Case Report

Renal angiomyolipoma in horseshoe kidney – A surprise diagnosis at laparotomy

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Introduction

The presence of renal tumor with pregnancy is rare (.07-.3%)¹.

Angiomyolipoma (AML) is a benign renal neoplasm derived from perivascular epitheloid cells and composed of fat, and vascular and smooth muscle element besides HMB-5 reactive spindle cells^{2,3}.

The horseshoe kidney is the most common type of renal fusion anomaly and it has increased incidence of renal tumors like renal cell carcinoma, Wilm's tumor and carcinoid tumors.

Here we present the rarest case of renal angiomyolipoma in horseshoe kidney with pregnancy. In worldwide literature, many cases of AML associated with pregnancy are reported, but no case is reported of AML in horseshoe kidney with pregnancy.

Case report

A 29-year-old G₃P₁+1 woman came for routine antenatal checkup in third trimester. She had no complaints. Her general and obstetric examination showed normality but there was a huge solid fixed lump in the right side of the abdomen. She was advised ultrasonography (USG) for the assessment of fetal well being as well as to evaluate the lump. USG showed a single live pregnancy of 35±1 weeks with asymmetrical fetal growth and more than mild hydramnios. It also revealed a large, ill defined, echogenic mass with small hypoechoic areas in the maternal right iliac and lumbar regions. The appearance was in favor of ovarian dermoid. She was kept under observation. An elective Cesarean section was done after 39 weeks of pregnancy and a male baby of 2.5kg, with apgar score 8 at 1 minutes and 10 at 5 minutes was delivered. After the closure of uterus a soft solid irregular mass was found on the right side of the abdomen. It extended upto the right subcostal margin from where it could be traced posteriorly. It originated from the right horn of the horseshoe kidney.

The tumor was resected with some renal tissue adherent to it. Proper hemostasis achieved and the abdomen closed. Postoperative period was uneventful and she was discharged on the 8th day. Histopathology reported angiomyolipoma with epitheloid reaction. Follow-up was done after 1 month.

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Figure 1. Tumor arising from horseshoe kidney.

Discussion

AML is a benign neoplasm composed of vascular and smooth muscle element. It has an incidence of about 13/10000¹. Two types are described (a) Isolated AML (b) AML associated with tuberous sclerosis². Characteristic absence of elastic tissue in tumor vessels predisposes the patient to aneurysm formation and spontaneous hemorrhage which is the most dangerous complication¹.

Isolated AML occurs sporadically, is often solitary and accounts for 80% of the tumors. Larger ones are more common in women^{1,2}. AML which are associated with tuberous sclerosis are bilateral, multiple and larger².

Horseshoe kidney is the most common type of renal fusion anomaly. It is more prone to recurrent infections. Certain tumors are more frequent in horseshoe kidney i.e. renal cell carcinoma, Wilm's tumor and carcinoid tumors.

Clinical presentation during pregnancy is similar to that in nonpregnant women. Majority of AML are asymptomatic². Main symptom is intratumoral and retroperitoneal hemorrhage in 87% of the patients and hematuria in 42% of the patients². There is only a palpable mass in 47% of the patients.

Abdominal pain hypotension or shock are the other symptoms. Angiomyolipoma grows rapidly during pregnancy^{1,2}. There are increased chances of

hemorrhage in pregnancy due to increased vascularity. Rupture does not occur solely in the first pregnancy but occurs especially during the first, second and third pregnancies. Average gestational age at occurrence of hemorrhage is 27 weeks with a minimum of 10 weeks.

Besides, routine diagnostic procedures and USG angiography is useful to plan the surgical approach which in principle should be organ sparing. MRI is done in complicated cases, although CT is the first method of choice for diagnosis².

Basic therapeutic algorithm of renal angiomyolipoma is follow-up, tumor size and the presence of symptoms are most decisive to decide best therapy alternative. Bleeding angiomyolipomas in pregnancy are rare³, but may be devastating and potentially fatal. Most of them are managed with urgent nephrectomy with cesarean simultaneously in patients of 28 weeks or more gestational age³. Nephrectomy is recommended in the first and third trimester. Polar nephrectomy and embolization are some less radical treatment options. Conservative management and vaginal delivery is one mode of treatment, which is followed very rarely and need strict monitoring of maternal and fetal status⁴. These women should have a medical follow-up period of closer intervals during their pregnancy and the postpartum phase.

Young patient with pulmonary lymphangioleiomyomatosis should be cautioned about becoming pregnant.

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