

PHEOCHROMOCYTOMA ASSOCIATED WITH PREGNANCY: ANAESTHETIC MANAGEMENT

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INTRODUCTION

Pheochromocytomas are the tumor of neural crest cells of adrenal gland secretes Catecholamines (norepinephrine being greater than epinephrine). It causes hypertension in 0.1% of cases.¹ 80-90 % are solitary tumors localized to single adrenal gland usually right side while 10% are bilateral. They may originate any where in neural crest side from neck to inguinal ligament e.g. right atrium, spleen, broad ligament of ovary, vagina, bifurcation of aorta and in 10% of cases it is inherited as familial autosomal dominant trait. In such cases it is associated with Men 2a, 2b,² Von hippel disease³ and neurofibromatosis.

PHEOCHROMOCYTOMA IN PREGNANCY

It is a rare complication of pregnancy but it is life threatening when it does exist. The exact prevalence in pregnancy is impossible to determine but somewhat more than two hundred cases have been reported.⁴ Association of tumor with pregnancy cause significant risk for both mother and fetus. Risk of maternal death is 40%, abortion is likely in 12%, intra uterine death in 23%, while the overall fetal loss may be up to 50%.⁵

Clinical Presentation: Pheochromocytoma with pregnancy is a protean disease that makes it both difficult and challenging to recognize.⁶

Signs and symptoms basically depend upon the predominant secreting hormone. Norepinephrine secreting pregnant patients mostly present a paroxysmal symptoms of hypertension 80-85%, headache 65-70%, palpitation 35-40%, sweating 30-35%, blurred vision 18-20% and anxiety 15%.⁵ Epinephrine secreting tumors present as episodic syncope, faintness and weakness. Most of these signs and symptoms are also present in normal pregnancy, so the first thing to diagnose this in pregnancy is to have a high index of clinical suspicion.

Other modes of presentation include myocardial infarction,⁷ heart failure, dysrhythmias, cardiomyopathy,⁸ and malignant hyperthermia.⁹

Diagnosis: The two main stay of diagnosis are biochemical testing to find out increase in level of hormones and their metabolites and imaging techniques to find out the localization of tumors.

Biochemical tests are most commonly used to establish the diagnosis of Pheochromocytoma.¹⁰ Measurement of urinary catecholamines and their metabolites metanephrine, normetanephrine, vanillyl mandelic acid and measurement of plasma level of catecholamines are the commonly performing tests. While Ultra sonography, CT scanning, Metaiodobenzylguanidine scanning and MRI are the commonly used imaging techniques.

Treatment: For pregnant patients presenting at or near term, tumor can be treated medically by using alpha-adrenergic blockage. Phenoxybenzamine (PBZ) is usually the initial drug of choice. It is a non-competitive long acting alpha adrenergic receptor blocker. Usually it is given orally but can be given intravenously in case of emergency situations. Beta-blockers have a role in predominating epineph-

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* Received for Publication: May 31, 2005

Accepted: October 5, 2005

rine secreting tumors or in the prevention of secondary arrhythmias after proper alpha blockage. The definite treatment is surgical resection of tumor and in cases of pregnancy it can be resected during first trimester or at the time of caesarian section.⁵ Maternal mortality is decreased with caesarian section as compared to vaginal delivery.⁵

Anaesthetic Considerations: Pheochromocytoma associated with pregnancy is a life threatening condition. Mortality rate decreases when the anesthetist knows the condition preoperatively. In fact 25-50% of hospital deaths with pheochromocytoma occur during induction of anesthesia for other disorders.

Preoperative Evaluation: Along with routine preoperative assessment including history, previous surgeries, drugs history, proper examination one should specially look for:

- * Adequacy of alpha blockage judged by resting BP, orthostatic hypotension and heart rate.
- * Signs of CHF, cardiomyopathy, ventricular ectopics, EKG evidence of ischemia and specially intra vascular volume status.
- * Treatment with PBZ for 10-14 days is usually required to achieve adequate alpha block. However 3-5 days therapy has also been reported.¹¹

Investigations: It includes routine tests like hemoglobin, hematocrit, electrolytes, urea, creatinine, chest Xray and EKG while other tests like echocardiography, stress test, etc. depend upon overall condition of the patient.

Preoperative medications: Patient should continue alpha blocking and other anti hypertensives till the day of surgery.

Aspiration prophylaxis has a major role in term of pregnancy.¹²

The following criteria shows the optimal preoperative condition for such patients

- * No hospital reading of BP higher than 160/90mmHg for 24 hours before surgery.
- * Orthostatic hypotension with readings above 80/45 mmHg should be present.
- * The patient should have no more than one PVC every five minutes

- * The EKG should be free of ST-T abnormalities.

Proper preoperative assessment is essential as it is associated with reduction in maternal as well as fetal out come.¹³

Choices of Anaesthesia: Choices are between sub-arachnoid block alone or in combination with epidural analgesia or general anesthesia along with control mode of ventilation. Regional techniques are useful in blunting stress response to surgeries, anticoagulating effects, problems associated with intubation extubation can be avoided, prolonged pain relief but again it should be kept in mind that it can lead to sympathectomy which can cause dangerous hypotension in such patients. General anesthesia is usually associated with adrenergic responses during intubation and extubation, which may be harmful in such patients.

Monitoring: Saturation pressure of Oxygen, EKG, temperature, Pulse rate are the routine standards of modern anesthesia practice, while invasive blood pressure, CVP, Pulmonary artery catheter (in case of cardiac pathology) are used in case of secondary pathologies.

Maintenance: It is important to ensure:

- * Adequate level of anesthesia and analgesia should be maintained during induction and maintenance.
- * Avoid unnecessary compression of abdomen as in 50% of patients it is associated with increase in BP.
- * Intraoperative use of Sodium nitroprusside 3 microgram per Kg to maximum 800 microgram per min for control of cardiovascular status.
- * Use of Magnesium sulphate 2-2.5 gram per minute for its cardiostable effects.¹⁴
- * Ketamine, suxamethonium and pancuronium should be avoided because of their effects on cardiovascular system.

Post Operative Management: Proper pain management has important role in such patients and the choice lies in narcotic infusions, patient controlled intravenous analgesia and epidurally administered infusions.

Postoperative BP control is very important, as 50% of patients remain hypertensive for 3-4

days and catecholamines level usually returns to normal within 10 days.

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