Adult giant hydronephrosis diagnosed in the second trimester of pregnancy: A case report and literature review

Fathi Ramly a,⁎, Noor Azura Noor Mohamad a, Akmal Zulayla Mohd Zahid a, Norhana Mohd Kasim b, Khai Yeong Teh c

a Obstetrics & Gynaecology, Medical Faculty, UiTM Sungai Buloh, Malaysia
b Obstetrics & Gynaecology, Hospital Sungai Buloh, Malaysia
c Urology, Hospital Selayang, Malaysia

ABS T RACT

Adult giant hydronephrosis is an unusual finding during pregnancy. The most frequent cause is congenital obstruction at the ureteropelvic junction. Ultrasound accompanied by magnetic resonance imaging (MRI) can help in reaching the correct diagnosis. We report a case of giant hydronephrosis in a woman who presented at 23 weeks of gestation with abdominal distension. She was managed conservatively. Unfortunately, the pregnancy was complicated by severe pre-eclampsia at 32 weeks of gestation, necessitating delivery via emergency caesarean section. The literature and previously reported cases of giant hydronephrosis in pregnancy are reviewed.

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1. Introduction

Sterling first defined giant hydronephrosis (GH) as a gigantic dilatation of the pelvicalyceal junction that occupies a large part of the abdominal cavity, or dilatation of renal system that is filled with more than 1 l of urine [1,2]. The radiological definition of GH is hydronephrosis that spans to the hemiabdomen across the midline and which spans at least 5 vertebral bodies [3].

GH is a rare finding during pregnancy as most of patients are diagnosed and treated earlier in their life [3]. It should be included in the differential diagnosis when the origin of an abdominal mass is inconclusive. Ultrasound and magnetic resonance imaging (MRI) can help in reaching the correct diagnosis [4]. Managing such cases is challenging, as there is little discussion on the management and outcome of GH in pregnancy due to its rarity.

We report a case of adult GH diagnosed in the second trimester of pregnancy. We also review the key management strategies and their obstetric outcome in the previously reported cases in English literature.

2. Case

A 27 year-old woman, a primigravida at 23 weeks of gestation, presented to the emergency department with one-week history of left iliac fossa pain. The pain was exacerbated by movement, with pain score of 3. She denied any symptoms of urinary frequency or dysuria, and her urine output was normal. There was no per vaginal bleeding and no uterine tightening. The fetal movement was felt.

She was overweight (BMI 28.2 kg/m²) and had gestational diabetes diagnosed since the early second trimester. Her blood sugar control was normal. There was no significant congenital disease or medical illness in her family.

On examination, there was a mass palpable at 30 week size spanning over the left suprapubic area extending to the left hypochondrium. The mass was cystic in nature, non-mobile and tender on deep palpation. There was no rebound tenderness. The uterus corresponded to 24 week size and pushed to the right hemiabdomen. Serum full blood count, renal function test and urine microbiology investigations were within normal limits.

A transabdominal ultrasound scan (Fig. 1) revealed a viable single intrauterine fetus, which was pushed to the right by a huge cyst. The cyst was multiloculated, multiseptated and hypoechoic with no solid area. The outline of the cyst was smooth, and there was no Doppler uptake. There was also mild right hydronephrosis; however, the left kidney could not be visualised. MRI of the abdomen and pelvis revealed...
gross left hydronephrosis and a paper-thin renal cortex with a collapsed left ureter (Figs. 2 and 3).

Diagnosis of left GH was made, and she was managed conservatively following consultation with the urologist. Based on the joint multidisciplinary discussion, ureteric stenting was reserved for later, in case of severe urinary tract infection. The left-sided abdominal pain did not recur after the first episode.

The pregnancy continued until 32 weeks of gestation, when the patient was diagnosed with severe pre-eclampsia necessitating delivery via emergency caesarean section. A right paramedian skin incision and lower segment uterine incision were made to deliver a normal 1.8 kg male baby with good APGAR scores. Intraoperatively, we minimised manipulation of the visualised left GH that occupied the left abdomen. During post-operative recovery, the patient’s blood pressure returned to normotensive level, and there were no pre-eclampsia complications. Her baby had respiratory distress syndrome and was admitted to NICU for observation and was later discharged well. Four months after the caesarean section, the patient had diuretic renography with renal MAG3 (mercaptu-acetyltriglycine) scan which showed a non-functioning left kidney. Computed tomography (CT) of the abdomen and pelvis showed normal right and left renal artery vasculature and a normal right kidney. Nevertheless, the size of the left GH remained the same, with no dilated ureter or contrast demonstrated during the excretory phase. She was scheduled for left nephrectomy 12 months later, and remained asymptomatic. The surgery was delayed due to the COVID-19 pandemic.

3. Discussion

More than 500 cases of GH in adults have been reported in the literature since Sterling described it in 1939 [1,3]. The male to female ratio is 2.4 to 1, and, thus, the case is seldom encountered in pregnancy. To date, there are only a few reported cases of GH diagnosed during pregnancy in the English literature [5–8].

The majority of the reported GH occurred due to congenital obstruction of the ureteropelvic junction. The acquired causes include urolithiasis, external compression, abnormal kidney variant, iatrogenic and urethral strictures [2,3,9–11]. In the present case, the most likely cause of GH is congenital obstruction at the ureteropelvic junction, which was not detected before pregnancy. Some patients remain asymptomatic due to the slow progression of the disease and compensation by the contralateral functioning kidney. The diagnosis becomes apparent during pregnancy as physiological changes may aggravate the existing obstruction [6].

Abdominal pain and mass are the most common presentation during pregnancy, as in our case. Others may present with recurrent urinary tract infection, loin pain, haematuria or renal insufficiency [8,12,13]. Subsequently, GH can lead to the development of hypertension, cyst rupture, pain, or compression effect [3]. These signs and symptoms can be similar to the common pathophysiological effects of pregnancy. Therefore, GH can easily be mistaken for other differential diagnoses, such as ovarian tumour and hydrosalpinx [8].

Routine practice in identifying maternal kidneys when dealing with an abdominal mass in pregnancy enabled us to establish this rare pathology. This practice has been recommended by Malhotra N et al.
avoid misdiagnosis or surgical dilemma [14]. MRI is an invaluable tool to demarcate the anatomy further and confirm the diagnosis [4,15]. Ultrasound and MRI were used in all reported cases in pregnancy.

Cancer Antigen 19–9 (CA19–9) has been evaluated as a non-invasive diagnostic tool for GH in the adult population. Patients with GH have significantly raised serum and urine CA19–9 pre-operatively. The level returns to normal during post-operative follow-up [16]. However, the diagnostic value for this condition of CA19–9 in pregnancy has not been evaluated.

Upon diagnosis, multidisciplinary involvement, including urology, is warranted. Additional assessment includes; the mechanical effect of the mass towards adjacent organs, the function of the affected as well as contralateral kidney, and the overall renal function. Abnormality of the renal tract is associated with a 36% increased risk of concurrent urinary tract stone, 5.5% increased risk of synchronous renal-ureteral stones, pain, and secondary hypertension that requires timely monitoring [17]. Prophylaxis antibiotic is recommended for the recurrent urinary tract infection in pregnancy to avoid obstetric complications. Our patient did not experience any significant urological complication during the interval between diagnosis and delivery.

The association between reflux nephropathy and the development of the hypertensive disorder in pregnancy is a result of functional ischaemia promoting the removal of rennin-mediated hypertension [8,18]. This association has not been studied in GH during pregnancy. With the exception of our case, no reported cases developed a hypertensive disorder in pregnancy, although the majority of them had the risk factor of being primigravida [5–8].

Nephrectomy is the mainstay treatment for massive non-functioning renal tissue [19]. Intervention during pregnancy depends on the severity and function of the affected kidney, the function of the contralateral kidney, and symptoms or complications that arise from this pathology. Our patient was managed conservatively as she denied any significant warning signs and the fact that she had a normally functioning contralateral kidney. There are reported interventions that aim to relieve compression symptoms in order to allow for the pregnancy to progress. The other management options that have been tried with success include pyeloplasty, insertion of a ureteric stent, percutaneous drainage of the GH, and nephrectomy during the first trimester [5–8].

The timing and mode of delivery in pregnancy complicated by GH are dictated by obstetric indication. Our patient had iatrogenic prematurity delivery due to severe pre-eclampsia. However, most of the previous cases of pregnancy with GH were delivered vaginally at term with good fetal outcome. During vaginal delivery, care should be taken to avoid fundal pressure as there is a reported case of spontaneous rupture of the cyst [20]. As GH can cause significant anatomical distortion in the abdomen, surgical access during caesarean section should be planned. In our case, a paramedian incision was made contralateral to the pathological kidney. Intraoperatively, manipulation of the GH was minimised as iatrogenic cyst rupture and vascular injury posterior to retroperitoneal hydronephrosis kidney may occur [14].

In the present case, the management of an adult GH in pregnancy was presented together with a review of crucial management strategies from previously documented cases. As a standard recommendation in managing the case in pregnancy cannot be made from a single case, this paper outlines the management approach and outcome from previous case reports.

4. Conclusion

GH in a pregnancy is rare. A conservative approach or a decompression procedure are plausible options, while nephrectomy can be reserved until the postpartum period. The obstetric outcome remains good; however, care during delivery is needed to avoid potential urological complications.

Contributors

Fathi Ramly is the main author and contributed to the literature review and discussion.

Noor Azura Noor Mohamad contributed to the literature review and discussion.

Akmal Zulayla Mohd Zahid contributed to the case writing.

Norhana Mohd Kasim contributed to the image editing, and the introduction section.

Khai Yeong Teh contributed to the discussion section.

Conflict of Interest

The authors declare that they have no conflict of interest regarding the publication of this case report.

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Patient Consent

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References

[1] W.C. Stirling, Massive hydronephrosis complicated by hydro-ureter: report of three cases, J. Urol. 42 (4) (1939) 520–533.

[2] S. Ennaciri, M.H. Farhid, Giant hydronephrosis associated with ureterocele complicated by ureteral lithiasis, Pan Afr. Med. J. 31 (2018) 205.

[3] K.S. Kaura, M. Kumar, A.K. Sokhal, A.K. Gupta, B. Purkait, D. Saini, et al., Giant hydronephrosis: still a reality! Turk J. Urol. 43 (3) (2017) 337–344.

[4] Y.J. Lin, Y.C. Ou, L.C. Tsang, H. Lin, Diagnostic value of magnetic resonance imaging for successful management of a giant hydronephrosis during pregnancy, J. Obstet. Gynaecol. 33 (1) (2013) 91–93.

[5] R.L. Bernstein, G.A. Leblanc, J.F. Richardson, Giant hydronephrosis complicating pregnancy, Am. J. Obstet. Gynecol. 78 (2) (1959) 431–433.

[6] H.-H. Peng, C.-J. Wang, C.-F. Yen, C.-C. Chou, C.-L. Lee, Huge maternal hydronephrosis: a rare complication in pregnancy, Eur. J. Obstet. Gynecol. Reprod. Biol. 108 (2) (2003) 223–225.

[7] E. Hecht, Hydronephrosis complicating pregnancy, Am. J. Obstet. Gynecol. 64 (3) (1952) 684–685.

[8] R.B. Neri, A. Munargwadi, A.S. Mudegowdar, A. Patil, M.B. Hiremath, S. Ghagane, A giant hydronephrosis mistakenly diagnosed as ovarian tumor in a pregnant woman, Urol. Case Rep. 4 (2016) 20–21.

[9] R. Fakurpoto, T. Ohtoshi, K. Kobayashi, F. Akhiro, R. Inamura, Y. Tsujimoto, et al., A case report: retroperitoneoscopic nephrectomy for a giant hydronephrosis of a horseshoe kidney, Hinyokika Kiyo 55 (10) (2009) 615–618.

[10] R.A. Gaddekarareem, M.F. Abdelhafez, A.M. Moreen, A.A. Shahaat, M.M. Gadelmoula, M.M. Osman, et al., Experience of a tertiary-level urology center in the clinical urological events of rare and very rare incidence. IV. Urological surprises: 2. Clinically visible giant hydronephrosis in adults: is there a significant function? Afr. J. Urol. 26 (1) (2020) 1–6.

[11] S. Das, A.D. Amar, Ureteropelvic junction obstruction with associated renal anomalies, J. Urol. 131 (5) (1984) 872–874.

[12] A. Ardigoil, V. Vuzjec, M.K. Atikeler, E. Ozdemir, A case of adult giant hydronephrosis as unusual cause of intraabdominal mass, Int. Urol. Nephrol. 35 (1) (2003) 7–8.

[13] M. Kunimatsu, Bilateral giant hydronephrosis with acute renal insufficiency after childbirth, Japan. J. Clin. Urol. 52 (1998) 419–422.

[14] N. Malhotra, K.C. Roy, P.K. Garg, D. Takkar, Ectopic hydronephrotic kidney masquerading as an ovarian cyst during pregnancy, Eur. J. Obstet. Gynecol. Reprod. Biol. 97 (2) (2001) 239–240.

[15] J. Spencer, R. Chahal, A. Kelly, K. Taylor, I. Eardley, S. Lloyd, Evaluation of painful hydronephrosis in pregnancy: magnetic resonance urographic patterns in physiological dilatation versus calculous obstruction, J. Urol. 171 (1) (2004) 256–260.

[16] I. Banerjee, V. Tomar, S.S. Yadav, N. Vyas, S. Yadav, B. Sathan, Role of urinary and serum carbohydrate antigen 19–9 as a biomarker in diagnosis of adult giant hydronephrosis, J. Clin. Diagn. Res. 10 (9) (2016) [PCOB].
[17] A. Hemal, M. Ansari, D. Doddamani, N. Gupta, Symptomatic and complicated adult and adolescent primary obstructive megaureter—indications for surgery: analysis, outcome, and follow-up, Urology 61 (4) (2003) 703–707.

[18] R. Attini, I. Kooij, B. Montersino, F. Fassio, M. Gerbino, M. Biolcati, et al., Reflux nephropathy and the risk of pre-eclampsia and of other adverse pregnancy-related outcomes: a systematic review and meta-analysis of case series and reports in the new millennium, J. Nephrol. 31 (6) (2018) 833–846.

[19] S. Kausik, J.W. Segura, Surgical management of ureteropelvic junction obstruction in adults, Int. Braz. J. Urol. 29 (1) (2003) 3–10.

[20] S.S. Hwang, Y.H. Park, C.B. Lee, Y.J. Jung, Spontaneous rupture of hydronephrotic kidney during pregnancy: value of serial sonography, J. Clin. Ultrasound 28 (7) (2000) 358–360.