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GENERAL UROLOGY

Case Report

Renal artery aneurysm in pregnancy presenting as an arteriovenous fistula: an uncommon presentation

Gebelikte arteriyovenöz fistül olarak gelen renal arter anevrizması-alışılmadık bir prezantasyon

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ABSTRACT

Renal artery aneurysms (RAAs) are uncommon. RAA is generally an incidental finding; however, in certain instances, it may be a devastating pathology. This is particularly true in case of pregnant females where the incidence of rupture is high if untreated, with high mortality rates for both the mother and the fetus. Early intervention in this particular high-risk group is advocated.

Key words: Aneurysm; pregnancy; renal artery.

ÖZET

Renal arter anevrizmaları (RAA) nadirdir. Tanımlandığı zaman, bu genellikle rastlantısal bir bulgudur, ancak bazı durumlarda, tahrip edici bir patoloji olabilir. Bu özellikle hamile kadınlarda böyledir, eğer tedavi edilmezse rüptür insidansı yüksektir ve hem anne hem de fetüs için yüksek mortalite hızları ile birliktedir. Bu problemden kaçınmak için, bu özel yüksek-risk grubunda erken müdahale önerilmektedir.

Anahtar kelimeler: Anevrizma; gebelik; renal arter.

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Introduction

Renal artery aneurysms (RAAs) are rare and account for 0.01%-0.5% of all aneurysms,[1] with an incidence of approximately 0.09%.[2] RAA is generally an incidental finding, although the incidence may increase as more people undergo imaging studies. The management of this pathology remains controversial. In many patients in whom it is identified incidentally, treatment should be considered if the size of the aneurysm is >2 cm.[3-8] Symptomatic patients are also offered treatment. The majority of patients are females. This particular group is at a higher risk of aneurysm rupture, particularly in pregnancy. The mortality rate due to ruptured RAAs in the general population is approximately 10%; however, in pregnant women, the mortality rate for the mother and the fetus can be as high as 50% and 80%, respectively.[9] Thus, in young women of childbearing age, surgical or endovascular repair of RAAs is recommended irrespective of the size of the aneurysm.[10,11] This is even more important in rare instances of solitary kidneys with RAAs.[12] The natural history of this condition in pregnancy involves progressive weakening of the arterial wall, as a result of the effect of increased circulating estrogens, and hyperdynamic circulation with increased cardiac output leading to further weakening and eventual rupture.[10] Leakage and subsequent development of an arteriovenous fistula (AVF) has been postulated to occur but has not been reported in the modern literature.

Case presentation

We present the case of a young 30-year-old woman (gravida 3, para 2) who initially presented to a tertiary center at 14 weeks of pregnancy complaining of right-sided abdominal pain with mild fever. She was assessed and a diagnosis of acute appendicitis was made. She underwent emergency surgery via a Lanz incision under general anesthesia. Intraoperatively, the appendix was noted to be normal. However, retroperitoneal hematoma

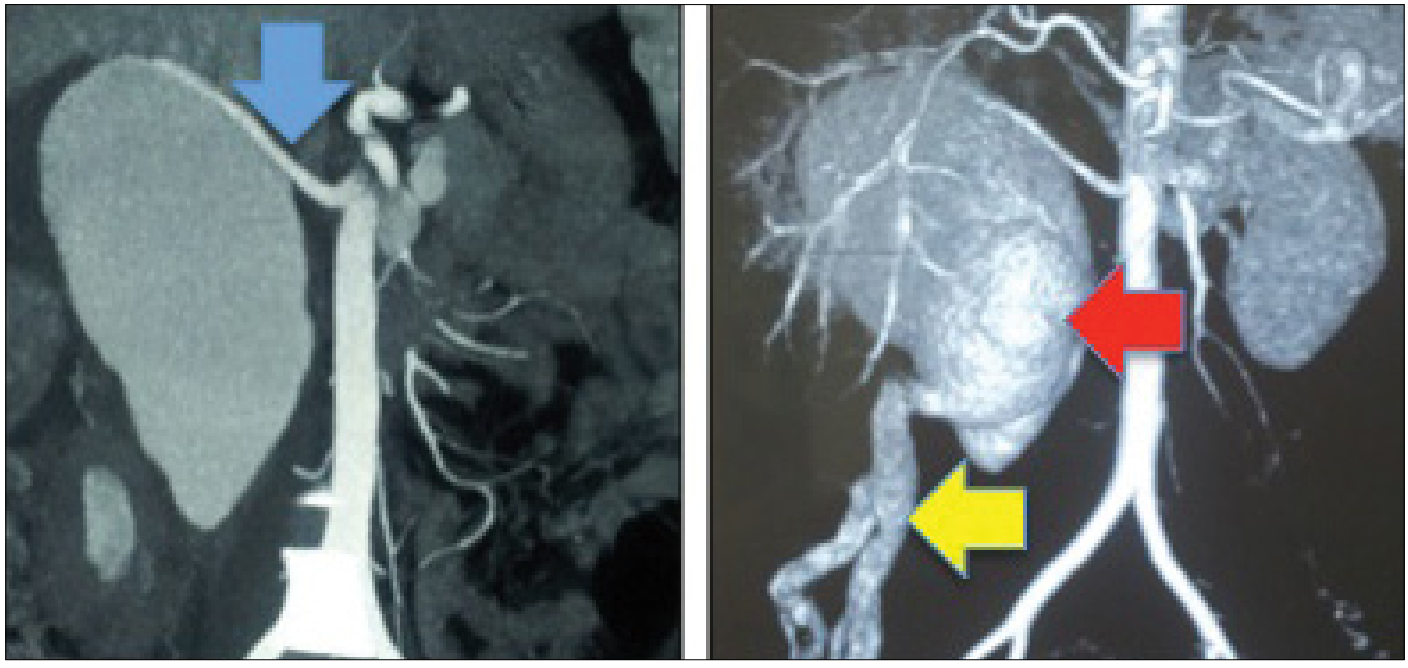

Figure 1. Right renal artery pseudoaneurysm showing the right renal artery (blue arrow), the aneurysm (red arrow), and the draining fistula through the right ovarian vein (yellow arrow)

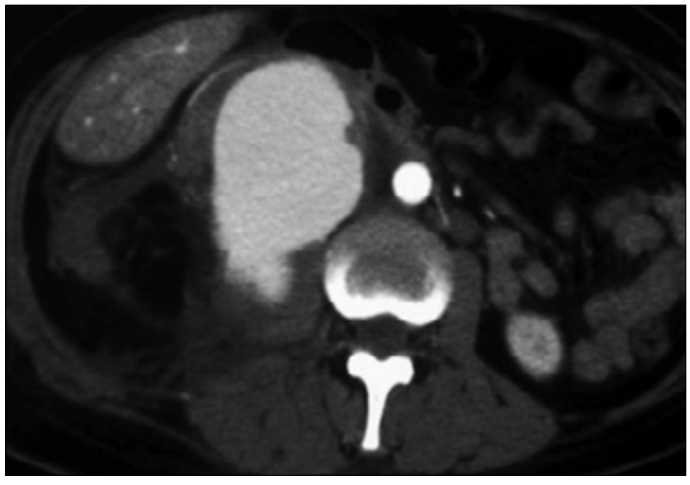

Figure 2. Axial CT angiogram showing right renal artery aneurysm
CT: computed tomography

was observed over the right side. Appendectomy was performed and the surgery was completed. Postoperatively, the patient recovered well and underwent ultrasonography (USG) of the kidneys, which showed a small right RAA measuring 2.0 x 2.6 cm. The patient's hemoglobin levels were 11.6 g/dL and her renal function was normal. She denied any prior trauma and her previous pregnancies were normal with no hypertension. The primary team at that hospital decided to manage her conservatively and to wait for her pregnancy to come to term and allow delivery before attempting any repair. She was warned to come back immediately if there was any sudden onset of pain or hematuria. She was subsequently discharged home well at 16 weeks of pregnancy. No Doppler USG was performed at that point nor was there subsequent follow-up at the primary center after the obstetrician consulted the vascular surgeon at the national vascular referral center.

However, she again presented to the same tertiary hospital at 31 weeks of pregnancy, 1 day prior to visiting our institution, complaining of sudden onset of pain that worsened progressively. She had no hematuria. Her hemoglobin levels were 8 g/dL. She was clinically pale with a pulse rate of 90 beats/min and normal blood pressure.

Urgent Doppler USG of the kidneys was performed, revealing a large (10 x 10 cm) right RAA with turbulent prominent flow in a vein medial to the right kidney. The patient was then referred to our institution for further combined multimodal management. After discussion with the pediatrician, obstetrician, and urologist, the patient and her family agreed for a semi-emergent cesarean section on the following day after optimizing the patient's hemoglobin levels with blood transfusion. A baby girl was delivered with a good Apgar score (8 at 0 min and 9 at 5 min). The patient underwent CT angiography postoperatively, which revealed a large right RAA that had eroded into the right ovarian vein leading to the formation of an AVF (Figure 1-3). Further discussion with the interventional radiologist ruled out the possibility of embolization of the fistula and aneurysm.

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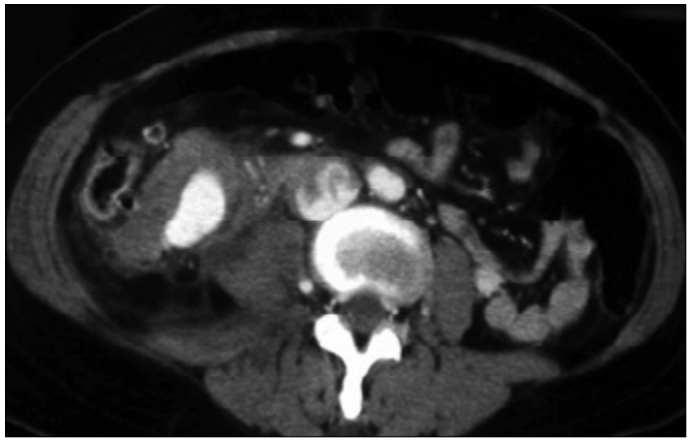

Figure 3. Axial CT angiogram (lower cuts) showing flow through the right ovarian vein suggestive of a right arteriovenous fistula
CT: computed tomography

Because of the size and presentation of the aneurysm, it was decided that she required emergency nephrectomy.

The patient underwent right nephrectomy on the same day. Intraoperatively, the right kidney was grossly enlarged and pulsating. The right ovarian vein was grossly dilated and also pulsatile. There were numerous adhesions, particularly to the inferior vena cava and at the region of the hilum. Identification of the right renal artery (which was pushed away superiorly and thinned out) was difficult. Eventually, with the help of the vascular surgeon, the right kidney was removed and the right ovarian vein was ligated and cut.

The patient required a total of 6 pints of blood and transfusion of other blood products intraoperatively and postoperatively. She recovered well postoperatively. The baby was well and both mother and child were discharged 2 weeks later. Postoperatively, creatinine levels remained normal and the blood pressure continues to be within the normal range without any medication.

Discussion

Pregnancy-related development of RAAs has been well documented in the literature but still remains uncommon. Management guidelines are available in terms of the timing of intervention in this select group of patients. However, in the present case, the decision of not intervening at the initial presentation, though seemingly justified at that time, led to progressive dilatation and the likelihood of erosion into the right ovarian vein, leading to the formation of an AVF.

Early intervention may have prevented the development of this complication, which nearly resulted in the patient's death; however, the justification for all patients to undergo surgery in pregnancy with an aneurysm size of 2 cm may be a little premature and even possibly dangerous to both the mother and the unborn child. It is, however, generally recommended that women of childbearing age who are diagnosed with RAAs of ≥2 cm should undergo surgical or endovascular treatment to avoid a scenario such as that encountered in this patient.[10-12]

Some researchers have advocated termination of pregnancy and repair of the aneurysm if it is diagnosed in the first trimester of pregnancy.[13] However, if it is diagnosed later in pregnancy, decision-making becomes more difficult, particularly in patients who do not have a life-threatening rupture at presentation. The question remains on how to monitor the aneurysm's progression through the pregnancy. Possible suggestions include more frequent USG or Doppler USG, i.e., monthly or fortnightly. This may enable calculation of the rate of enlargement or progression to estimate the risk of rupture.

In the present case, the patient may have been fortunate that her aneurysm enlarged and eventually eroded into the right ovarian vein rather than rupture and cause immediate exsanguination with possible death for her and her unborn child. However, this particular occurrence was possibly by chance and will very likely not happen in another patient with an almost similar presentation.

An attempt to embolize the feeding artery, i.e., the right renal artery, may have been an option if it was not due to the large size and nature of the aneurysm that had developed into a large AVF. If this had been successful, it would have reduced the amount of blood loss intraoperatively. In some centers with early detection, angioembolization would have been the only treatment required.[14] Successful treatment of ruptured RAAs in pregnancy using angioembolization is rare, with only 2 cases reported in the literature, the most recent being in 2009.[15]

Peripartum diagnosis (either after cesarean section or vaginal delivery) is also extremely rare and has so far been documented in only 3 cases. The most recent case reported a ruptured RAA, diagnosed 1 day after successful cesarean section and successfully treated with simple nephrectomy.[16]

In conclusion, RAAs in pregnancy are uncommon. Management during pregnancy remains a challenge for the treating physician as well as a dilemma for the patient due to the high risk of maternal and fetal death. An early combined discussion among obstetricians, vascular surgeons, urologists, and the patient must be initiated. An individualized close follow-up plan with frequent Doppler USG must be initiated. Early surgery during the second trimester may be advantageous and must be discussed in a multi-disciplinary meeting involving the patient. Interventional radiological embolization may be an option in

select individuals. The chances of another patient developing a similar "escape mechanism" of an AVF instead of direct rupture is unlikely and very remote; thus, early intervention after counseling may be a safer option.

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