 2011 Apr;86(4):297–303.
2. Van Bommel EF, Jansen I, Hendriksz TR, Aarnoudse AL. Idiopathic retroperitoneal fibrosis: prospective evaluation of incidence and clinicoradiologic presentation. *Medicine (Baltimore)*. 2009 Jul;88(4):193–201.
3. Zen Y, Onodera M, Inoue D, Kitao A, et al. Retroperitoneal fibrosis: a clinicopathologic study with respect to immunoglobulin G4. *Am J Surg Pathol*. 2009 Dec;33(12):1833–1839.
4. Warnatz K, Keskin AG, Uhl M, Scholz C, 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 Jun;64(6):828–833.
5. Gershman B, Kulkarni N, Sahani DV, Eisner BH. Causes of renal forniceal rupture. *BJU Int*. 2011 Dec;108(11):1909–1911.