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Case Report & Review of the Literature

Right Sided Bochdalek Hernia with Ureteric Involvement – A Case Report and Review of Management Options

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ARTICLE INFO

Article history:
Received: 1 December, 2020
Accepted: 16 December, 2020
Published: 25 December, 2020

Keywords:
Bochdalek hernia
diaphragmatic hernia
ureteric hernia
ureteric obstruction
rare ureteric obstruction
conservative management

ABSTRACT

Ureteric herniation through a diaphragmatic defect in adults (Bochdalek hernia – BH) is rare, with less than 15 cases reported in the literature. Most patients present with non-specific abdominal symptoms with or without worsening of renal function, and therefore, the clinical picture can be masked [1]. Management of ureteric involvement ranges from conservative to reconstructive surgery. We report a case of an elderly (>80-year-old) patient who presented acutely with symptoms mimicking an acute abdomen and on imaging was found to have a BH associated with herniation of right ureter causing hydro-ureter. We managed the patient conservatively, and she improved symptomatically, which was evident from her follow-ups. We provide a detailed case report of this situation as well as discuss the literature on the topic to guide treatment decision-making.

Background

Ureteric herniation can occur at various anatomical sites, including inguinal canal, femoral canal, sciatic foramen, and diaphragmatic hiatus, with diaphragmatic hiatus being the least common [2, 3]. Florid obstructive uropathy is rarely the predominant clinical picture, with the presentation commonly that of non-specific symptoms including abdominal pain, dyspnea, chest pain, and nausea and vomiting, sometimes leading to ileus [4, 5]. Management of this can therefore be challenging with a missed diagnosis potentially leading to sepsis and multi-organ dysfunction. There is very little indication in the literature to guide the management of ureteric involvement in BH in terms of acute and definitive management. This case report illustrates the complexities associated with the diagnosis and management of BH with ureteric involvement and deranged renal function. The accompanying review of the literature attempts to guide decision-making in this challenging scenario.

Case Report

An elderly patient presented with a 12-hour history of sudden onset right-sided abdominal pain, vomiting, bowels not opening and not passing flatus. It was her second admission in the month for similar complaints. She was known to have hypertension, chronic low backache for which she took codeine, and constipation for which she took lactulose. On admission, she was alert with a national early warning score (NEWS) of 1 for a pulse rate of 104. Abdominal examination revealed a non-distended, soft abdomen, with some tenderness in the right iliac fossa with diminished bowel sounds. Her urine dip test showed 1+ for blood, sugar, WCC, and ketones. Her blood tests showed a lactate of 2.1, elevated WCC 13.7 (normal- 4.00-11.0×109/L), creatinine- 125 (normal- 45-84 µmol/L) and eGFR- 36 (normal- 70-130 ml/min).

CT Abdomen and pelvis with contrast diagnosed herniation of the proximal right ureter through a posteromedial defect in the right hemidiaphragm (Figures 1 & 2). There was associated moderate right hydronephrosis with good contrast uptake in the right kidney. There was also herniated fat just above the right hemidiaphragm. Urgent urology review was sought. She was offered emergency stenting vs. conservative management. A joint decision was taken to consider a trial of conservative management with potential escalation to intervention if the patient deteriorated. She was managed conservatively with IV fluids, anti-emetics, and analgesics. On her repeat blood tests, the WCC was down to 9.7, creatinine 102, and eGFR-45. She was discharged home with an outpatient referral to urology for further work up and management.

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http://dx.doi.org/10.31487/j.SCR.2020.12.24

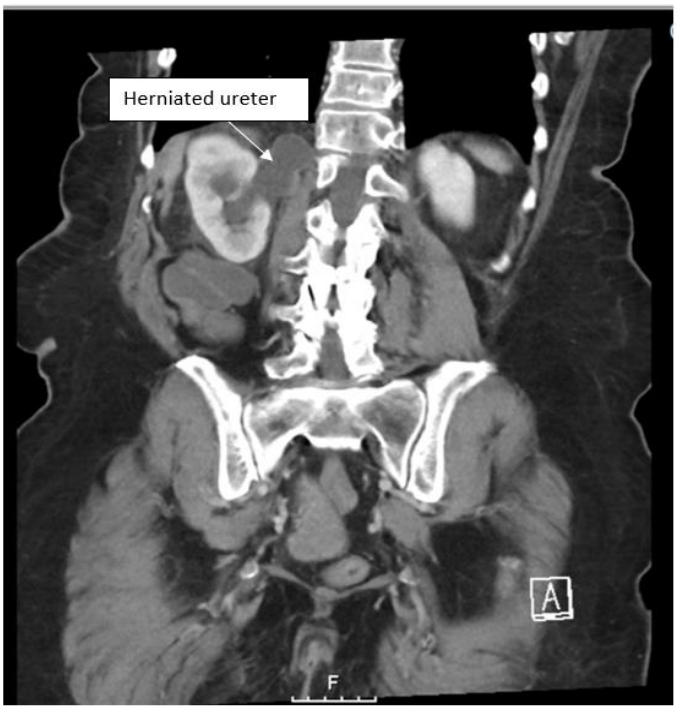

Figure 1: Coronal plane showing the right sided herniated ureter.

Figure 2: Sagittal view showing the posterior diaphragmatic defect through which the ureter is coursing.

She further had 2 visits to A&E, once in June and then in august of 2018, with pain in the right abdomen and no other bowel obstruction symptoms. During out-patient urology review, her workup showed a serum creatinine of 117 and eGFR-38. The urology team performed a MAG3 scan (Figure 3), which demonstrated a hydronephrotic, poorly draining but non obstructed right kidney with only mildly impaired function (equal spilt function). Conservative management was continued and future urology out-patient clinic appointment for renal function follow-up were set up, and these continue. She hasn't had any worsening of renal function or episodes of pyelonephritis on follow up.

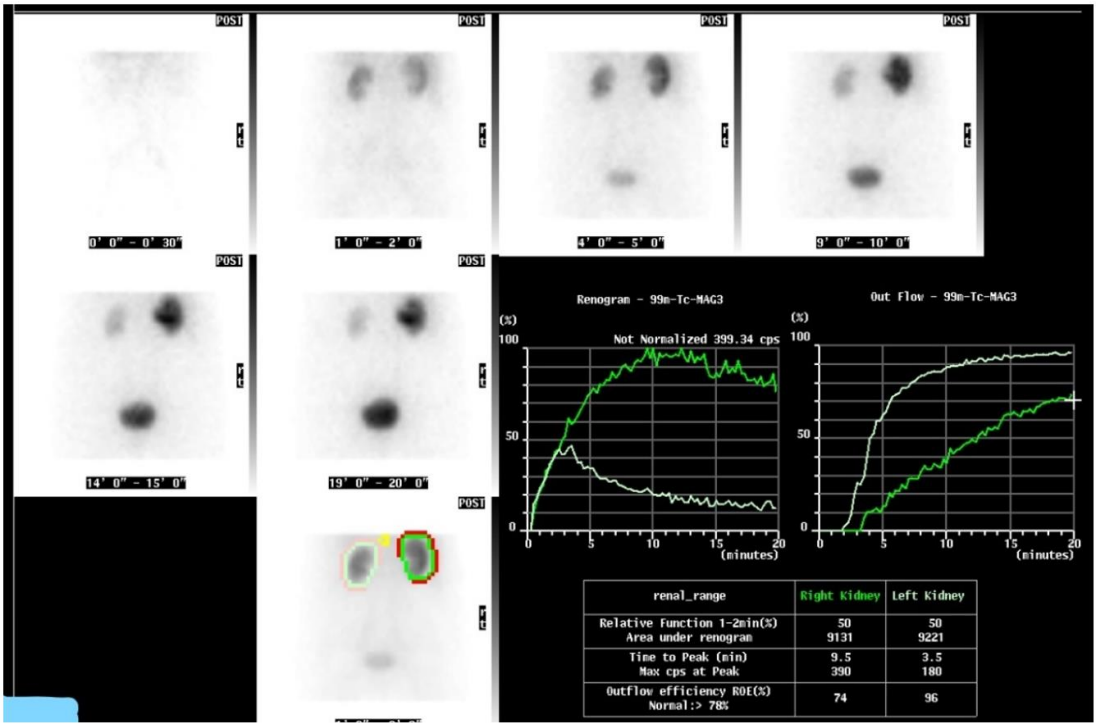

Figure 3: MAG3 renogram curves showing reduced uptake and outflow but equal split function.

Discussion

Bochdalek hernia is predominantly seen in children less than 1-year-old with respiratory symptoms and has a poor prognosis [6]. It is caused due to a failure of fusion of the pars lumbalis and pars costalis parts of the diaphragm [7]. Most adults with BH are diagnosed incidentally on imaging. Studies using MDCT and multiplanar reconstruction estimate a prevalence of up to 10.5% in patients having scans for other reasons [8]. Asymptomatic cases most often present equally on the right and left side, whereas symptomatic cases are more common on left with an incidence of 9:1 compared to the right [9]. However, ureteric involvement seems to always present on the right, as noted in the 14 cases reported in the literature. Table 1 summarizes the literature review.

Table 1: This summarizes the management plans for patients with diaphragmatic hernia with ureteric involvement as reported in the literature. Table modified from Abou Heider *et al*. [14].

Authors (Year of publication)	Age/gender	Laterality	Presentation	Acute Management	Outcome of acute management	Investigations	Indication for definitive management	Definitive management	Outcome of definitive management
Swithinbank (1958)	60/F	Right	Intermittent right flank pain with radiation to pelvis	Morphine, spontaneous resolution	Pain subsidence	Intravenous pyelogram showing loop of ureter with intrathoracic route, hydronephrosis	Refractory symptoms	Surgical repair – herniation reduced, and hiatus closed	Intra- venous pyelography, showed the right ureter pursuing a more normal, sub-diaphragmatic course
Paterson and Lupton (1989)	75/M	Right	Right hypochondrial pain, nausea and vomiting	Morphine, spontaneous resolution	Pain subsidence	IVP, CT showing herniation of PUJ Renogram confirming obstruction	Refractory symptoms	Surgical repair – side to side pelviureteric anastomosis	IVP showed normal ureteric course
Chawla and Mond (1994)	56/M	Right	Intermittent flank pain on both sides	Pain relief	Pain subsidence	IVP, CT showing herniation but no obstruction	none	Conservative	Intermittent symptoms with spontaneous resolution

Catalano *et al*. (1998)	63/F	Right	Upon workup for renal stones	ESWL for stone, pain relief	Pain subsidence	IVP, CT showing abnormal course of ureter with some dilatation	To prevent obstruction with stone fragment after ESWL, and to prevent further stones	Surgical removal of renal stone, reduction of hernia and repair of the diaphragm.	No further stones, normal course of ureter
Sukumar *et al*. (2010)	75/F	Right	Incidental finding on workup for renal failure and respiratory infection	Supportive management	Pain subsidence	CT, retrograde studies showed Curlicue sign. Prompt drainage suggesting no obstruction	None	Conservative	No report of further respiratory or renal compromise
Balakrishnan and Neerhut (2011)	83/F	Right	Intermittent Right flank pain	Retrograde ureteric stent insertion	Resolution of pain	CT showing herniation	none	6 monthly stent changes	No recurrence of pain
Song *et al*. (2011)	75/M	Right	Right upper quadrant pain	Retrograde ureteric stent insertion. Removal of stent	Resolution of pain and obstruction	CT showing herniation	Patient opted not to have surgery, no recurrence of hydronephrosis	conservative	No recurrence of pain or hydronephrosis
Hatzidakis *et al*. (2014)	86/F	Right	Septic obstructive pyelonephritis	Nephrostomy followed by antegrade ureteric stenting	Relief of sepsis and obstruction	CT showing obstruction and herniation	none	Stent insertion and change	Straightening of course of ureter.
Almeida *et al*. (2015)	82/F	Right	Incidental on PET scan for workup of lung nodule	None mentioned	None mentioned	CT showing a knuckle of ureter in chest	None mentioned	Not mentioned	None mentioned
Dru and Josephson (2016)	94/F	Right	Sharp right flank pain	Cystoscopy and retrograde stent insertion	Resolution of obstruction	CT showing herniation and obstruction	None mentioned	None mentioned	None mentioned
Lin *et al*. (2017)	81/F	Right	Right flank pain and renal obstruction	Initial conservative management, then retrograde ureteral stenting on progression of hydronephrosis and MAG3 showing obstruction	Resolution of obstruction	CT showing herniation. Mag3 showed reduced function on the right	Worsening herniation despite stent, presence of stented ureter in thorax	Surgical repair of defect, nephropexy, excision of redundant ureter and pelviureteric end-to-end anastomosis	No herniation or obstruction
Beland *et al*. (2019)	84/F	Right	Obstructing ureteric stone in a herniated ureter	Flexile ureteroscopy and LASER fragmentation of the stone	Removal of stone, persistence of herniation	CT showed stone and ureteric herniation	None mentioned	None mentioned	None mentioned
Abou Heidar *et al*. (2019)	71/M	Right	Abdominal pain, vomiting	Conservative	Improvement in symptoms	CT showing herniation	None	Conservative	None

			and lose stools						
Current case	>80/F	Right	Right sided abdominal pain	conservative	Improvement in symptoms	CT showing herniation, MAG3 showing no obstruction and normal split renal function	none	Conservative	none

The pathophysiology for this has never been completely elucidated. The origin is undoubtedly embryonic as the presence of BH is an embryonic occurrence [10]. As this is a non-dependent location for herniation, the presentation is probably related more to episodes of sustained raised intra-abdominal pressure as well as weakening of the diaphragm, which probably becomes more apparent with age [10]. Most of the cases report herniation of retroperitoneal fat along with the ureter correlating the pressure hypothesis. Most of the patients reported in the literature had some sort of pulmonary issues such as COPD, bronchitis etc., which could have precipitated the herniation over time.

Of these reported cases, 2/14 presentations were incidental while investigating for other pathologies. The remaining patients presented with mild right sided abdominal discomfort. Only one patient in the literature presented with septic obstructed system [4]. Management varied from conservatively (5 cases), surgical repair (4 cases), and ureteral stenting (4 cases). In two cases reports, the management wasn't mentioned [3, 4, 10-19]. The range and median age of presentation were 56-94 years and 75 years respectively demonstrating that the ureteric herniation had very little impact on these patients for the most part of their lives. Male to female incidence was 5:9.

Indication for treatment has not been clearly defined in the literature. There are 5 cases in the literature that shows that conservative management is feasible without any adverse outcomes. Our case shows this similarly. Patients undergoing ureteric stent placement seem to have had them placed for resolution of pain and to treat the episode of acute obstructive uropathy [17]. There is conflicting evidence for the use of ureteric stents in the treatment of obstructive uropathy, where pain is the driving factor for management, as stents themselves can cause pain. Reports that have suggested the use of stents have advised this in the presence of objective evidence of renal obstruction on imaging. They have also cautioned readers on the use of stents as the ureter can be very tortuous, necessitating the use of stiff guide wires and longer stents to prevent upward migration. In some patients, stents reduced the herniation, whereas in others, persistence of herniation despite stent was an indication for reconstructive surgery. In our discussion with the patient, her main symptom was pain, with some element of acute obstructive uropathy. Despite these findings, a trial of conservative management with IV fluids and IV antibiotics proved fruitful.

It is essential for those presenting with a septic obstructed system to have drainage either in the form of a nephrostomy or with a ureteric stent. Hatzidakis *et al*. further illustrate how retrograde stent insertion can be challenging in these situations because of the tortuosity of the ureter and the abnormal anatomical path it traverses [4].

The indication for major reconstructive surgery has been reported to be to reduce the potential for further incarceration and to get patients stent free. Long-term follow-up has not been reported, and therefore the effectiveness of this approach to management needs to be ascertained. The surgical treatment most reported involves plication of the diaphragm with one case mentioning nephropexy, excision of redundant ureter and ureteroplasty. It is interesting to note that most descriptions of major reconstructive surgery were from reports before 2000, and the more recent reports tend to treat this with conservative management.

Decision for major reconstructive surgery needs to be taken with caution as most of these patients seem to be older with comorbidities. We used a MAG3 renogram to guide us in the decision-making process, and as its results were reassuring, we proceeded with conservative management and managed to avoid the risks of major surgery.

Conclusion

Ureteric herniations through BH are uncommon and are picked up on non-invasive imaging for asymptomatic patients or patients with non-specific bowel and chest symptoms. Our case illustrates that uncomplicated patients without objective evidence of obstructive, decline in renal function or sepsis can be managed conservatively with imaging and blood test follow up without intervention. This is particularly attractive as most patients presenting with these findings are of older age with potential underlying comorbidities in whom surgery might not be safe. The literature review attempts to guide clinicians in the management of this challenging and rare condition.

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