CASE REPORT

Transcaval ureter: a rare embryological anomaly causing obstructive uropathy

Stephanie Giddy,¹ Isaac Thangasamy,^{1,2} Antonio Vega Vega²

¹School of Medicine, University of Queensland, Herston, Queensland, Australia

²Department of Surgery, Rockhampton Base Hospital, Rockhampton, Queensland, Australia

Correspondence to
Stephanie Giddy,
swgiddy@gmail.com

Accepted 29 January 2015

SUMMARY

A 21-year-old woman presented with a longstanding history of episodic right flank pain related to fluid consumption and recurrent urinary tract infections. On examination, there was right costovertebral angle tenderness. Renal tract ultrasound demonstrated hydronephrosis and CT urography confirmed transcaval ureter. The patient's clinical presentation was likely due to obstruction associated with the right ureter passing through the opening created by the duplicated segment of the inferior vena cava. While asymptomatic transcaval ureter can be managed conservatively, severe symptoms or complications of ureteric obstruction may require surgical treatment. In this case, treatment comprised a laparoscopic ureteroureterostomy with an end-to-end anastomosis between the proximal and distal sections of the right ureter. The patient's postoperative recovery was uneventful and she reported resolution of the flank pain during the one year of follow-up.

BACKGROUND

Transcaval ureter is a rare embryological abnormality of the inferior vena cava (IVC) in which a segmental duplication of the IVC creates a venous ring that encircles the right ureter. It is a unique cause of ureteric obstruction, which requires appropriate management to prevent complications. Few cases of transcaval ureter have been reported in the literature. This is the first report to describe a case of transcaval ureter managed laparoscopically.

CASE PRESENTATION

A 21-year-old woman was referred to an urologist with severe episodic right flank pain occurring intermittently for the past 2 years. The flank pain was precipitated by consuming considerable amounts of fluids and alleviated by limiting intake. The patient also described a history of recurrent urinary tract infections, for which she frequently required treatment with antibiotics. There were no other associated urinary, gastrointestinal or gynaecological symptoms, and the patient denied any relevant medical or family history. The patient reported minimal alcohol intake and was a non-smoker. Examination revealed only mild right costovertebral angle tenderness; the remainder of the examination was found to be unremarkable.

INVESTIGATIONS

Results of a full blood count, liver function tests, urea, electrolytes and creatinine and a urinalysis were within normal ranges. The Society for Fetal Urology (SFU) grade I hydronephrosis on the right

side was demonstrated on ultrasound of the renal tracts. Subsequently, CT urography was performed and revealed transcaval ureter, in which the right ureter emerged between the two vessels of a partially duplicated IVC ([figure 1](#)).

TREATMENT

As the patient's symptoms became more unbearable, surgery was preferred over conservative management. Cystoscopy and insertion of a right double-J ureteric stent were performed, followed by a laparoscopic ureteroureterostomy ([figure 2](#)). An indwelling catheter was inserted and the patient was placed in the left lateral decubitus position. During the procedure, the colon was retracted and the duodenum was dissected to access the IVC. The left and right common iliac veins were joined together to form the proximal portion of the IVC and then divided into two vessels, before eventually fusing to give rise to the single IVC again. The right ureter was dissected proximal and distal to the transcaval segment, sectioned and spatulated before executing an end-to-end anastomosis with anterior and posterior running sutures. The patient was monitored in the high-dependency unit for 24 h as part of routine protocol at the hospital.

OUTCOME AND FOLLOW-UP

Postoperative convalescence was uneventful and the patient was discharged a few days after the procedure. The stent was removed after 6 weeks. The patient was reviewed 1 month following the stent removal and then at 3 months with CT and MAG3 scans, which demonstrated resolution of the hydronephrosis. She expressed complete resolution of symptoms during the 1 year of follow-up.

DISCUSSION

IVC embryogenesis is an intricate process involving the coordinated formation and regression of several pairs of primitive veins.¹ Any deviation from normal development may result in various congenital IVC abnormalities, including transcaval ureter.

The most accepted theory that attempts to explain IVC embryogenesis is the supracardinal model.¹ This model proposes that formation of the IVC relies on the coordinated development and regression of anastomoses between three pairs of veins, namely the posterior cardinal, subcardinal, and supracardinal veins.² Initially, the posterior cardinal veins develop and drain blood from the embryo caudal to the heart.² As they regress to form the iliac bifurcation, the subcardinal veins appear and assume the role of the posterior

CrossMark

To cite: Giddy S, Thangasamy I, Vega Vega A. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2014-208076

Figure 1 (A) Axial and (B) coronal CT images demonstrating progression of right ureter (arrow) between the vessels of the partially duplicated inferior vena cava (*).

cardinal veins in draining blood from the caudal embryo.² The right subcardinal vein merges with the right hepatic vein to give rise to the hepatic segment of the IVC.² Meanwhile, the left subcardinal vein atrophies.² Finally, the supracardinal veins appear and contribute to the formation of the infrarenal IVC and azygos system.¹

There have been few reports on transcaval ureter in the medical literature, with variable explanations for its embryological development.^{3–7} According to the supracardinal model of IVC embryogenesis, transcaval ureter is more commonly thought to arise when the right posterior cardinal and right supracardinal veins fail to regress adjacent to the level of the kidney (figure 3). The resulting partial duplication in the infrarenal IVC creates a venous ring through which the right ureter can pass.

Other well recognised congenital anomalies of the IVC include left IVC, double IVC and retrocaval ureter.²

In particular, retrocaval ureter, in which the right ureter passes posteriorly to the IVC, should be considered in the differential diagnoses for this clinical presentation.

In this case, transcaval ureter was detected by CT urography, which also demonstrated a right hydronephrosis and proximal hydroureter. The patient's symptoms most likely resulted from increased venous return causing dilation of the IVC and subsequent compression of the transcaval segment of the ureter.

Management of patients with transcaval ureter is either conservative or surgical, and depends on the severity of symptoms, which is in turn determined by the degree of ureteric obstruction. Reported symptoms that patients may experience include flank pain,^{3–6} urinary frequency⁶ and haematuria.^{3 4 7} Although treatment generally comprises conservative management, the presence of complications, such as recurrent urinary tract infections,³ impaired renal function and urolithiasis,⁵ necessitates surgical correction. Surgical management involves resection of

Figure 2 Intraoperative photographs showing (A) the course of the right ureter (arrow) between the vessels of the duplicated segment of inferior vena cava (IVC) and (B) the reconstructed right ureter transposed anterior to the IVC (*).

Figure 3 Schematic diagram of embryology of (A) normal inferior vena cava and (B) transcaval ureter.

the ureter and an ureteroureteric anastomosis anterior to the IVC, which was performed laparoscopically in this case.

This first Australian report of transcaval ureter describes this rare embryological IVC anomaly as a unique cause of ureteric obstruction and as a differential diagnosis for flank pain. Imaging

modalities, such as CT urography, thus play an imperative role in the identification of transcaval ureter. This subsequently facilitates appropriate management to prevent complications of obstructive uropathy.

Twitter Follow Isaac Thangasamy at @iThangasamy

Contributors SG drafted and revised the report. IT conceived the idea for the report, assisted with the care of the patient and revised the report. AVV managed the patient and revised the report.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

Learning points

- ▶ Transcaval ureter arises as a consequence of abnormal development of the inferior vena cava, and is thought to involve the unusual persistence of the right postcardinal and right supracardinal veins, which would otherwise normally regress.
- ▶ It is a unique cause of ureteric obstruction and can potentially lead to impaired renal function, recurrent urinary tract infections and urolithiasis.
- ▶ Abdominal CT is a useful imaging modality for diagnosing transcaval ureter and assessing the degree of ureteric obstruction.
- ▶ Treatment involves conservative management or surgical intervention. Surgical correction, which comprised a laparoscopic ureteroureterostomy in this case, is indicated when obstructive uropathy is evident.

REFERENCES

- 1 McClure CFW, Butler EG. The development of the vena cava inferior in man. *Amer J Anat* 1925;35:331–83.
- 2 Bass JE, Redwine MD, Kramer LA, *et al*. Spectrum of congenital anomalies of the inferior vena cava: cross-sectional imaging findings. *Radiographics* 2000;20:639–52.
- 3 Dharman K. Transcaval ureter. *J Urol* 1980;123:575–6.
- 4 Gong J, Jiang H, Liu T, *et al*. Imaging of partial right double vena cava with ureter crossing through its split, confirmed at surgery. *Clin Imaging* 2011;35:148–50.
- 5 Gupta NP, Nayyar R, Sahay SC. Periureteric venous ring with renal calculi and transitional cell carcinoma: report of a rare case. *Surg Radiol Anat* 2010;32:405–7.
- 6 Rosen MP, Walker TG, Brennan JF, *et al*. Transcaval ureter with hydronephrosis: radiologic demonstration. *AJR Am J Roentgenol* 1989;152:793–4.
- 7 LePage JR, Baldwin GN. Obstructive periureteric venous ring. *Radiology* 1972;104:313–15.

Copyright 2015 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