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Renal angiomyolipoma during pregnancy a rare: Case report

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Abstract

The incidence of renal angiomyolipoma (RA) is 0.3% in the general population and even more frequent during pregnancy. Pregnancy can increase the risk of rupture although the casual mechanism is still not clearly defined. We report our case Mrs. M 24 years, ICSI pregnancy quadtriplet reduced to twin delivered by emergency LSCS at 26 weeks of gestation due to abruption and underwent renal artery embolisation after 2 months of LSCS and underwent definitive treatment (Nephrectomy) after one year. We need further enhance the knowledge about the management of these rare tumour during pregnancy.

Keywords: Renal angiomyolipoma, abruption, renal artery embolisation, pregnancy, emergency c-section, nephrectomy

Introduction

Renal angiomyolipoma, one of the rare tumours which is sporadic, but rarest to see during pregnancy. RA has an incidence of 0.3% generally but very uncommon to see in pregnancy. Generally these are hormone sensitive hence flare up during pregnancy and have serious complications of rupture. Origin of these tumours is from epithelioid cells that surround the blood vessels or sometimes found either with tuberous sclerosis or pulmonary lymphangiomyomatosis. Even though it may have symptoms such as flank pain, hematuria, it is commonly detected during routine imaging in pregnancy. These tumours are sensitive to hormones like estrogen and progesterone and hence increase in size during pregnancy very few cases reported in literature hence optimal treatment strategies is still controversial, and can be individualized.

Case detail

Mrs. M, 24 Years female married for 5 years, was taking infertility treatment, tried with 2 IUI cycles failed then proceeded with ICSI. She became positive with quatriplets and was on regular follow up. At 11 weeks gestation, she underwent Twin Fetal reduction. Elective cervix encirclage done at 14 weeks gestation. She had no other co-morbidities and was on regular follow up. At 22 weeks gestation she came with mild abdomen pain and was found to have cervical incompetence and was advised admission and on strict bed rest and symptomatic management. At 24 weeks she developed breathing difficulty and abdomen discomfort and hence shifted to ICU on evaluation. She is found to have angiomyolipoma of (Right) kidney of size 10.9 x 6.3 cm in routine ultrasound. MRI abdomen taken results showed a lobulated heterogeneously contrasted 11x11 cm soft tissue mass lesion arising from the upper segment of the Right kidney. No other hall mark signs were found to suggest tuberous sclerosis. Multidisciplinary approach done with obstetrician, urologist, nephrologist, intensivist and vascular surgeon. Renal function test was within normal range and all other biochemical parameters normal. As the patient's condition had stabilized and given the substantial risk of prematurity before 26 weeks and risk of rupture of tumour, patient was planned for conservative management with close monitoring.

Two weeks after close monitoring, patient developed sudden severe abdomen pain with severe bleeding per vaginum with mild hemodynamic instability. USG done that revealed a retro placental clot of 6 x 6 cm, features Suggestive of abruption placenta. Patient immediately taken up for emergency LSCS with 2 units blood and urologist standby and delivered alive 1st Male,

800 grams and 2nd twin a female 850 grams. Retro placental clots was 300 grams. Since the tumours is hormone sensitive and in this case it was un ruptured, termination of pregnancy may regress the size of the tumours, patient planned for conservative management with informed consent. Postnatally patient was under regular follow up with urologist and renal artery embolisation done 2 months after LSCS.

Tumours size decreased from 11 x 11 to 5.2 x 3.0 cm. She was under regular follow up with urologist with serial USG. One year after Renal artery embolisation, the tumour size again increased to 11 x 7 cm and in view of rupture, she underwent (Right) nephrectomy. She is doing well now with Twin babies. Pathological evaluation of specimen showed epithelioid angiomyolipoma with intact tumour free borders around mass.

Discussion

25% RA have progesterone and estrogen receptors, hence these tumours grow with pregnancy or any hormonal drug usage. Since these tumours arise from epithelioid cells around blood vessels, they can also be seen in other organs such as liver, spleen and sometimes uterus. Usually these tumours are found as incidental findings during routine ultrasound in pregnancy, hence they are mostly asymptomatic. Rarely some patients present with features of sudden abdomen flank pain or even some times breathlessness or acute abdomen and also with features of hypovolemic shock, which indicates rupture. If rupture occurs and patients presenting with hemodynamic instability, stabilization of the patient and emergency laprotomy is the first choice of treatment. As most of these patients are asymptomatic, conservative management is adopted and definitive management can be done after post-partum period. But close follow up of these pregnant patients is needed as there is risk of rupture.



Fig 1: MRI of Renal tumour coronal section



Fig 2: Frozen section of nephrectomy specimen showing tumour in

upper pole of kidney



Fig 3: Nephrectomy Specimen

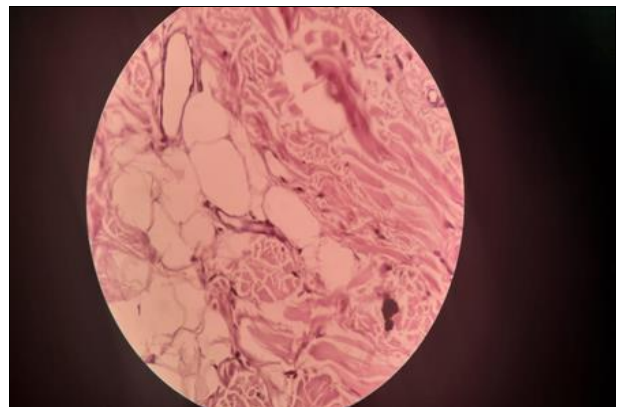


Fig 4: histology showing features of Renal angiomyolipoma

Most of these patients with Renal Angiomyolipoma is delivered by Cesarean section as in our case and only few patients are reported to be delivered vaginally in literature. Vaginal delivery is considered safe for these patients, as Cesarean section does not reduce the risk of rupture. However delivery can be vaginal or Cesarean based on obstetric indication. But in vaginal delivery it is better to cut short the second stage of labour by vacuum extraction or forceps application.

Differential diagnosis for this tumours includes Renal cell carcinoma, oncocytoma or any metastatic lesion from primary tumours. Radiography alone is enough to distinguish all of these tumours but however in case of difficulty, biopsy can be done based on clinical implications. Definitive treatment for RA can be partial or total nepectomy (as in our case) Cryoablation, radiofrequency ablation or even sometimes arterial embolisation for shrinkage of the tumour can be done (as we tried in our patient).

Conclusion

Renal Angiomyolipomas are rare hormone sensitive tumours, that tends to grow during pregnancy and becomes either symptomatic or asymptomatic that decides the mode of treatment. Mostly USG and MRI is enough for diagnosis but rarely biopsy needed for definitive diagnosis. Since very few cases are reported, treatment can be individualized based on patients condition at the time of diagnosis for better maternal and neonatal outcomes. As in our case with conservative management and later nephrectomy after one year will bring additional scope & multidisciplinary approach in mode of

treatment for these patients with good maternal and neonatal Outcomes.

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