Case Report

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Bilateral Wunderlich syndrome causing Page kidney during pregnancy: a rare case report

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Introduction and importance: Wunderlich syndrome is a rare and life-threatening condition that is characterized by spontaneous renal hemorrhage into the subcapsular and perinephric regions. This case report describes the diagnosis and management of bilateral Wunderlich syndrome during pregnancy, resulting in Page kidney.

Case presentation: The patient presented with complaints of left flank pain and breathlessness. After stabilization, an emergency lower cesarean delivery was performed, and a percutaneous drainage procedure was carried out to alleviate the compression on the left kidney. The patient was treated with blood transfusion, methyldopa, and perindopril. Follow-up examinations performed 3 months later revealed a significant decrease in fluid volume surrounding the left kidney.

Clinical discussion: Lenk's triad provides the primary description of the classical manifestations of this syndrome. Some instances have been connected to the Page kidney phenomenon. The relationship between pregnancy and Wunderlich syndrome has not been extensively studied, primarily because the symptoms can resemble other complications related to pregnancy. Due to the scarcity of evidence in the literature, there is no definitive guideline for managing Wunderlich syndrome during pregnancy. Consequently, each patient is treated on an individual basis. Conservative treatment is recommended once malignancy has been ruled out.

Conclusion: The case highlights the importance of considering Wunderlich syndrome as a differential diagnosis in pregnant patients with abdominal or flank pain, a palpable mass, and hypovolemia. Furthermore, the case illustrates the successful management of Wunderlich syndrome during pregnancy.

Keywords: bilateral Wunderlich syndrome, Lenk's triad, pregnancy complications, spontaneous retroperitoneal hemorrhage

Introduction

Wunderlich syndrome is an unusual and lethal condition that is associated with spontaneous renal hemorrhage into the subcapsular and perinephric region in patients with no history of trauma[1]. Symptoms of Wunderlich syndrome can range from mild to life-threatening[2], and patients can present with a triad of symptoms known as 'Lenk's triad', which consists of acute flank or abdominal pain, a palpable flank mass, and hypovolemia[3]. It is a rare clinical finding in pregnancy, and there are no established evidence-based guidelines to manage this condition[1]. Even more rare is for it to cause the Page kidney phenomenon, which is when the kidney is compressed by a hematoma or a mass, resulting in high blood pressure, likely due to the activation of the renin–angiotensin–aldosterone system[4]. We present a case of a 23-year-old primigravida who developed bilateral Wunderlich syndrome during pregnancy, resulting in Page kidney.

HIGHLIGHTS

- It is a case of bilateral Wunderlich syndrome.
- Wunderlich syndrome caused Page kidney phenomenon.
- Wunderlich syndrome happened during pregnancy.
- The case illustrates the successful management of Wunderlich syndrome during pregnancy.
- The importance of considering Wunderlich syndrome as a differential diagnosis in pregnant patients with abdominal or flank pain, a palpable mass, and hypovolemia.

Case presentation

A 23-year-old nonsmoking woman who was over 34 weeks pregnant presented to the emergency department with complaints of left flank pain and breathlessness. She had been experiencing repeated episodes of vomiting and weight loss for the last 4 months. Her medical history was significant for celiac disease, which was diagnosed 3 years earlier by biopsy, and has been managed through a specialized diet since. No significant surgical, family, or medication history was reported.

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This case report (Bilateral Wunderlich syndrome during pregnancy, resulting in Page kidney) is very rare and uncommon due to several points highlighted in the manuscript.

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Physical examination

On physical examination, the patient complained of crampy abdominal pain. Her heart rate was 92 beats per minute, blood pressure was 160/90 mmHg, and respiratory rate was 30 breaths per minute. Additionally, she exhibited mild dehydration, likely caused by frequent vomiting, and dryness of the mucous membranes was observed. The patient's hemoglobin was 6.2 g/dL before fluids resuscitation and blood transfusion, a finding consistent with chronic hemorrhage. blood creatinine was within normal range at 0.7 mg/dL, indicating no signs of acute kidney injury resulting from dehydration. Her white blood cell count was 9200/μl, and platelet count was 127×10^3/μl. Lactate dehydrogenase was slightly elevated at 642.3 U/l. Globulin levels were normal at 2.76 g/dl, whereas albumin levels were low at 2.38 g/dl. High proteinuria was detected at 49.8 mg/dl after investigating the urine. An ultrasound of the abdomen and kidneys was performed, revealing under-capsule hemorrhage in both kidneys. Free fluid was also detected in the subcapsular region of both kidneys and in the upper abdomen. To further confirm the findings, a computed tomography (CT) scan of the chest/abdomen/pelvis that was done after the delivery showed an increased fluid volume around both kidneys compared to the ultrasound examination. The fluid significantly compressed the left kidney, and a 5–8 mm thick layer of fluid was detected in the pericardial cavity; however, the electrocardiogram results were within normal limits, indicating that the heart was not adversely affected by the accumulated fluid. A small amount of fluid was also found in the pelvis and abdomen, but no masses were detected in previous scans. Therefore, these changes were suspected to be subcapsular hemorrhage, leading to a final diagnosis of bilateral spontaneous Wunderlich syndrome that is also causing Page kidney phenomenon (Figs 1, 2).

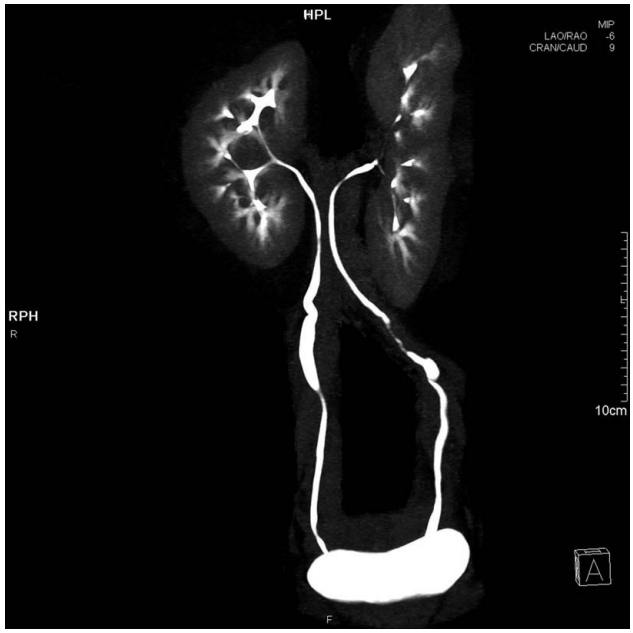

Figure 2. Computed tomography urogram reveals a mild decrease in the function of the left kidney.

Treatment

To address the patient's low levels of albumin and hemoglobin and prevent a hypovolemic shock, three units of blood were administered via transfusion. Methyldopa was also given to lower her blood pressure during pregnancy. After stabilizing her condition, an emergency lower cesarean delivery was performed. During the procedure, a drainage was placed in the Douglas pouch. Both the mother and baby were reported to be in good health. To alleviate the compression on the left kidney, a percutaneous drainage procedure was performed under real-time CT guidance. And perindopril was prescribed to lower her blood pressure.

Outcomes and follow-up

Follow-up examinations performed nearly 3 months later revealed a significant decrease in the fluid volume surrounding

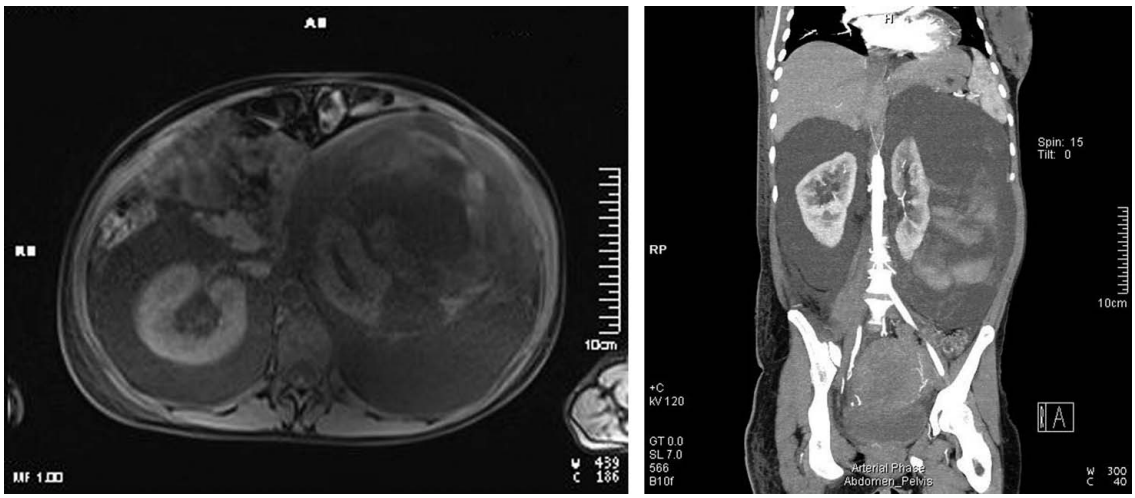

Figure 1. Depicts a contrast-enhanced computed tomographic scan revealing extensive perirenal hematomas enveloping both kidneys. On the right, the maximum thickness measures 41 mm with an upper/lower diameter of 211 mm, while on the left, the maximum thickness measures 106 mm with an upper/lower diameter of 305 mm. These hematomas cause compression of the adjacent kidney tissue.

the left kidney from 3500 to 600 ml. This fluid completely disappeared over the next few days, along with hypertension, and the patient's general condition improved and stabilized. However, ~400 ml of fluid remains around the right kidney and may require drainage in the future if conservative treatment is not effective.

Discussion

The case report presented a rare and potentially life-threatening medical condition known as bilateral Wunderlich syndrome, which occurred during pregnancy and resulted in Page kidney.

Wunderlich syndrome is characterized by spontaneous renal bleeding into the subcapsular and perinephric space in patients with no history of trauma. The condition is named after Carl Wunderlich, a German pathologist who first described it in 1856[1,3].

The classical manifestations of this syndrome are mainly described in Lenk's triad as insidious abdominal or flank pain, a palpable tender mass, and symptoms of internal bleeding. Furthermore, vomiting, nausea, hematuria, anemia, and hemodynamic instability leading to hypovolemic shock, are all possible presenting symptoms[5]. In addition, a few cases have been associated with the Page kidney phenomenon, which leads to secondary systemic hypertension due to extrinsic compression of the renal parenchyma that activates the renin–angiotensin–aldosterone system[4].

The most common cause of spontaneous retroperitoneal hemorrhage is renal angiomyolipoma. However, there are various other causes such as malignant and benign tumors, polyarteritis nodosa, renal infections, underlying hematological conditions, and, less frequently, renal cysts, blood dyscrasias, or anticoagulant therapy[2]. Yet, occurrences with no apparent influencing factors can also be found[5].

Currently, there is limited research on the association between pregnancy and Wunderlich syndrome[1], as symptoms can mimic other pregnancy-related complications, such as preeclampsia, placental abruption, and uterine rupture. Therefore, clinicians should consider Wunderlich syndrome as a potential cause of abdominal pain and shock in pregnant women, especially those without risk factors for other pregnancy-related complications. Similarly, when establishing a differential diagnosis for atrial hypertension during pregnancy, Page phenomenon should be taken into consideration. Further studies on these matters are highly recommended.

A solid diagnosis and characterization using multidetector CT or MRI is essential to permit optimal patient management[3].

The management of Wunderlich syndrome during pregnancy lacks a definite guide due to the scarcity of evidence in the literature. Therefore, each patient is dealt with independently[1]. Decisions must be made after reviewing the patient's clinical status, laboratory findings, hemodynamic stability, and degree of kidney rupture, as well as the size of any tumors that may be present[1,2].

Unstable cases may require surgical procedures such as nephrectomy to be performed. Moreover, minimally invasive options such as nephron-sparing surgery and selective arterial embolization should be considered[2].

For clinically stable patients who respond to fluid resuscitation and have a self-limiting hemorrhage, conservative treatment is the appropriate approach after malignancy has been excluded[2]. In the current study, we opted to terminate the pregnancy out of concern for potential kidney damage. We also administered ACE inhibitors as the optimal drug to lower blood pressure in Page kidney and relieved the compression of the kidney using CT-guided drainage. It is important to note that active monitoring and care by a nephrologist is recommended in this scenario[1].

This work has been reported in line with the CARE criteria[6].

Conclusion

This rare case of bilateral spontaneous renal hemorrhage highlights the importance of repetitive imaging and effective minimally invasive treatment, including timely percutaneous drainage and supportive therapy. The case also emphasizes the need for conducting more research on this subject.

Ethics approval

This retrospective review of patient data did not require ethical approval in accordance with local/national guidelines.

Patient consent

Written informed consent was obtained from the patient for the publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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Author contribution

S.A.: supervised and helped in writing the manuscript; Z.A.A.A., B.R.J., D.B., and M.H.H.: wrote the manuscript; Z.A.A.A.: critically revised the manuscript. All authors read and approved the final manuscript.

Conflicts of interest disclosure

The authors declare that they have no conflicts of interest.

Research registration unique identifying number (UIN)

Not applicable.

Guarantor

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Data availability statement

All data generated or analyzed during this study are included in this published article and its supplementary information files.

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