Hypertension secondary to pheochromocytoma. We report a series of young pregnant women presenting with intrauterine hypoxia, thus imposing a serious risk to the fetus [6]. Maternal catecholamine exposure may be responsible for the occurrence of placental abruption and have deleterious effects on the uteroplacental circulation and of catecholamines, in patients with pheochromocytoma may catecholamine levels are within normal limits. Even in patients with pre-eclampsia, although it may imitate other clinical conditions during pregnancy. Early diagnosis, timely and appropriate management reduces possible maternal and fetal complications. We report a series of three pregnant women diagnosed of hypertension secondary to pheochromocytoma. Following removal of the tumor, all patients recovered from high blood pressure. Multidisciplinary approach is of utmost importance and essential during the management of this life threatening condition during pregnancy.

INTRODUCTION

Hypertension during pregnancy remains among the most understudied areas despite the advances in medical care and management [1]. Although the most common cause remains pregnancy-specific, preecclampsia, there is paucity of data regarding the other potentially disastrous hypertensive disorders such as pheochromocytoma. Pheochromocytoma occurring during pregnancy is potentially disastrous to the mother and fetus. Its ambiguous presentation is often mistaken for pre-eclampsia, although it may imitate other clinical conditions during pregnancy. Early diagnosis, timely and appropriate management reduces possible maternal and fetal complications. We report a series of three pregnant women diagnosed of hypertension secondary to pheochromocytoma. Following removal of the tumor, all patients recovered from high blood pressure. Multidisciplinary approach is of utmost importance and essential during the management of this life threatening condition during pregnancy.

Case report 1

A 23 year old moderately built primigravida was detected to have hypertension during her regular early third trimester (28 weeks) antenatal check-up. She was put on antihypertensive medications and advised rest till the end of pregnancy. Further assessment of blood pressure during the rest of pregnancy was on the higher side but with no acute episodes. Urine examination revealed no proteinuria. The weight of the patient in the early part of third trimester was 46 kgs and the patient had no pedal edema. The patient went into labor at the end of 40 weeks. The blood pressure shot up during the delivery, however the Obstetricians were able to manage it and the patient delivered (normal vaginal) a healthy 2500 gms female child. Post-operatively the patient continued to have high blood pressure for which she was referred to our center. Routine ultrasonography of abdomen revealed normal sized kidneys on both the sides. There was a 4 cm diameter mass lesion in the region of right suprarenal gland. Twenty-four hour urinary excretion of VMA and cortisol were within normal range. The patient was put on a combination of alfa (Prazocin 10mg/day) and beta blockers (Metoprolol 50mg/day) to control hypertension. A clinical diagnosis of pheochromocytoma was made and the patient underwent laparoscopic right adrenalectomy two weeks after delivery. Histopathological examination of the lesion revealed a well-defined adrenal tumor with adrenal rests separated by fibrovascular stroma. The tumor cells were large, polyhedral and had abundant granular cytoplasm. The features were diagnostic of pheochromocytoma. Post-operatively the patient’s blood pressure was well controlled without the need for further antihypertensive medications.
Case report 2

A 27 year old gravida 2 woman presented at 18 weeks with uncontrolled hypertension and proteinuria. She had been recently diagnosed with gestational diabetes mellitus with no prior medical illnesses. She remained asymptomatic but had episodes of palpitation. Her blood pressure was labile with episodes of tachycardia during admission. A clinical impression of pregnancy with pre-eclampsia was made. However the physicians treating her hypertension thought of evaluating the cause of her labile BP and difficult-to-control hypertension that was associated with tachycardia. A routine abdominal ultrasonography was done to image the kidneys and other organs. A left sided adrenal mass of 5X4 cms was noticed. Urinary and blood metanephrines were elevated. A magnetic resonance imaging confirmed the lesion. The patient was put on antihypertensive medications (alpha and beta adrenergic blockers), blood/fluid transfusions given and advised to proper hydration. The patient was counselled regarding the surgery, complications and outcome of pregnancy. Under general anesthesia and sodium nitroprusside infusion, laparoscopic left adrenalectomy was performed (Figure 1). Post-operative period was uneventful, though the patient had episodes of very high BP (200/120 mm Hg) during surgery. The patient was gradually tapered of her antihypertensives in the post-operative period. The patient continued her pregnancy and delivered normally a healthy male baby at 38 weeks.

Case report 3

A 31 year old primi was diagnosed to have hypertension during her first trimester (12 weeks) antenatal check-up. She was put on antihypertensive medications and advised rest. She had episodes of palpitations, excessive sweating and syncopal attacks. A routine abdominal ultrasonography revealed a right sided 4X3 cms right adrenal mass. MRI confirmed the lesion and the mass showed heterogeneous T2 hyperintensity (Figure 2). Urinary and blood metanephrines were within normal range. In view of the clinical symptoms and MRI findings, it was decided to excise the tumor. The patient was counselled regarding the surgery and outcome. The Patient underwent open right adrenalectomy (decision of open nephrectomy was based on financial reasons) in the 16 week of pregnancy (Figure 3). The intraoperative and postoperative period was smooth. Histopathological examination of the tumor confirmed the diagnosis of pheochromocytoma (Figure 4). Post-operatively the patients BP was easily controlled with low doses of calcium channel blockers. The patient continued with her pregnancy and delivered a healthy baby girl at the end of 42 weeks.

DISCUSSION

The classical presentation of pheochromocytoma [1,7] with paroxysmal hypertension, headaches, sweating, and palpitation may not always be the way it presents during pregnancy. The initial diagnosis of hypertension during pregnancy is frequently attributed to pre-eclampsia rather than pheochromocytoma. The diagnosis of pheochromocytoma in pregnant patients is often missed because these tumors have the ability to produce signs and symptoms that mimic other forms of hypertension, including the new-onset hypertensive syndromes in pregnancy, gestational hypertension, and preeclampsia. In fact, the appearance of hypertension can be insidious, and the patient may even appear asymptomatic until delivery [8]. Preeclampsia is usually the first condition that the clinician considers when hypertension appears de novo or accelerates rapidly. However, preeclampsia is usually accompanied by proteinuria and, often, sudden weight gain and edema, as well as liver and coagulation abnormalities [5]. Obesity and overweight contribute to the risk of both preterm preeclampsia and severe preeclampsia, with several studies revealing that women with high pre-pregnancy body mass index were more likely to develop severe preeclampsia [9]. With pheochromocytoma, excessive urinary protein excretion and liver or coagulation abnormalities occur rarely, and sudden weight gain and edema are unusual.

The difficulty in diagnosing pheochromocytoma in pregnancy is because of the inability of the medical team to include pheochromocytoma as a differential diagnosis. Clinical clues, such as labile hypertension, head-aches, and the failure to respond to conventional treatment, should prompt physicians in the possibility of this condition in pregnant patients [1]. Several
mechanisms can trigger clinically overt pheochromocytoma during pregnancy. These include increases in intra-abdominal pressure, fetal movement, uterine contraction, and process of delivery, an abdominal surgical intervention, and even general anesthesia [1,4,5].

Clinical diagnosis, biochemical tests, and imaging are important to make a diagnosis of pheochromocytoma. Blood and urine metanephrines are highly sensitive and yield a significant negative predictive value. In two of the three patients in our series, the urine and blood metanephrines were within normal ranges. The clonidine suppression test is used when metanephrines are slightly increased and diagnosis is dubious. However, it cannot be used in pregnant individuals due to possible adverse side effects [6]. Computed tomography and scintigraphy with metaiodobenzylguanidine are highly sensitive, but contraindicated for pregnant patients. Therefore, the imaging test of choice is nuclear magnetic resonance for its high sensitivity, the bright T2 images, and for not emitting ionizing radiation [10]. Once pheochromocytoma is diagnosed during pregnancy, the goal of medical management is to treat hypertension, prevent paroxysms, and correct hypovolemia.

The best time to surgically remove the tumor is within the first 24 weeks of gestation or after delivery, the second trimester being the safest moment as the risk of miscarriage during the first trimester is high. Patients diagnosed with pheochromocytoma after 24 weeks of gestation should be treated with drugs as used in preoperative preparation for prolonged periods until the fetus is viable. The tumor may be removed during or after a C-section if performed. Laparoscopic adrenalectomy is rapidly becoming the surgical technique of choice for removal of diseased adrenal glands [11]. The small size of the adrenal gland, the benign nature of most adrenal tumors, and the difficulty of gaining access to the organ by open surgery makes the laparoscopic approach particularly suitable for adrenalectomy. Within a short period, this procedure has gained wide acceptance among laparoscopic surgeons and is considered the procedure of choice for the resection of most adrenal tumors [11-13]. Surgery for pheochromocytoma has specific risks that are not associated with other adrenal tumors such as catecholamine induced cardiomyopathy and pneumoperitoneum induced hypertensive crisis [11].

CONCLUSIONS

A pheochromocytoma in a pregnant patient is one of the most threatening medical conditions for mother, fetus, and physician. Though extraordinarily rare, pheochromocytoma is notorious for its devastating consequences. The signs and symptoms are quite variable and nonspecific, with hypertension being one of the most prominent signs. Confusion with the much more prevalent forms of pregnancy-related hypertension is the main cause of overlooking the diagnosis. MRI is the most suitable investigation for reliable localization. The prognosis of pregnant patients with a pheochromocytoma has considerably improved due to technical progress in terms of further evolvement of surgical and anesthesiological techniques. However early awareness remains the crucial factor.

REFERENCES


Cite this article