Paragangliomas: Under pressure
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Background

- Paragangliomas are rare (2-8 per million) extra-adrenal neuroendocrine tumors of the autonomic ganglia, with many similarities to pheochromocytomas.
- Catecholamine-secreting tumors can be rare causes of paroxysmal hypertension and hypertensive crises.
- Surgery is the definitive treatment, but pre-operative blood pressure management is essential to prevent significant morbidity and mortality.
- No official guidelines exist, but management of hypertension typically includes alpha-blockers, beta-blockers, calcium channel blockers, metyrosine and fluids.

Characteristics of paragangliomas

<table>
<thead>
<tr>
<th>Parasympathetic</th>
<th>Sympathetic</th>
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<tbody>
<tr>
<td>Cause</td>
<td>Most sporadic, 30% with mutations of SDHB, RET, VHL, NF1</td>
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<td>Location</td>
<td>Sympathetic chain, para-aortic ganglia (esp. abdomen)</td>
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<td>Activity</td>
<td>Majority functional, secrete norepinephrine</td>
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<td>Malignant potential</td>
<td>15-25%</td>
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HPI: A 34 year old female presented initially to an outside hospital via ambulance after bystanders noticed unusual behavior and severe vomiting. The patient later endorsed 20 years of paroxysmal headache, blurred vision, palpitations, dysphonia, nausea and vomiting, all of which had been worsening over the previous month.

The patient presented delirious, hypertensive (159/111), tachyphagic and tachycardic with heart rates in the 150's. Laboratory analysis revealed leukocytosis and lactic acidosis. Broad spectrum antibiotics were initiated for E. coli pyelonephritis and suspected pneumonia. Computed tomography of the abdomen and pelvis with contrast revealed a heterogeneously-enhancing 7.4 x 7.0 x 6.6 cm left retroperitoneal mass abutting the abdominal aorta.

The patient subsequently developed an NSTEMI, cardiogenic and hepatic shock, acute renal failure, and acute respiratory distress syndrome requiring intravenous vasopressors, hemodialysis, intubation and mechanical ventilation.

Post-op Days 2-5

- Phenoxybenzamine dose increased by 10 mg per dose every 1-2 days to a maximum dose of 30mg twice daily.
- Started on oral labetolol with gradual increase in dose to maximum of 400 mg twice daily.
- Start on phenoxybenzamine 10mg PO twice daily

Clinical Course

Day 1:
- Started on phenoxybenzamine 10mg PO twice daily
- Started on oral labetolol with gradual increase in dose to maximum of 400 mg twice daily, with intermittent IV labetolol and hydralazine boluses for persistent hypertension (SBP 180/103)

Day 2-5:
- Phenoxybenzamine dose increased by 10 mg per dose every 1-2 days to a maximum dose of 30mg twice daily.
- Patient taken for exploratory laparotomy with successful tumor excision. Blood pressures ranged from 120's/80's to 150/100 with heart rates in the 80's.

Post-op:
- Required intravenous vasopressors for several hours, then developed hypertension which was managed with oral carvedilol and atenolol. Carvedilol was discontinued at outpatient follow-up.

Paragangliomas carry significant risk for peri-operative mortality that is drastically improved with proper blood pressure management:
- 45% operative mortality without pre-operative BP control
- 2.9% operative mortality with careful pre-operative BP control.

Discuss

Paragangliomas can present with hypertension, which may lead to severe morbidity and mortality. Pre-operative blood pressure management is crucial to avoid peri-operative complications. When managing hypertension, it is important to consider the patient's overall health and medical history. Monitoring blood pressure closely and adjusting medication as needed is essential to prevent complications. Effective pre-operative management can significantly reduce the risk of peri-operative mortality.

References