Pheochromocytoma: catecholamine-producing tumor of the adrenal gland medulla.

Extraadrenal pheochromocytomas = paragangliomas. Tumors in the sympathetic ganglia.

Secrete two hormones: epinephrine (adrenaline) and norepinephrine (noradrenaline) (catecholamines).
What is a Pheochromocytoma?

Signs and symptoms

Very variable!

Spells
Headaches
Palpitations
Sweating
Chest pain
Pallor
Anxiety
Shortness of breath
High blood pressure
Trembling

Chronic problems
High blood pressure
Heart failure
Elevated glucose/diabetes
Weight loss
Visual impairment (retinopathy)
Pheochromocytoma in Pregnancy

- One of the most threatening medical conditions for mother, fetus, and physician!
- Very rare with a frequency of 0.002% of all pregnancies: 1 in 54,000 pregnancies.
- Notorious for its devastating consequences:
  If undiagnosed, maternal and fetal mortality is around 50%.

Risks for Fetus

- During pregnancy, transient excessive maternal levels of catecholamines can have deleterious effects on the circulation of the placenta.
- Extreme constriction of the blood vessels can lead to placental abruption and intrauterine hypoxia (low oxygen), thus imposing a serious risk to the fetus.
Risks for Mother

- Potential risks to the mother include hypertensive crisis, syncope (loss of consciousness), myocardial infarction, arrhythmias, heart failure, stroke and neurological deficits.
- Women with unexplained heart failure before or after delivery need to be evaluated for pheochromocytoma.
- Highest risk: peripartum period due to labor, abdominal palpation, anesthesia, delivery or the use of certain medications (e.g. analgesics, anti-emetics).

Impact of therapy

- Early detection and proper treatment during pregnancy decrease the maternal mortality to <5%, and the fetal mortality to about 15%.
Biochemical diagnosis: plasma or urinary metanephrines (metabolites of catecholamines are the tests of first choice.

Imaging: MRI
(CT and MIBI-scans need to be avoided in pregnancy)

Stepwise evaluation of hypertension in pregnancy

Hypertension

Before week 20
Evaluate for essential hypertension or secondary hypertension
ECG
Kidney function
Renal Doppler
Thyroid function
Cortisol
Metanephrines
Vanillyl mandelic acid
Renin
Aldosterone

After week 20
Evaluate for preeclampsia and gestational hypertension
Urinary proteins
Present
Preeclampsia
Absent
Evaluate for secondary hypertension
If the tumor is diagnosed in the first 24 weeks of gestation, it should be removed by laparoscopic adrenalectomy after 10–14 days of medical preparation with antihypertensive medications (phenoxybenzamine, doxazosin).

Preferred time point: second trimester.
If the tumor is diagnosed in the third trimester, the patient should be managed until the fetus is viable using the same drug regimen as for regular surgical preparation.

Cesarean section with tumor removal in the same session or at a later stage is then preferred. Vaginal delivery is possibly associated with higher mortality.

All patients should be followed up every year for at least 10 years after surgery.

Patients with extra-adrenal or familial pheochromocytomas should be followed up indefinitely.

Genetic screening for hereditary pheochromocytoma in young pregnant women is indicated because of the implications for the children and the family.
Known genetic defects associated with hereditary pheochromocytomas/paragangliomas

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