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Renal Angiomyolipoma in a Pregnant Patient at the Youngest Age: A Case Report

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ABSTRACT

Angiomyolipomas are the most common benign tumor of the kidney composed of smooth muscle, vascular and adipose tissue. We presented a case of 19 years young primigravidae at 32 weeks with complain of flank pain and hematuria. On examination, she was having right renal mass with uterus of 32 weeks and normal fetal heart sound. Her USG showed single live intrauterine fetus of 31 weeks 4 days with reduced liquor and a large 12.2 × 8 cm size, mixed echogenic lesion with slight vascularity seen in maternal right kidney distorting the normal renal architecture suggestive of renal neoplasm. The patient underwent caesarean section at 34 weeks after steroid cover. CECT of abdomen, done after delivery, showed a large heterogeneous enhancing solid mass lesion in right kidney, measuring 8.7 × 8.8 × 12.8 cm. The patient underwent right radical nephrectomy and the histopathology showed angiomyolipoma of kidney.

Keywords: Renal mass, Angiomyolipoma

Abbreviations: AML: Angiomyolipoma

INTRODUCTION

Angiomyolipomas are most common benign tumor of the kidney composed of smooth muscle, vascular and adipose tissue which typically present with flank pain and hematuria. Although being benign, they may grow significantly so as to impair kidney function. The blood vessels in it may dilate and burst, leading to retroperitoneal hemorrhage. Presentation of this tumor in pregnancy can be devastating to both mother and baby and require emergent delivery at premature gestational ages.

CASE REPORT

A 19 years young patient, primigravida at 32 weeks presented with chief complain of right flank pain and on and off hematuria from last 10 days in Obstetrics and Gynecology Department. On examination, vitals were stable with pulse 98/min, blood pressure of 102/70 mm Hg and she was having mild pallor. On per abdominal examination uterus was of 30 weeks, cephalic presentation and fetal heart sound was regular. A mass in right flank region of size approx. 11 × 8 cm with firm consistency is felt below right sub costal margin with upper border cannot be reached. Her USG showed single live intrauterine fetus of 31 weeks 4 days with reduced liquor (AFI=6 cm) and a large 12.2 × 8 cm size, mixed echogenic lesion with slight vascularity seen in maternal right kidney distorting the normal renal architecture suggestive of renal neoplasm. Her HB was 8.5

g/dl. Renal function tests were normal. Urine routine examination showed 50-60 RBC/HPF. Although MRI is to be done ideally for further evaluation of renal mass but because of poor resource setting in developing country and cost factor it was not done. Decision to terminate the pregnancy after fetal lung maturity was planned after a discussion with urologist and pediatrician. Steroid cover was given for lung maturity of fetus and one unit blood was transfused to mother in view of anemia.

Trial of normal labor was given but the patient land up into caesarean section because of failed induction at 34 weeks and given birth to a healthy female child of 1.8 kg. After 4 days of caesarean, CT scan of whole abdomen was done which showed a large heterogeneous enhancing solid mass lesion in right kidney, measuring 8.7 × 8.8 × 12.8 cm, involving predominantly upper and mid pole with areas of

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hemorrhage and necrosis with surrounding fat planes appears normal (**Figures 1a and 1b**). Left kidney was normal.



Figure 1. CECT of whole abdomen of patient after delivery showing right renal mass shown by arrow. (a) Transverse view. (b) Coronal view.

Patient underwent open radical nephrectomy of right kidney after 6 days of caesarean. The gross feature of specimen showed kidney measuring $13.5 \times 9 \times 7.5$ cm and sectioned kidney showed a well circumscribed mass measuring 12×8 cm, occupying one pole and middle portion of the kidney in cortex and medulla. Cut surface of mass is solid grayish white with areas of necrosis and hemorrhage. Microscopic sections reveal a neoplasm showing islands of mature adipose tissue, large thick walled blood vessels and spindle cells with blunt nuclei circling these vessels. The histopathological features were consistent with angiomyolipoma. The patient was discharged asymptotically with a healthy live baby (**Figure 2**).

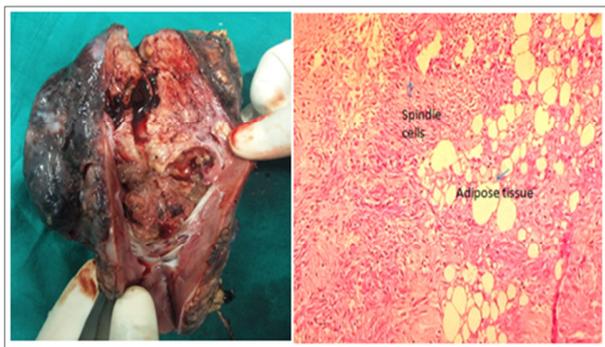


Figure 2. (a) Gross specimen of renal tumor with well circumscribed mass with areas of necrosis and hemorrhage after radical nephrectomy; (b) Histopathology (H&E, x40) of renal mass showing angiomyolipoma.

DISCUSSION

Angiomyolipomas are the most common benign tumour of the kidney and these are most commonly found in middle-aged women [1]. Histopathologically, it is composed of composed of variable proportions of adipose tissue, spindle

and epithelioid smooth muscle cells and abnormal thick walled blood vessels. The two organs most commonly involved organs are the kidney (77%) and liver (14%). [2] They are found either sporadically (80-90%) or in patients with tuberous sclerosis. The prevalence of AML in the general population is around 0.13% [3] and is more common in females. AMLs have a greater tendency to increase in size and rupture during pregnancy and is thought to be because of expression of estrogen and progesterone receptors in AMLs [4]. It may present as flank pain, hematuria or Wunderlich syndrome which is spontaneous, non-traumatic renal hemorrhage confined to the sub capsular and peri-renal space. On extensive search of literature using keywords renal angiomyolioma, renal hamartomas, kidney tumors, kidney angiomyolipoma, AML, rupture, hemorrhage of renal angiomyolipoma in pregnancy till date and on review of literature (from year 1978 till now) total 29 cases have been reported, the age found to be varying from 21 to 45 years with mean age 32.4 years [5,6,8]. 24 cases have hemorrhage in pregnancy [5,6,8]. We have reported the case of angiomyolipoma with pregnancy at the youngest age that is at 19 years. The age at first pregnancy is still at a lower side in developing country. For asymptomatic patients, a conservative approach may be chosen, especially during pregnancy [5]. For these patients, definitive treatment may be postponed until the postpartum period [7]. In pregnant patients; close follow-up may be preferred because of the high risk of rupture. Vaginal delivery can be considered as a safe approach for these patients. Although most patients with angiomyolipomas in the literature delivered their babies via caesarean section and only a few were delivered vaginally [5,6], caesarean section to be done for obstetric indications as it does not reduce the risk of rupture. Vacuum extraction can also be an alternative option for these patients in order to shorten the second stage of the labor. The gestational age at presentation vary from 9 weeks to 38 weeks [5,6]. Therapeutic termination was advised by Ferianec et al. [8] in a case presented at 9 weeks (first trimester). Our case reported at 32 weeks and delivered by caesarean at 34 weeks. The size of renal tumor varies from 4 cm to 21 cm with the largest tumor reported by Ferianec et al. [8] of 21 cm. In our case, the size of renal tumor was $13.5 \times 9 \times 7.5$ cm after right radical nephrectomy. Management during pregnancy is still a dilemma due to few cases. The various management options include embolization, nephrectomy and conservative approach [6]. In cases of rupture, in hemodynamically unstable patients, emergency surgery or arterial embolization (if available) are the main options of treatment.

CONCLUSION

Renal masses presenting during pregnancy should be evaluated and managed properly. Angiomyolipoma should be kept in mind as they can grow and be symptomatic for the first time during pregnancy. Careful follow up is needed as they can rupture during pregnancy.

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