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Pheochromocytoma and Paraganglioma
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Introduction
What is the topic?

Pheochromocytoma and Paraganglioma (PPGL)

Who choose PPGL?

• Possible malignant catecholamine secreting tumor
• Incidence in general population: 0.005-0.0005%
• Incidence in adult hypertensive population: 0.005-0.2%

Underlying Pathophysiology

• Exact cause is unknown (Hills & Marshall, 2016)
• 90% are an isolated Jacksonian phenomenon
• 9% inherited as an autosomal dominant
• Chromaffin cell tumours originate from neural crest cells from the adrenal medulla and extra-adrenal sites (Momotani, Khan, Bharati, Thal, & Anuradha, 2017)
• 80-85% from adrenal medulla
• 10-20% extra-adrenal: organ of Zuckerkandl, neck, thorax, bladder, and prostate

• Most pheochromocytoma secretes norepinephrine and epinephrine at a 85:15 ratio (Nagabush & Elisa, 2018, Hills & Marshall, 2016)
• 15% secret epinephrine predominantly
• Can also secrete dopamine, though less common
• Neoadjuvant chemotherapy to reduce hormone release due to lack of innovation
• “Rule of two,” both adrenal glands involved in 10% of adults with tumor, 10-15% extraadrenal, and at least 10% are malignant

• Malignant spread typically through venous and lymphatic channels to liver (Nagabush & Elisa, 2018)

Pathophysiologic Processes

• Anatomical classification falls short of providing sufficient insight into pathogenesis, clinical presentation, and potential management approaches
• Functional, pathogenesis-based classification may offer better understanding of actual tumorigenesis, expected biochemical profiles, tumor location, and malignant potential
• Functional classification of well-established susceptibility genes
• Phaeochromocytoma group
• Paraganglioma group
• Neuroendocrine group
• Other less commonly involved

• Genetic knowledge helps design new therapeutic approaches that target specific gene pathways
• Example: Hypoxia-inducible factor 2α antagonism (HIF-2α) which promote HIF hydroxilation and degradation of von Hippel-Lindau gene promotes tumor growth at a specific pathogenic point

• Additional clinical presentations and genetic mutations provide unique opportunities to further understand the pathophysiology of pheochromocytoma and paragangliomas (Kanterovich & Pacak, 2019)

What is your question?

Pathophysiology of Pheochromocytoma

• Main signs and symptoms (catecholamine secreting tumors)
• Signs can occur once a month or multiple times per day ranging in intensity in a spectrum
• 24-hour urine collection of metanephrines and catecholamines or measurement of plasma free metanephrines
• Telemetry monitoring for ablation
• Elevation or depression of ECG interval
• Bradycardia or tachycardia
• Hypertensive crisis
• Arrhythmia

• Surgical resection is the primary treatment strategy
• High risk for mortality and morbidity in the periprocedural period (Bai et al., 2018)
• Can be as high as 5% (Bai, Sun, Xue, Zhu, Li, Zhang, Wang, & Wu, 2018)
• Improvements in perioperative medical preparation, anesthesia, and surgical techniques have reduced risk to 3%-5% (Bai et al., 2018)

• Cardiovascular related complications occurred in 24% of patients who underwent removal in a study by Bai et al. 2018 which compared similarly to previous research
• Postoperative hypertension
• Myocardial infarction
• Peripheral vasculature impairment

• Adrenal medulla
• Cerebrovascular disorder
• Cardiac arrhythmia
• Hypertensive crisis

Removal

• Surgical resection is the primary treatment strategy
• High risk for morbidity and mortality in the perioperative period (Bai et al., 2018)
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• Cardiovascular related complications occurred in 24% of patients who underwent removal in a study by Bai et al. 2018 which compared similarly to previous research
• Postoperative hypertension
• Myocardial infarction
• Peripheral vasculature impairment

Implications for Nursing Care

• Close monitoring of BP, HR, pulse oximetry
• Signs can occur once a month or multiple times per day ranging in intensity in a spectrum
• 24-hour urine collection of metanephrines and catecholamines or measurement of plasma free metanephrines
• Telemetry monitoring for ablation
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