Spontaneous Rupture of Renal Cell Carcinoma in Pregnancy, Surgical Management With Fetal Preservation:A Case Report

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Spontaneous Retroperitoneal hemorrhage in pregnancy is a rare condition. Renal angiomyolipoma (RA) is the most common cause of this hemorrhage. To the best of our knowledge, this is the first reported case of Wunderlich syndrome (WS) due to renal cell carcinoma (RCC) diagnosed in the second trimester of pregnancy.

Keywords: nephrectomy; pregnancy; renal cell carcinoma

INTRODUCTION

Wunderlich syndrome with spontaneous non-traumatic retroperitoneal hemorrhage in pregnancy is a rare condition which is clinically characterized by Lenk's triadas, acute flank pain, flank mass, and hypovolemic shock.(1) The incidence of cancer during pregnancy is rare and occurs in approximately 1:1000 pregnancies.(2) The most common urological cancer during pregnancy is RCC.(3)
Spontaneous rupture of the kidney is very rare as the first presentation of RCC and to the best of our knowledge this is the first report.(4) We report a case of spontaneous rupture of the renal tumor in pregnancy with retroperitoneal hemorrhage and hypovolemic shock as the first manifestation of RCC that was treated by nephrectomy .

CASE REPORT

A 20 year old woman in the 20th week of pregnancy (Gravid 1, para1) referred to our center with severe generalized abdominal pain and flank pain. She described a history of 8 hours of severe, constant pain with sudden onset and radiation to the right flank that progressively aggravated. She did not have any predisposing factors or history

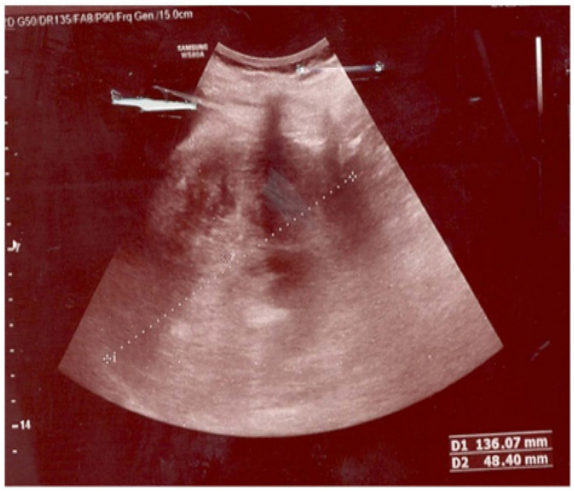

Figure 1. Ultrasound showed a 136*48 mm heterogeneous collection.

Figure 2. Kidney with site of rupture at upper pole(Arrow).

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Received November 2017 & Accepted February 2018

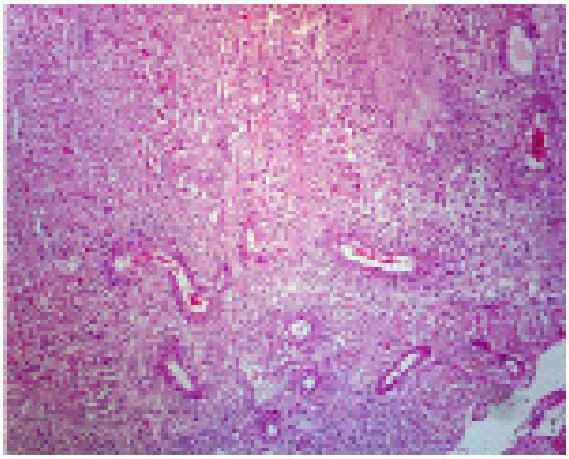

Figure 3. Clear cells and areas of sarcomatoid feature (H&E original magnification x40)

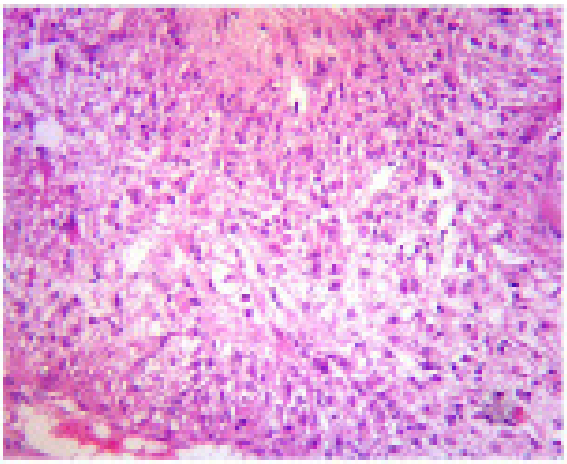

Figure 4. Transitional areas between clear cells and spindle shape ones. (H&E original magnification x100)

of trauma, did not have previous medical disease, did not take any medication and was non-smoker. She was referred to our center in a shock state. BP: 80/55, PR: 110, RR: 18; also, she was febrile (oral temperature: 38.5c)

Blood investigation such as (complete blood cell, electrolyte profile, liver function test and coagulation screen) was done, showing only a drop in the serum hemoglobulin from 6.6 g/dl to 5.5 g/dl after just one hour with a decrease in the concentration of hematocrit level from 33.5% to 21.7%. Urine sediment showed many RBC and 12-4 WBC with little bacteruria. Resuscitation with intravenous fluid and 3 units of crossed match packed cell was done immediately. Abdominal ultrasound showed a 136*48 mm heterogeneous collection in the anterior right pararenal space (**Figure 1**). Spontaneous rupture of the renal artery aneurysm and rupture of a renal mass were at the top of differential diagnoses. Written informed consent was taken before the operation and the patient was transferred to OR within 4 hours after admission. Midline abdominal incision was done and after mobilization of the right colon and duodenum, a large hematoma was evacuated and the right kidney was exposed. A large renal mass between the mid to upper pole was seen; also, significant fresh bleeding was seen through the ruptured site of the mass, so due to hemodynamic instability, early clamping of the pedicle was done and simple nephrectomy performed. It should be considered that radical nephrectomy was not done due to the possibility of renal angiomyolipoma. The patient was admitted in the intensive care unit for close observation. The patient did well post-operatively and was discharged one week later.

The pathology report revealed a 14x6x3.5 cm, which was slightly distorted with an area of rupture at the upper pole (**Figure 2 arrow**). The cut section of the organ revealed a well-defined ovoid mass with variegated cut surface.

Microscopic examination of the tumor showed sheets of cells with moderate amounts of clear to eosinophilic cytoplasm and nuclei with nucleoli visible at 100x magnification, as well as areas in those tumor cells which became spindle shape (**Figures 3, 4**). Operative time was 1.5 hours and total transfusion was 5 crossed match red packed cell and 3 platelet.

DISCUSSION

Wunderlich syndrome is a rare clinical disorder with spontaneous non-traumatic kidney rupture, significant retroperitoneal hemorrhage with sudden onset or flank pain, palpable flank mass, and hypovolemic shock with or without hematuria.[1,5] The first description of these symptoms was in 1856 by a German physician called Carl Reinhold August Wunderlich.[1] The etiology of Wunderlich syndrome may be neoplastic, as the most common cause (angiomyolipoma, RCC), or non-neoplastic, such as vascular causes (renal artery aneurysms, AV malformations, renal vein thrombosis), cystic renal diseases, calculus diseases, nephritis, and coagulation disorders.[6] The current management options range from observation and minimally invasive measures, such as embolization, radiofrequency ablation, and cryoablation to partial or radical nephrectomy.[7]

There is no a guideline in favor of either approach. The choice of the management method is influenced by the patient's condition, availability, expertise, and surgeon judgment. Maternal safety should always be a priority although management should be performed based on the patient's wish regarding having a child. The management should be based on hemodynamic status, week of gestation, requirement of blood transfusion and association with tuberous sclerosis.[8]

The patient in our study was hemodynamically unstable; huge hematoma and renal mass was observed during the operation, so nephrectomy was necessary to save her life.

The safe radiological modality in pregnant women is ultrasonography with no teratogenicity or carcinogenesis.[9] Ionizing radiation from the CT scan results in increased risk of cancer in the mother and fetus. It results in teratogenic effect as to the fetus at high up to 15 weeks post-conception.[10] However, in our case due to severe hemodynamic instability and unsuccessful aggressive intravenous hydration and progressive drop of hemoglobin, it was not possible to proceed for angioembolization, or even extend the diagnostics to MRI or CT scan and the patient was indicated for urgent operative exploration.

ACKNOWLEDGEMENTS

The authors would like to thank Shiraz University of Medical Sciences, Shiraz, Iran and also Center for Development of Clinical Research of Nemazee Hospital and Dr. Nasrin Shokrpour for editorial assistance.

CONFLICT OF INTEREST

The authors have no conflict of interest declare.

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