PHEOCHROMOCYTOMA

By: Belkys Cota, Priscilla Carpio, Melissa Houchin, Leslie N. Beltran.

Pheochromocytoma

Rare neuroendocrine tumor of the medulla of the adrenal glands (originating in the chromaffin cells), or extra-adrenal chromaffin tissue that secretes excessive amounts of catecholamines (epinephrine and norepinephrine) -- hormones that regulate heart rate and blood pressure.

Pheochromocytoma

May occur as a single tumor or as more than one growth.

It usually develops in the center (medulla) of one or both adrenal glands.

Sometimes this kind of tumor occurs outside the adrenal gland.

Factors associated with pheochromocytoma include:

- A family history of pheochromocytoma
- Tumors in other glands of the body
- Other hormonal disorders
- Genetic diseases including:
  - Von Hippel-Lindau disease
  - Multiple endocrine neoplasia, type 2
  - Neurofibromatosis type 1
  - Paraganglioma syndromes

Extra-Adrenal Sites

Within the sympathetic nerve chain along the spinal cord (orange spots)

Overlying the distal aorta (the main artery from the heart) (green spots)

Within the ureter (collecting system from the kidney) (yellow spot)

Within the urinary bladder (blue spot)

Remember, 90% are in the adrenal glands (red spots on the kidneys)

Signs and Symptoms

- Hyperglycemia
- Tachycardia
- Anxiety
- Chest pain
- Palpitations
- Severe headache
- Flushing
- Diaphoresis
- Abdominal pain
- Increased appetite
- Weight loss
Exams and Tests:
- 24 hr Urine Sample
- Plasma levels of Catecholamines
- Glucose test
- Adrenal biopsy
- Abdominal CT scan
- MRI of abdomen
- Ultrasonography

Laboratory analysis will reveal increased urine catecholamines, vanillylmandelic acid (VMA) and metanephrine.

Exams and Tests:
- 24 hr Urine Sample
- Plasma levels of Catecholamines
- Glucose test
- Adrenal biopsy
- Abdominal CT scan
- MRI of abdomen
- Ultrasonography

Laboratory analysis will reveal increased urine catecholamines, vanillylmandelic acid (VMA) and metanephrine.

Treatment

<table>
<thead>
<tr>
<th>Pharmacologic Therapy</th>
<th>Surgical Management</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Decrease BP:</strong></td>
<td></td>
</tr>
<tr>
<td>Alpha-adrenergic blocking agents: -</td>
<td>Adrenalectomy</td>
</tr>
<tr>
<td>eg. Phentolamine (Regitine)</td>
<td></td>
</tr>
<tr>
<td>Smooth muscle relaxants: -</td>
<td>1/5 Corticosteroid replacement:</td>
</tr>
<tr>
<td>eg. Na nitroprusside (Nipride)</td>
<td>Solu-Medrol</td>
</tr>
<tr>
<td><strong>Before and During Surgery:</strong></td>
<td>Oral Corticosteroids:</td>
</tr>
<tr>
<td>Phenoxybenzamine (Dibenzyline)</td>
<td>Prednisone</td>
</tr>
<tr>
<td>Ca Channel Blockers: -</td>
<td></td>
</tr>
<tr>
<td>Nifedipine (Procardia)</td>
<td></td>
</tr>
<tr>
<td>Beta-adrenergic blocking agents: -</td>
<td></td>
</tr>
<tr>
<td>Propranolol (Inderal)</td>
<td></td>
</tr>
<tr>
<td>Cathecholamine synths. inhibitors: -</td>
<td></td>
</tr>
<tr>
<td>Methyrosine</td>
<td></td>
</tr>
</tbody>
</table>

Prognosis

- 1/3 Patients continue to be hypertensive:
  1) Not all tissue removed
  2) Recurrence
  3) Blood vessels damaged by severe & prolonged hypertension
- The tumors come back in less than 10% of these patients.
- Release of the hormones norepinephrine and epinephrine returns to normal after surgery.
- Less than 50% of patients who have cancerous tumors that spread to the bones, liver, or lung are alive after 5 years.

Assessment Findings

- **Hypertension** (may be persistent, fluctuating, intermittent, or paroxysmal)
- Pounding headaches
- Hyperglycemia and glucosuria
- Tachycardia, apprehension, palpitations
- Profuse sweating, cold extremities
- Nausea, vomiting
- Dilated pupils

Blood sugar
- Hypoglycemia (after surgery)
- Hyperglycemia (before and during surgery)

Blood pressure
- Hypertension (before and during surgery)
- Hypotension (after surgery)

Nursing Assessment:

- The “Five Hs”
  - Hypertension
  - Headache
  - Hyperhidrosis
  - Hypermetabolism
  - Hyperglycemia

Presence of these signs has a 93.8% specificity & 90.9% sensitivity for pheochromocytoma.

Nursing Assessment cont…

- Blood sugar
  - Hypoglycemia (after surgery)
  - Hyperglycemia (before and during surgery)

- Blood pressure
  - Hypertension (before and during surgery)
  - Hypotension (after surgery)
Nursing assessment cont…
- Other vital signs
- Hemodynamic parameters
- Fluid and electrolyte status—including intake and urinary output—and urine catecholamine levels.
- Assess the patient for bleeding and infection
- Assess the patient for pain

Nursing Dx:
- Risk for injury related to potential for hypertensive crisis.

Patient Outcomes:
- Identify early signs and symptoms of hypertensive crisis, and seek medical treatment immediately if they develop
- Avoid factor known to precipitate hypertensive crisis
- Remain free for injury

Nursing Dx:
- Anxiety related to potential seriousness and associated complications of pheochromocytoma.

Patient Outcomes:
- Identify and express her/his feelings about the diagnosis
- Perform activities that help lower anxiety
- Demonstrate positive coping method

Nursing Dx:
- Ineffective renal tissue perfusion related to adverse effects of hypertension in renal vascular system.

Patient Outcomes:
- Maintain adequate renal function
- Have normal renal function studies

Planning & Implementation
- Stabilize patient:
  1) Prescribed Bedrest
  2) Pharmacologic Treatment
- Patient teaching:
  Treatment
  Prognosis
- Prepare patient for treatment
- Monitor: ECG changes, arterial pressures, fluid and electrolyte balance, and blood glucose levels.
- Patient teaching: Self-Care, Follow-up visits.

Evaluation:
- BP stable
- Hyperglycemia controlled
- Patient reports pain relief and improves comfort
- Lungs are clear to auscultation
- No signs of resp. problems
- There is no evidence of infection
- Follow-Up
Question #1

90% of all pheochromocytomas are found?

a) Within the sympathetic nerve chain along the spinal cord
b) In the medulla of the adrenal glands
c) Within the urinary bladder
d) All of the above

Question #2

Pheochromocytoma disturbs the secretion of which hormones?

1) TSH
2) FSH
3) Epinephrine & Norepinephrine
4) ACTH

Question #3

Which is the most common symptom related to pheochromocytoma?

1) Pyuresis
2) Nausea & Vomiting
3) Anxiety
4) Hypertension

Question #4

What is the most conclusive test for the diagnosis of pheochromocytoma?

1) CT scan
2) MRI
3) 24 hr Urine Sample
4) Ultrasonography

Question #5

What is the primary treatment for this tumor?

1) Adrenalectomy
2) Chemotherapy
3) Insulin Therapy
4) Radiation