

*East African Medical Journal Vol. 93 No. 2 February 2016*

#### PHEOCHROMOCYTOMA IN PREGNANCY: CASE REPORT

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### PHEOCHROMOCYTOMA IN PREGNANCY: CASE REPORT

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#### SUMMARY

**This is a case presentation of a 32 year old woman with pheochromocytoma diagnosed at 27 weeks of gestation, she was managed till term, induced and had assisted vaginal delivery. The pheochromocytoma was surgically re-sected successfully at six weeks postpartum.**

#### CASE REPORT

Pheochromocytoma is a neuroendocrine tumour of the medulla of the adrenal gland. It is a catecholamine producing tumour (1-3). The tumour may also be found in the extra-adrenal paragangliomas (extra-adrenal pheochromocytomas), though less common (1). The prevalence of pheochromocytoma in patients with hypertension is 0.1-0.6 %, but in pregnancy it is even rarer 1:54,000 pregnancies. Most of the tumors are benign, 10% are unilateral and 10% are bilateral (2,3).

Majority of the cases are diagnosed at autopsy, and if undiagnosed in pregnancy can lead to adverse maternal and foetal outcomes, with about 50% mortality and fetal loss (4). This calls for a high index of suspicion. A multidisciplinary team made up of an Obstetrician, Endocrinologist, Anaesthesiologist, Paediatrician and a Surgeon is needed in the management of pheochromocytoma in pregnancy (5).

There has been very few (n=200) diagnosed cases of pheochromocytoma in pregnancy who have had a vaginal delivery reported in the world literature. Cesarean section has been the preferred mode of delivery for most centers.

#### CASE PRESENTATION

We present a case of a 32 year old female patient, para 2 +1 gravida 4, who presented for her first antenatal clinic at fourteen weeks gestation. In this current pregnancy, her antenatal profiles, and vital signs including blood pressure (BP) were all within normal limits during her first antenatal visit. At 27 weeks gestation however, she presented with headache, fainting spells, sweating and a fluctuating blood pressure that ranged from 110/80mmhg to 200/125

mmHg in 15 to 30 minutes intervals. She had no history of hypertension within her family or in her previous pregnancies.

On Physician review, she was admitted to the High Dependency Unit (HDU) for continuous BP monitoring. Full blood count, renal function tests, liver function tests, obstetric ultrasound, and MRI / MRA / MRV of the brain were normal. Vanillyl Mandelic Acid (VMA) test was elevated (4,000umols/24hrs; lab ref range 9-35umols/24hrs) and an abdominal MRI was reported as: 'There is a 42x35x30mm well marginated left supra renal lesion, the left adrenal gland is not identified separate from the lesion. The lesion is high signal on T2W and low signal T1W. There is no nulling of signal on SPAIR indicative that it is lipid poor. There is no blooming or diffusion restriction. The right adrenal gland, liver, gallbladder kidneys spleen and pancreas are normal in appearance. There is no ascites or para aortic lymphadenopathy. The lung bases were normal with an intrauterine foetus noted. Conclusion Lipid poor left adrenal mass suspected to be Pheochromocytoma'.

A diagnosis of hypertension secondary to pheochromocytoma at 28 weeks gestation was made. She was commenced on methyl dopa 500mg three times a day and Nifedipine R 20mg twice a day. The patient remained admitted in HDU for 10 days and was discharged back to the maternity unit after blood pressure stabilisation. Surgical review recommended conservative management as long as the BP stabilizes, for elective resection of the mass at six weeks postpartum. Methyl dopa, nifedipine R phenoxybenzamine, calcium supplements and multivitamins were the compliment of medications given while in the wards. At 35 weeks gestation she was allowed home and reviewed every two weeks, for induction of labor at 38 weeks gestation.

At 38 weeks gestation induction of labor was carried

out using prostaglandin E2 pessaries (prostin), twice six hours apart. During induction of labor, her blood pressure remained stable ranging from 120/70 mmHg to 130/80 mmHg. Successful induction was achieved, artificial rupture of membranes was done at four centimeters of cervical dilatation with clear liquor. Labor was augmented with syntocinon 10 IU in one litre of dextrose in normal saline (DNS). Labor progressed well. Assisted vacuum vaginal delivery was performed at second stage. Outcome was a live female baby birth weight 3520g and placental weight of 980 grams.

During the postpartum period the blood pressure stabilized at 130/83 mmHg, no headache sweating or fainting episode were reported. She continued methyl dopa, nifedipine and phenoxybenzamine. She was counselled not to breast feed because phenoxybenzamine is excreted in breast milk. She was discharged home on the fifth postnatal day in stable condition.

At six weeks postpartum, she was re-admitted for surgical resection of the adrenal mass. Intra-operatively, induction of anesthesia was uneventful. An oblique incision was made over the left 12<sup>th</sup> rib to access the perinephric fat and the upper pole of the kidney. A large left suprarenal tumor within the suprarenal gland was exposed, the tumor was carefully dissected and completely excised with very minimal manipulation. During the surgery the blood pressure was fluctuating and the anti hypertensives were stopped soon after the successful removal of the tumour. The post-operative period was uneventful and she was discharged from hospital on the fourth post-operative day.

Histology was reported as '*Sections show a tumor composed of well-defined nests bound by a delicate fibrovascular stroma. The cells are moderately pleomorphic and have abundant glandular eosinophilic cytoplasm. Some of the cells have prominent nucleoli. No mitotic figures are noted. Histopathological diagnosis of Pheochromocytoma*'

## DISCUSSION

A high index of suspicion is required for timely diagnosis and management of pheochromocytoma in pregnancy. The main clinical presentations are headaches palpitations, sweating, fainting episodes and paroxysmal hypertension which can occur suddenly. Due to its non-specificity, and also despite the high level of awareness of the clinicians it can be easily missed. The case presented had all these symptoms and the suspicion of the condition led to a timely diagnosis and management.

Transient excessive maternal levels of catecholamines, which can lead to extreme vasospasm, are associated with uteroplacental placental insufficiency in these cases and may be responsible for

the occurrence of placental abruption and intrauterine hypoxia (5). In this case, phenoxybenzamine was used for alpha adrenergic blockage to reduce the effect of catecholamines. Pheochromocytoma can be unmasked by pregnancy due to the pressure effect of the uterus, (13) and should be ruled out in cases with hypertensive disease of pregnancy.

Diagnosis of pheochromocytoma in pregnancy poses a challenge and starts with a high index of suspicion. Demonstration of excess secretion of catecholamines and or its metabolites is key in diagnosis. The tests include 24-hour urinary fractionated metanephrines and catecholamines, plasma catecholamines, urinary vanillylmandelic acid, or plasma chromogranin A (3,10). Biochemical confirmation of pheochromocytoma is usually followed by imaging to locate the tumor. In pregnant women, T2-weighted magnetic resonance imaging (MRI) without gadolinium enhancement is the imaging procedure of first choice since, CT scanning involves radiation exposure (3, 11, 12).

Surgical resection of the tumor is the gold standard of management of pheochromocytoma. The surgical removal of the tumour should be done before 24 weeks of gestation if diagnosed early in pregnancy, otherwise the surgery should be performed six weeks after delivery. There are centers however, who do deliver the patients by Caesarian section near term and perform tumor resection in the same sitting. Recent cases of concurrent tumor resection at caesarean sections have had good outcomes (9). Where feasible, laparoscopic surgery is the best surgical approach due to the less complications associated with it (6, 7).

Caesarean section is considered to be the best mode of delivery for these patients due to the risks associated with labor and vaginal delivery. Some studies have reported a higher maternal mortality in vaginal delivery (31%) than caesarean delivery (19%) (4,8). In our patient we opted for the conservative approach and surgery after the puerperium with good outcome.

In conclusion, for pregnant women suspected to have pheochromocytoma, a multidisciplinary team approach is essential for timely diagnosis, institution of the right medical therapy and rational and individualised timing of curative surgery as well as planning of mode of, and timing of delivery for optimal outcomes for mother and baby. In patients in whom the condition has been well controlled there is room for vaginal delivery.

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