Embolization of renal angiomyolipoma in pregnancy: case report

Przypadek embolizacji angiomyolipoma nerki w ciąży

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Summary

Background: Renal angiomyolipoma is a rare benign tumour composed of adipose tissue, blood vessels and smooth muscles. However it can locally grow to a great size and its numerous blood vessels may cause major bleeding requiring immediate intervention.

Case: At 20th week of pregnancy a previously healthy 26-year old pregnant woman with an episode of sudden and severe pain in the left flank followed by fainting was diagnosed with a bleeding tumour of the left kidney. The diagnosis was based on ultrasonography and magnetic resonance imaging (MRI). Diagnostic angiography was followed by selective embolization of the tumour blood vessels. At 38th week of pregnancy elective caesarean section was performed and after the puerperium the tumour was resected.

Conclusion: Embolization of renal angiomyolipoma bleeding vessels during pregnancy can be an effective therapeutic approach protecting against further bleeding and haemorrhagic shock thereby obviating the need to perform urgent surgery and allowing the woman to carry her pregnancy to term safely in outpatient setting.

Key words: angiomyolipoma / pregnancy / bleeding /

Streszczenie

Wstęp: Angiomyolipoma nerki jest łagodnym, rzadko występującym guzem, zbudowanym z tkanki tłuszczowej, naczyń i mięśniówki gładkiej. Może on jednak osiągać duże rozmiary, a obecność licznych naczyń może być przy czyną obfitych krwawień wymagających natychmiastowej interwencji.

Przypadek: U 26-letniej, dotychczas zdrowej ciężarnych 26-letniej ciężarnych z epizodem nagłych silnych bólow w lewej okolicy lędźwiowej i zasłabnięciu, na podstawie badania ultrasonograficznego i rezonansu magnetycznego (MRI) rozpoznano krwawiący guz lewej nerki. Po angiografii diagnostycznej wykonano selektywną embolizację naczyń guza. W 38 tygodniu przeprowadzono planowe cięcie cesarskie, a po okresie połogu usunięto guz.

Wnioski: Embolizacja krwawiących naczyń angiomyolipoma nerki w ciąży może być skutecznym postępowaniem leczniczym, które chroni przed dalszym krwawieniem i wstrząsem krwotocznym, a zarazem koniecznością wykonywania naglej operacji i umożliwia bezpieczne donoszenie ciąży pod kontrolą ambulatoryjną.

Słowa kluczowe: angiomyolipoma / ciąży / krwawienie /

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Introduction

Renal angiomyolipoma is a benign tumour composed of adipose tissue, blood vessels and smooth muscle elements. The incidence of this tumour is low (0.24% in general population) [1]. 1/5 of angiomyolipoma tumours are diagnosed in patients with tuberous sclerosis. These tumours are large, multiple, often bilateral and affect both sexes almost equally. However great majority i.e. 80% of all angiomyolipoma cases are isolated tumours of varied size, several times more frequent in women than in men, usually located on the right side. Angiomyolipoma does not have a capsule but is usually well separated from the kidney, contains varyingly abundant adipose tissue which is typical of this tumour, and has rich vascular supply. The blood vessels are thick and tortuous because their wall is devoid of elastic fibers, which results in greater susceptibility to aneurysm formation and spontaneous blood extravasations. In general the tumour growth is slow but pressure caused by the tumour may lead to the kidney damage. Usually the tumour grows outwards exceeding the capsule of the kidney and spreading into the perirenal space. Hormonal and haemodynamic changes occurring during pregnancy are very likely to stimulate rapid growth of angiomyolipoma, which is associated with increased risk of bleeding from the tumour [2]. Individual cases of the tumour expansion into the renal vein and inferior caval vein have been reported [3]. 60% of the lesions are asymptomatic and are incidental findings on ultrasound images (incidentaloma). If their diameter is smaller than 4cm they seem to require only periodical evaluation with an imaging examination. In diagnosis of angiomyolipoma various imaging techniques are used including plain abdominal radiography, ultrasonography, computed tomography, intravenous urography, magnetic resonance imaging, arteriography and isotope renography with DMSA. A very characteristic sonographic feature of most angiomyolipoma tumours is their increased echogenicity caused by the presence of fat tissue, which is not found in a normal kidney, except renal sinus. Obviously tumours with low fat content and with blood extravasations and necrotic lesions look differently. Unfortunately the presence of adipose tissue is not pathognomonic for angiomyolipoma as the adipose tissue can also be found in renal cell carcinoma, lipoma, liposarcoma, teratoma and Wilms tumour [4, 5].

Growing lesions can manifest with flank pain and haematuria whereas bleeding into the tumour or into the retroperitoneal space is life-threatening and requires immediate surgical intervention involving resection of the tumour or even the whole kidney. Embolization of the tumour blood vessels can be equally effective alternative to stop bleeding.

Case

A 26-year old primigravida, who was 20 week pregnant suddenly experienced severe pain in her left flank followed by fainting and was admitted to the Department of Obstetrics and Gynaecology. An ultrasound examination showed a heterogeneous mass 155x110x75mm large in the left retroperitoneal space, which originated from the posterior part of the kidney and displaced the kidney to the front. The ultrasound image of the tumour was described to contain subcapsular, 25mm thick, hypoechogenic tissues and tissues of similar echogenicity, 99x65x21mm large, located centrally. The right kidney was found to be free of abnormal lesions and typically located. Magnetic resonance imaging of the kidneys was performed in a biphasic mode and T1 and T2-weighted images in axial and coronal planes were obtained. The MRI scans depicted a large tumor (120x124x174mm) located in the retroperitoneal space at the posterior margin of the kidney, with smooth outer margin and heterogeneous structure, modelling blood vessels of the left kidney although with no signs of abnormalities in the left renal vein or inferior caval vein. The tumour occupied the left periaortal space, most probably originated from the left kidney and was devoid of typical features of renal cell carcinoma. Hyperintense areas seen on T1-weighted images suggested considerable amounts of fatty tissue in the tumour. Peripherally, on the left side of the tumor there was a 12mm thick area that might correspond to the...
bleeding site. In the remaining areas of the kidney there were two further, small (10mm) foci hyperintense on T1-weighted images. The left suprarenal gland was invisible, compressed by the tumour. Laboratory test results showed moderate anaemia (Hb - 9.4 g/dl, Ht - 29.3%, RBC - 3.40 M/uL), renal function parameters in normal range (creatinine - 0.6mg/dl), K+ - 4.9mmol/dl, Na+ - 138mmol/dl and no abnormalities in urinalysis. An examination by an Obstetrician confirmed normal pregnancy development. On subsequent days increase in severity of anaemia was seen (Hb – 8.6g/dl, Ht -27.4%, RBC – 3.10M/μL). Having the patient consulted with an Urologist a decision was made to perform embolization of the tumour blood vessels. The first step was a diagnostic angiography which showed a hypervascular tumour with severe arteriovenous shunting, aneurysmally dilated interlobal vessels and a network of small abnormal blood vessels located peripherally. Then embolization with medium-sized PVA particles (500) was performed with superselective catheterization of blood vessels supplying the tumour. One of the upper interlobal arteries which supplied normal renal parenchyma was kept intact during the embolization procedure. Three days after the procedure the patient in good condition was discharged from hospital. Follow-up ultrasound images of the kidneys showed no changes in size and location of the tumour. The patient was followed-up in out-patient setting up to the 38th week of pregnancy, when she was qualified for elective caesarean section. During the caesarean section the left kidney was evaluated – it was markedly enlarged but smooth and movable. After the puerperium the patient was admitted to the Urology Department and underwent computed tomography of the kidneys which showed no changes to the tumour. Technical aspects of the surgical procedure resulted in the decision to resect the tumour together with the whole left kidney. Histopathologic examination confirmed the diagnosis of angiomyolipoma; in this case the tumour had a well-marked capsule while the renal parenchyma and the ureter were free of any significant lesions.

Comment

The reported case involved a rare renal tumour - angiomyolipoma which accompanied pregnancy. The tumour was diagnosed at 20th week of gestation, when bleeding from the tumour blood vessels to the tumour and retroperitoneal space occurred which manifested with severe pain in the flank and development of anaemia in previously healthy pregnant women. Considering stage of pregnancy surgical resection of the tumour was not undertaken but instead a less invasive approach involving embolization of the tumour vessels was chosen. This approach was highly effective – the bleeding was stopped, the pain subsided within a few days and moderate anaemia was treated with administration of oral iron products. The pregnancy developed well and ultrasound images of the tumour showed no changes.
In literature we met with very few reports of bleeding to renal angiomylipoma during pregnancy. These patients usually underwent immediate laparotomy and nephrectomy, often with simultaneous caesarean section, if the foetus was viable. When conservative management was appropriate the baby was delivered by caesarean section in order to avoid the risk of haemorrhage into the tumour during normal delivery. Although individual cases of normal delivery after spontaneous inhibition of angiomylipoma bleeding were reported [6, 7], due to scarce data on safety of this approach found in medical literature we did not make such a decision.

References