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A case report of Pheochromocytoma and neurofibromatosis during pregnancy

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Objective: To describe a patient diagnosed with Pheochromocytoma in the third trimester of pregnancy and discuss the multidisciplinary team management.

Background: Pheochromocytoma is a rare neuroendocrine tumour with a highly variable clinical presentation. Its prevalence in patients with hypertension is only 0.1-0.6% and 0.002% of pregnancies. Most common presentations are episodes of headache, sweating, palpitation, and hypertension. Due to the effects of catecholamine secreted, potentially lethal cardiovascular complication can occur. Approximately 24% of these tumours are inherited, associated with neurofibromatosis type 1 and Von Hippel-Lindau syndrome, or occur in cases of multiple endocrine neoplasia type 2. Pheochromocytoma can cause maternal and fetal deaths in up to 50% of the cases if undiagnosed. In 20% of the cases, diagnosis is not made during pregnancy.

Case: A 34-year-old diagnosed patient with Neurofibromatosis, weighing 35 kg and in her third pregnancy at 35 weeks of gestation, who had two previous uncomplicated pregnancies with normal vaginal deliveries presented with on and off headaches, palpitations, vomiting suggestive of hyper-adrenergic spells for five years. The blood pressure values were normal until the time of surgery. The Ultrasound scan revealed a large left supra renal mass 7.5x8.0 cm in size and the fetal assessment showed evidence of growth restriction. Magnetic resonance imaging demonstrated a well-defined left sided supra renal mass. 24 hour urinary metanephrine excretion was 5.4mg/24hours (normal up to 1 mg/24hours). Short Synacthen test, Parathyroid hormone, Serum phosphate and Calcium levels were evaluated to find association with multiple endocrine neoplasia and all were found to be in normal range. Echocardiogram was normal. A multidisciplinary team including the endocrinologist, surgeon, anaesthetist, neonatologist and obstetric team planned for elective caesarean section and tumour removal surgery at the same time at 37 weeks of gestation. Alpha and beta blockades were started prior to surgery. A healthy baby delivered and open adrenalectomy along with removal of the tumour was done.

Conclusion: The main objectives in managing Pheochromocytoma during the pregnancy are early diagnosis, usage of alpha and beta blockers, and avoidance of hypertensive crisis during delivery and surgery. This case illustrates that with prompt diagnosis, advanced methods of tumour localization and multi-disciplinary team management, Pheochromocytoma in pregnancy can be treated successfully. Thus, it is important that obstetricians are aware of this rare possibility during pregnancy.