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CASE REPORT

Pregnancy with Adrenal Cortical Carcinoma

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ABSTRACT

Adrenal cortical carcinoma (ACC) during pregnancy is one of the rarest diagnoses and requires a high index of suspicion as symptoms and signs overlap with that of normal pregnancy. Adrenal cortical carcinoma is associated with various fatal maternal and fetal complications. Thus, a multidisciplinary approach with timely management is the key component to prevent grave consequences to mother and the fetus. Here, we present a case of primigravida female with Cushing syndrome diagnosed as ACC.

Keywords: Adrenal cortical carcinoma, Cushing syndrome, Multidisciplinary approach, Pregnancy.

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INTRODUCTION

Adrenal cortical carcinoma (ACC) is an extremely rare condition with an incidence of 1 to 2 cases per million per year in adults.[1,2] Most of these tumors are functional and associated with Cushing syndrome and virilization. However, pregnancy may account for many of the symptoms associated with Cushing syndrome, such as weight gain, edema, obesity, abdominal striae, glucose intolerance making the diagnosis difficult.[3] Complications associated with ACC include: hypertension, pre-eclampsia, gestational diabetes, abortion and preterm delivery. Magnetic resonance imaging (MRI) is the best diagnostic modality during pregnancy and complete surgical excision of the tumor is the most effective treatment.

Here, we report a case of pregnancy with Cushing syndrome secondary to adrenal cortical carcinoma and associated with diabetes and pre-eclampsia.

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CASE REPORT

A 26-year-old primigravida female with 28 weeks pregnancy was referred from a private clinic for diabetes and pre-eclampsia. She presented with headache and left flank pain on and off for 15 days. On admission, she was conscious, cooperative and well-oriented to time, place and person. Her blood pressure was 160/100 mm Hg, pulse 102/min and respiratory rate (RR) 20/min. She had a body mass index (BMI) of 27, bloated fascies with acne, hirsutism, central obesity, generalized edema and marked purple striae on lower abdomen and thighs. Cardiovascular system (CVS) and respiratory system were unremarkable. Fetal growth was normal as per the gestational age. Ultrasound examination of abdomen revealed a healthy live intrauterine pregnancy of 28 weeks with a left adrenal mass of 9.5 × 9 cm size. There was no ascites.

Her investigations were: Hemoglobin—10.2 gm%, total leukocite count—11000/mm^3, platelets—2.5 lakh/mm^3, red blood cells—197 mg/dl, serum creatinine—0.4 mg/dl, urine protein 150 mg/dl, S Na^+—136 m Eq/l, SK^+—1.7 mEq/l, serum glutamic-pyruvic transaminase—42 IU/ml.

Morning serum cortisol (8.00 AM) was >63 µg/dl (6.2–19), testosterone—402 ng/dl (14–76), adrenocerticotropic hormone—17.4 pg/ml (3.6–60) and 24 hours urine vanillylmandelic acid—4 mg/24 hours (2–13). Thyroid-stimulating hormone was normal 0.35 µIU/ml.

A multidisciplinary team consisting of a gynecologist, endocrinologist, urosurgeon, anesthesiologist was involved in the treatment of the patient. Blood pressure was controlled with antihypertensives (nifedipine) and diabetes was managed with insulin (regular insulin) therapy. She had persistent hypokalemia for which continuous potassium infusion was given. Patient developed two successive eclamptic fits for which magnesium sulfate was administered by Pritchards regime and continued for 24 hours with monitoring of knee jerk, respiratory rate and urine output. Once stable, her MRI abdomen was done which revealed a 9.5 × 9.4 × 8.5 cm well-defined lobulated solid mass arising from left adrenal gland with preserved fat planes with tail of pancreas and splenic vessels and loss of fat planes with upper pole of left kidney (Figs 1 to 3). Lesion was heterogeneously iso- to hyperintense on T2W1 and heterogeneously hypointense on T1W1. Pregnancy was terminated in view of eclampsia and she delivered vaginally a live female baby weighing

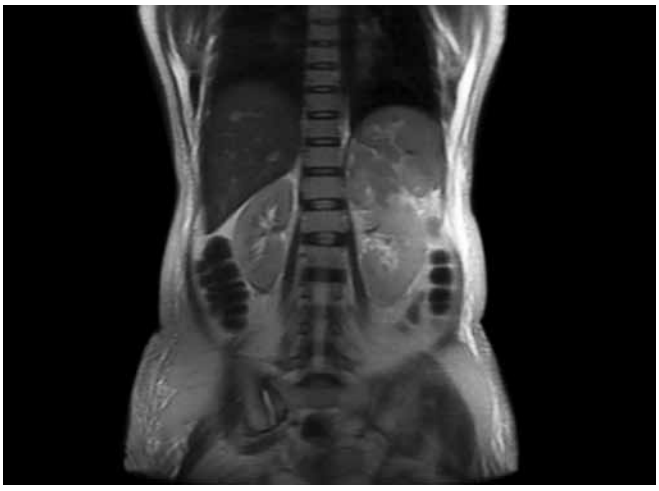

Fig. 1: Coronal section on MRI showing left adrenal tumor

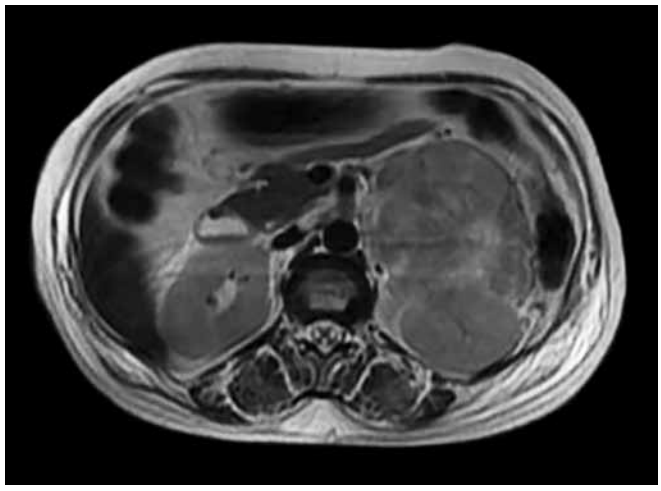

Fig. 2: Cross-section on MRI

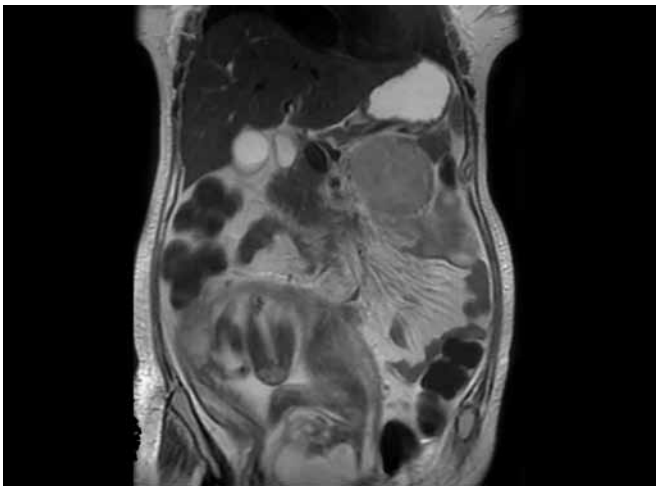

Fig. 3: Coronal section on MRI showing intrauterine pregnancy with adrenal tumor

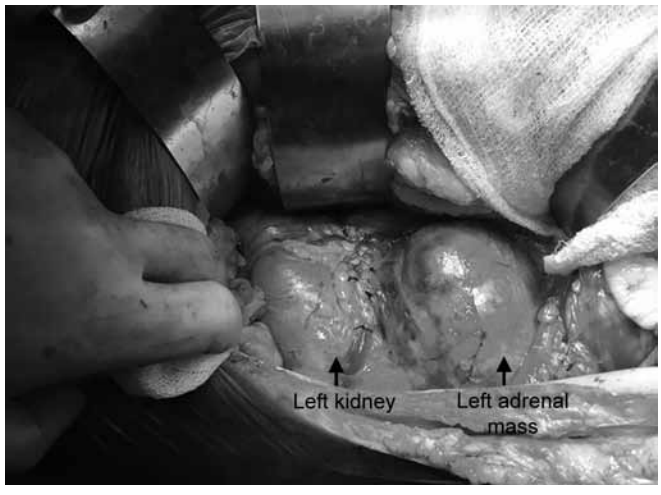

Fig. 4: Intraoperative left adrenal mass with left kidney

1 kg. Baby cried immediately after birth and was shifted to neonatal intensive care unit (NICU). After delivery, her blood pressure remained persistently high for which prazosin 5 mg 12 hourly was added. Hypokalemia was persistent while sodium levels were normal. Two days following delivery, computed tomography thorax showed bilateral minimal pleural effusion with a 7 mm soft-tissue density lesion in supralingular ligament of left lung. Fundoscopy was normal. Two-dimensional echo revealed concentric left ventricular hypertrophy (LVH), normal LV size, systolic function and compliance.

After thorough counseling and obtaining informed consent, laparotomy with left adrenalectomy was performed in collaboration with the urosurgical team (Fig. 4). Postoperatively, her blood sugar and potassium levels were normal and her blood pressure stabilized on nifedipine retard 20 mg 12 hourly. Histopatholgy confirmed the diagnosis of ACC with tumor cells showing pleomorphism and enlarged hyperchromatic nuclei with prominent nucleoli. All surgical margins were free from tumor, venous invasion was absent and tumor necrosis was present. Immunohistochemistry markers were synaptophysin-diffuse positive, vimentin-focal positive; epithelial membrane antigen (EMA), chromogranin–negative. Patient was discharged on antihypertensives. At 1 month follow-up, patient and her baby were doing well.

DISCUSSION

Adrenal cortical carcinoma during pregnancy is an extremely rare entity associated with diverse clinical manifestations. These tumors have a survival rate of 30% at 5 years.[4] Cushing syndrome and virilization are the commonest clinical features related to excessive hormone secretion by the tumor.[1] Our case also had the features of Cushing's syndrome (obesity, abdominal striae, acne, impaired glucose metabolism and hypertension) at the time of diagnosis.

During pregnancy, free plasma cortisol levels are 2 to 3 times higher than normal but the circadian rhythm is well preserved.[5] This patient had high serum cortisol levels with persistent hypokalemia. Low-dose dexamethasone

suppression test is not appropriate in pregnancy because of false-negative results. In this case, adrenal cortical tumor was suspected based on the clinical features of Cushing syndrome with high cortisol levels and MRI findings.

Pregnancy-associated ACC is associated with an extremely poor prognosis and shorter survival period as compared to nonpregnant females. Tumor stage at initial diagnosis is the best prognostic factor.[6] Cortisol hypersecretion is also associated with a poor prognosis. Adrenal cortical carcinoma during pregnancy is associated with an increased maternal and fetal morbidity and mortality and, thus, a multidisciplinary approach including an endocrinologist, obstetrician, anesthesiologist, neonatologist, surgeon and a nutritionist is required for the best management.[7]

Maternal complications include: glucose intolerance, gestational diabetes, hypertension, pre-eclampsia-eclampsia, pulmonary edema, heart failure, osteoporosis, myopathy, psychiatric alterations and death.[8] Fetal complications are abortion, intrauterine growth restriction, prematurity and perinatal death.[9]

Our patient had eclampsia and required termination of pregnancy in maternal interest. Baby was prematurely delivered but survived due to intensive neonatal care. Jairath and Aulakh reported a case of 4 month pregnancy with ACC who presented with headache, palpitation, hypertension and diabetes. Open adrenalectomy was performed following which spontaneous abortion occurred despite tocolytic therapy.[10]

Magnetic resonance imaging is the recommended diagnostic modality in pregnancy to avoid risk of radiation exposure to the fetus. Magnetic resonance imaging is useful to evaluate the surrounding fat planes and exclude metastases. Second trimester is the best time for surgery and can be performed by laparoscopy or an open technique.[11] Complete resection of the tumor is the only curative treatment for ACC and care should be taken to avoid tumor spillage. Termination of pregnancy may be considered in cases of ACC with pregnancy so that definitive treatment can be instituted. Vaginal delivery is preferred over cesarean section because of impaired wound healing associated with Cushing syndrome.

In this case, pregnancy was terminated in maternal interest and tumor was resected in the postpartum period. Diagnosis is confirmed by histopathology and immunohistochemistry. Timely diagnosis and multidisciplinary approach for management can offer the best possible outcome to the mother and fetus.

CONCLUSION

Adrenal cortical carcinoma during pregnancy is extremely rare which is difficult to diagnose and associated with a poor maternal and fetal prognosis. A multidisciplinary approach is essential to ensure the best possible clinical outcome for both the mother and the fetus.

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