Case Report

Adrenocortical carcinoma in pregnancy: A diagnostic dilemma

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ABSTRACT

Adrenocortical carcinoma is a rare disease. Additionally, in the case of coexisting pregnancy, there are diagnostic difficulties due to associated physiological hormonal changes as well as imaging limitations. Cushing's syndrome and virilization during pregnancy is a rare entity with few cases reported in the literature. Misdiagnosis is common as the syndrome may be easily confused with preeclampsia or gestational diabetes. We present the case of a 31-year-old pregnant woman with rapidly developing symptoms related to hormonally active adrenocortical cancer.

Key words: Adrenocortical carcinoma, cushing's syndrome, pregnancy

INTRODUCTION

Adrenocortical carcinoma (ACC) is a rare tumor, even rarer during pregnancy and has extremely poor prognosis both for mother and fetus. In pregnancy, it is a diagnostic dilemma as signs and symptoms overlap.[1-3] The most common clinical presentation, maternal and fetal complication of ACC is Cushing's syndrome (CS) with virilization,[4] hypertension, and prematurity.[5] However, it is unknown whether tumor progression is different in pregnant women due to the rarity of the disease. Magnetic Resonance Imaging (MRI) is the investigation of choice and the best time for a surgical procedure during pregnancy is the second trimester.[6]

CASE REPORT

A 31-year-old, 4½-month pregnant woman was referred to us with complaints of generalized weakness, persistent headache, palpitations, restlessness, nocturia, and increase in appetite since 2½ months. She had high blood pressure and was started on antihypertensive medication (methyldopa and amlodipine). Preeclampsia was excluded because of absence of proteinuria. Physical examination revealed facial and pedal edema, hirsutism, and purple abdominal striae. Fetal development was normal as per the gestational age.

During hospitalization, her blood pressure was 170/100 mmHg and fasting blood sugar/random blood sugar level were 200/408 mg/dl, respectively. Diabetes was controlled with insulin (regular and neutral protamine Hagedorn insulin). Laboratory studies revealed: Hemoglobin (Hb) -11.4 g%, hematocrit (Hct) -34.9%, red blood cell - 4.37 10^6/mm^3, mean cell volume (MCV): 70.7 fl, white blood cell- 10.0 × 10^3/mm^3, platelet count (PLT): 225 × 10^3/mm^3), coagulation tests: Prothrombin time (PT) -10.60 s (10.8-13.5), International normalized ratio (INR) - 0.83 (0.9-1.2), creatinine -0.4 mg/dL, potassium -3.1 mmol/L, sodium -135 mmol/L, bilirubin -0.39 mg/dL, aspartate aminotransferase (AST) -138 U/L (10-37), alanine aminotransferase (ALT) - 245 U/L (10-41), albumin-2.9 gm/dL (3.5-5.2).

24-hour urine for metanephrines and Vanillyl mandelic acid (VMA) were 269.5 mcg/24 hours (25-312) and 4.20 mg/24 hours (0-13.60), respectively. Morning cortisol level measured was 85.66 mcg/dl (6.20-19.40). Low-dose dexamethasone suppression test revealed cortisol level > 63.44 mcg/dL. Testosterone and Dehydroepiandrosterone sulfate (DHEA-S) levels were 602.87 ng/dL (14-76) and 312.90 µg/dL (74-410), respectively. Thyroid stimulating hormone (TSH) was normal, 0.41 µIU/mL (0.35-5.5).

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Abdominal ultrasound and MRI revealed a large heterogeneous tumor originating in the right adrenal gland measuring 77 × 64 × 64 mm, displacing the liver parenchyma superiorly and right kidney inferiorly with focal loss of fat planes suggestive of neoplastic adrenal tumor [Figure 1]. Neither enlarged lymph nodes in retroperitoneal space nor liver metastases was seen. An echocardiography study showed normal left ventricular size and systolic function with ejection fraction of 54% (50-80) and left bundle branch block with no regional wall abnormalities seen.

Hypokalemia (2.7 mmol/L) needed constant administration of intravenous potassium. Diabetes control was achieved by intensive insulin therapy (regular and NPH). The decision to treat was taken by a multidisciplinary team consisting of urologist, gastro-surgeon, gynecologists, endocrinologists, and anesthesiologists. Risk and benefit of laparoscopy *vs.* open surgery were explained to the patient and open adrenalectomy was performed.

One day after surgery, spontaneous abortion occurred despite continuous perioperative tocolytic infusion. Gradually, her general condition improved and potassium normalized (4.6 mmol/L). Anti-hypertensive drug dosage was reduced and gradually stopped. A histopathological examination of the resected tumor confirmed the diagnosis of the adrenal carcinoma with tumor cell showing prominent nucleoli [Figure 2]. On immunohistochemistry, the cells were strongly positive for expression of vimentin.

At 6 months follow up, she was doing well. Her blood pressure was under control without any anti-hypertensives. Chest X-ray and contrast-enhanced CT abdomen were within normal limits.

DISCUSSION

Adrenocortical cancer during pregnancy is a diagnostic dilemma as signs and symptoms overlap and are associated with various hormone overproduction syndromes most commonly being cortisol (30%), androgens (20%), estrogens (10%), and aldosterone (2%).[1-3] The most common clinical presentation of ACC is CS with virilization.[4]

Maternal complications of CS include hypertension (58 to 68%), diabetes or impaired fasting glucose (25%), preeclampsia (14%), osteoporosis or pathologic fractures (5%), psychiatric disorders (4%), cardiac failure (3%), wound infection (2%), and maternal death (2%). The most common fetal complications are prematurity (43%), still birth (6%), spontaneous abortion/intrauterine death (5%), intrauterine growth retardation (21%), and adrenal hypoplasia (2%).[5]

Free plasma cortisol in pregnant patients is 2 to 3 times higher than non-pregnant women, but its circadian rhythm is preserved.[3] Our patient had a very high cortisol (8 times) and testosterone levels with features of CS and virilization, which greatly facilitated the diagnosis.

Due to low incidence of ACC in pregnancy and high risk involved for fetus as well as mother, it requires a multidisciplinary approach to effectively treat this condition.[7] During pregnancy, MRI is the recommended imaging modality as radiation exposure to fetus is limited. The best time for surgical intervention during pregnancy is the second trimester.[6] However, the choice of surgical approach between open vs. laparoscopic remains debatable. Laparoscopy offers advantage of minimum invasive surgery but at the same time is associated with some increased theoretical risk associated with pneumoperitoneum but nevertheless it is safe in pregnancy.[8] In our view, it should be individualized based on patient and surgeon preference after explaining relative risk involved in each case and surgical expertise.

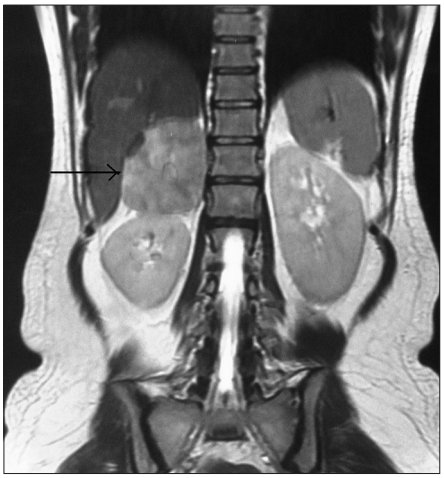

Figure 1: Large heterogeneous tumor originating in the right adrenal gland measuring 77 × 64 × 64 mm

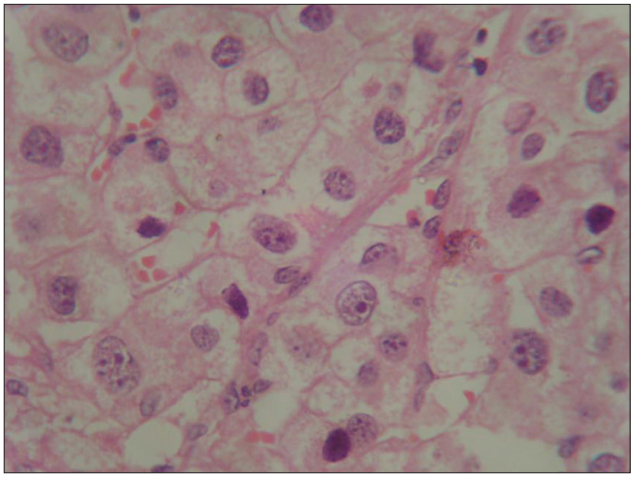

Figure 2: Adrenal carcinoma with tumor cell showing prominent nucleoli

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