

RENAL ANGIOMYOLIPOMA WITH RENAL VEIN EXTENSION

Goran Arandjelović, Claudio Milani

Department of Urology, Saint Giovanni e Paolo Hospital, Venice, Italy

A 31-year old female patient was diagnosed with small right renal mass with renal vein extension immediately after her caesarian delivery. Open right radical nephrectomy was performed with tumor thrombus removal. The histological findings showed simple angiomyolipoma. We present a very rare case of a small, plain angiomyolipoma with renal vein thrombus invasion and we discuss the possible causes and therapeutical options *Acta Medica Medianae* 2015;54(1): 48-52.

Key words: angiomyolipoma, renal vein extension, radical nephrectomy

Contact: Goran Arandjelović
Saint Giovanni e Paolo Hospital, Venice, Italy
E-mail: gorana_4@yahoo.it

Introduction

Angiomyolipoma (AML) represents the most common benign tumor of the kidney (1). It is a typically solid lesion that may also occur in the liver, ovary, fallopian tube, spermatic cord, palate and colon (2). It is composed of adipose tissue, smooth muscle cells and blood vessels. Until recently it was believed to belong to a family of lesions characterized by proliferation of perivascular epithelioid cells (1). In 20% of cases it is associated with tuberous sclerosis which is a genetic disorder that results in hamartoma formations in many organs (3,4). In those cases they are usually bilateral, multiple and small (5). AMLs can also occur in association with von Hippel Lindau disease, Sturge-Weber syndrome, von Recklinghausen disease, polycystic disease or pulmonary lymphangiomyomatosis (6,7). Its prevalence in the general population has been reported to be 0.3-3% in the female patients (8). The incidence has been found to be 0.1% in males and 0.2% in females, in the population without tuberous sclerosis (10).

Case presentation

We present the case of a 31-year-old woman who came to our observation through the gynecological department where she was previously treated for preeclampsy. After her caesarian delivery in the 40th week of gestation, she was readmitted to the hospital for breast feeding of her newborn son. A control renal ultrasound followed

by computed tomography (CT) scan of abdomen showed a right renal mass of 2cm in diameter, partially involving the renal pelvis with the tumor thrombus invading the entire renal vein to its confluence with inferior vena cava. (Figure 1, Figure 2). Magnetic resonance imaging (MRI) confirmed the diagnosis (Figure 3, Figure 4). Chest X-ray was negative. All biochemical parameters were in range including the renal function parameters.

Open right radical nephrectomy was performed with removal of tumor thrombus and anterior cavotomy.

The postoperative course was free of complications and the patient was discharged after 7 days.

The histologic findings showed angiomyolipoma with invasion of entire renal vein to the confluence with inferior vena cava and renal sinus and perinephric fat invasion. All surgical margins were free of tumor.

Immunohistochemistry showed weak human melanoma black (HMB-45) positivity.

Discussion

Due to uncertainty over their natural history, the gold standard management of AMLs is under debate. Factors affecting the treatment decision include symptoms, size, visible aneurysms, renal function, risk of rupture, patient compliance (11). Management of AMLs primarily consists of active surveillance if they are small and asymptomatic (2). Mostly the lesion of less than 4cm in diameter are managed conservatively, those greater than 4cm should be monitored closely and more than 50% of patients will undergo surgery (5,9). Although angiomyolipomas are typically benign lesions, they can -



Figure 1: Computed tomography showing renal angiomyolipoma with renal vein extension

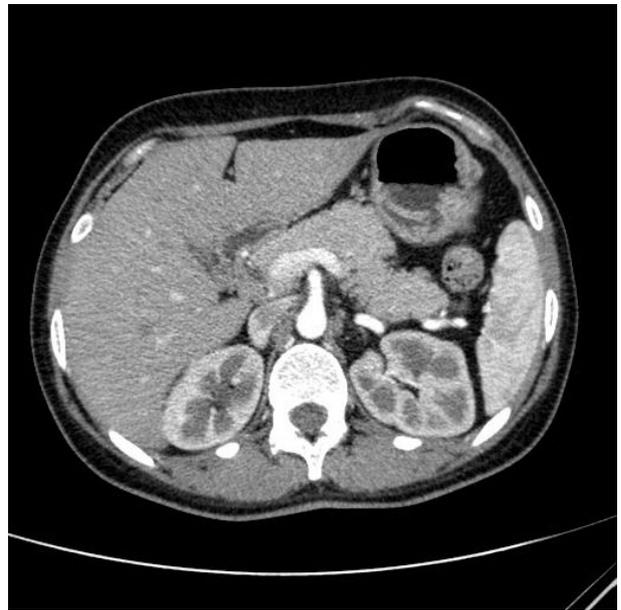


Figure 2: Computed tomography showing renal angiomyolipoma with renal vein extension

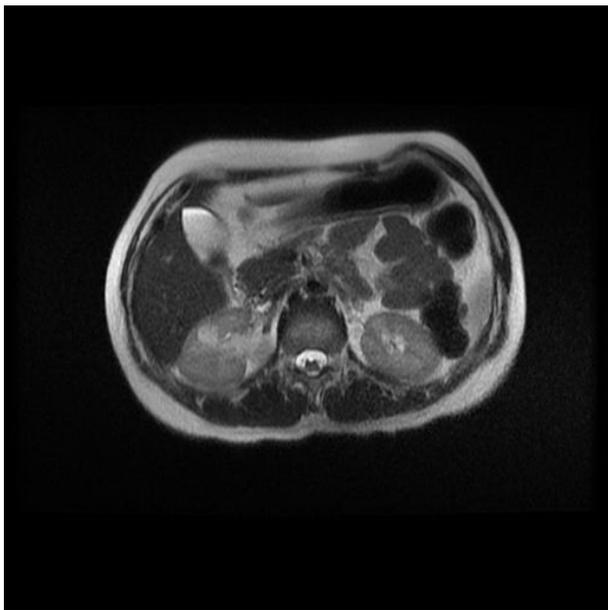


Figure 3: Magnetic resonance showing renal angiomyolipoma with renal vein extension

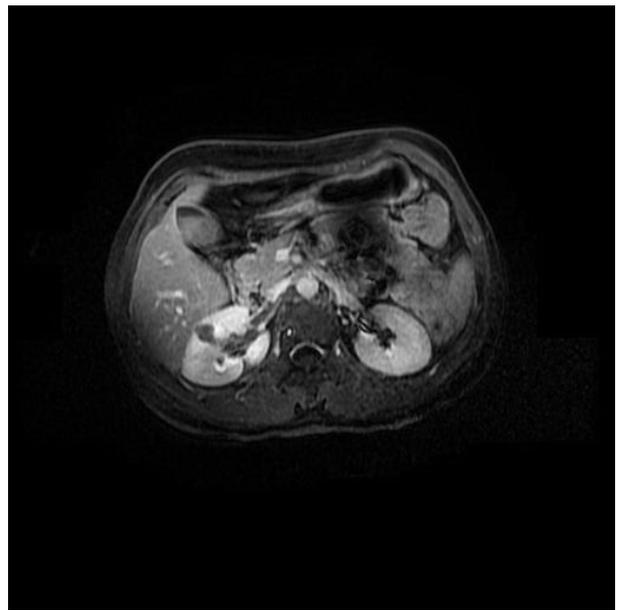


Figure 4: Magnetic resonance showing renal angiomyolipoma with renal vein extension

coexist with malignant lesions such as sarcoma and renal cell carcinoma (12), and they some times show extension into the renal vein and inferior vena cava (IVC). This is thought to be due to multifocal growth rather than metastasis (13). The formation of aneurism, facilitated by the fragility of blood vessels, might cause intraveneous extension (5). The only type of angiomyolipomas with capability of recurrence and metastasis is represented by epithelioid angiomyolipoma (EAML) (14). Approximately one third of EAML have been reported to develop metastasis. Eble at al. (15) reported 40 cases of AMLs with regional lymph node involment. This type of AML has been associated with aggressive

clinical behavior including extension into the vena cava or renal vein (10). The most common presentation is given by flank pain, although a quarter of the patients are diagnosed as a result of incidental findings (5,16). In 80% of the cases it involves the right kidney (3).

Though our histological findings showed simple angiomyolipoma with rather small dimensions, the tumor thrombus was present. It did not invade the inferior vena cava, but extended right to its confluence with the superior right renal vein. Therefore, the cavotomy had to be performed in order to avoid the incomplete thrombus removal and prevent thrombus dissemination with potential fatal cardiopulmonary embolism and death. Using tourniquets, vascular control of the inferior vena

cava and the left renal vein was achieved, and by performing the anterior cavotomy the tumor thrombus was removed intact and the cavotomy repaired. Thus the right radical nephrectomy, which had to be performed due to the central pelvic position of the tumor, was carried out safely.

Because of potentially fatal risks of AMLs with thrombi invading either the renal vein or IVC, the surgical approach of those lesions is indicated also when they are asymptomatic. Generally, AMLs larger than 8cm should be treated at the time of presentation, given that the size of tumor is directly correlated with the risk of rupture (17,18).

The patient, in our case, was diagnosed immediately after her pregnancy. Given the short period between the pregnancy and the diagnosis, it is reasonable to presume that the renal lesion was present also during the pregnancy. Case reports have demonstrated rapid growth of AML during pregnancy (11,19). Therefore, growth rates may be hormonally influenced and woman may be at higher risk of adverse outcomes if pregnant (2). AMLs have been shown to express estrogen and progesterone receptors (20) which supports this presumption. Gould Rothberg BE et al. (21) described rapid renal angiomyolipoma growth in a woman receiving progesterone contraception. It has been documented that the surgical approach of AML with vascular invasion in pregnancy is a safe and effective means of treatment which avoids the life-threatening complications to the patient without harm to the fetus (2).

In some rare cases, the tumor thrombus can reach the inferior vena cava. Lopater et al. (22) reported a case of a pregnant woman in the 30th week of gestation, with renal angiomyolipoma of 3cm of size with cava thrombus which was treated by surgical thrombectomy alone during pregnancy. The partial nephrectomy was performed 3 months after delivery. In this case, the benefit-risk ratio, regarding the risk of intraoperative bleeding as well as possible complications owing to fetal immaturity if the caesarean delivery

had to be done before thrombectomy, was in favour of primary thrombectomy.

In order to prevent thrombus extension and emboli, before or during the surgery, a vena cava filter can be applied. This cannot be done if the thrombus is located just below the suprahepatic veins (23).

Other surgical approaches for AMLs are represented by nephron sparing surgery, selective embolization, radiofrequency or cryoablation. The nephron-sparing surgery is contraindicated in case of a non-functioning kidney with uncontrolled hypertension, local tissue invasion, vascular invasion or potential malignancy (19). In those cases, the total nephrectomy has to be performed.

The angioembolization showed 31.6% of recurrence rate, recurrence defined as increase in size >2cm, which is probably due to adipose tissue resistance to embolization (24).

Tan YK et al. (25) reviewed a 10-year experience of radiofrequency ablation of incidental benign small renal masses, as an alternative to partial nephrectomy, which revealed to be an efficient treatment.

Percutaneous or laparoscopic renal tumour cryoablation could also be an efficient alternative to open surgical approach with minimal morbidity, encouraging treatment results and shorter hospital stay, although long-term follow-up is required (26,27).

Conclusion

Although considered as a purely benign lesion, angiomyolipoma and especially its epitheloid subtype, may be the cause of potentially fatal clinical outcomes. Its malignant behaviour, seen as a rapid growth, is especially observed in pregnancy because of its expression of estrogen and progesterone receptors. The rare cases of angiomyolipoma with vascular invasion are always to be treated surgically also when asymptomatic. In some cases, the thrombectomy can be carried out primarily, followed by nephron-sparing surgery or total nephrectomy.

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ANGIOMIOLIPOM BUBREGA SA TROMBOZOM BUBREŽNE VENE

Goran Arandjelović, Claudio Milani

Urološka klinika, Bolnica Sv. Đovanija I Pavla, Venecija, Italija

Kod 31-godišnje bolesnice, nakon 40. nedelje trudnoće, okončane carskim rezom, uspostavljena je dijagnoza male desne bubrežne mase sa trombom u bubrežnoj veni. Nakon radikalne nefrektomije i trombektomije, histopatološki nalaz pokazao je angiomiolipom. Predstavljamo redak slučaj malog angiomiolipoma sa invazijom bubrežne vene. *Acta Medica Medianae 2015; 54(1):48-52.*

Ključne reči: *Angiomiolipom, bubreg, tromboza bubrežne vene, radikalna nefrektomija*