Pheochromocytoma and Paraganglioma

Christopher Foltz
foltz1@otterbein.edu

Follow this and additional works at: https://digitalcommons.otterbein.edu/stu_msn

Part of the Nursing Commons

Recommended Citation
Foltz, Christopher, "Pheochromocytoma and Paraganglioma" (2019). Nursing Student Class Projects (Formerly MSN). 342.
https://digitalcommons.otterbein.edu/stu_msn/342

This Project is brought to you for free and open access by the Student Research & Creative Work at Digital Commons @ Otterbein. It has been accepted for inclusion in Nursing Student Class Projects (Formerly MSN) by an authorized administrator of Digital Commons @ Otterbein. For more information, please contact digitalcommons07@otterbein.edu.
Pheochromocytoma and Paraganglioma
Christopher Foltz, BSN, RN, CCRN
Otterbein University, Westerville, Ohio

Introduction
What is the topic?
Pheochromocytoma and Paraganglioma (PPGL)

Pathophysiological Processes
Underlying Pathophysiology
• Exact cause is unknown (Hilens & Marshall, 2018)
• 90% are an isolated finding
• 10% inherited as an autosomal dominant
• Chromaffin cell tumors originating from neural crest cells from the adrenal medulla and extra-adrenal sites (Moniencan, Khan, Bharath, Thosani, & Anuradha, 2018)
• 80-85% from adrenal medulla
• 15-20% extra-adrenal: organ of Zuckerkandl, neck, thorax, bladder, and prostate
• Most pheochromocytomas secrete noradrenaline and epinephrine at a 85-15% ratio (Nagelhout & Elisha, 2018, Hilens & Marshall, 2018)
• 15% secretes epinephrine predominately
• Can also secrete dopamine, though less common
• Neural stimulation can cause hormone release due to lack of inhibition
• “Rule of ten”, both adrenal glands involved in 10% of adults with tumor, 10-15% extramedullary, and at least 10% are malignant
• Malignant spread typically through venous and lymphatic channels to liver (Nagelhout & Elisha, 2018)

Genetic Significance in Pathophysiology
• Anatomic classification fall short of providing sufficient insight into pathogenesis, clinical presentation, and genetic implications, respect to malignant potential
• Functional, pathogenesis-based classification may offer better understanding of actual tumorigenesis, expected biochemical profiles, tumor location, and malignant potential
• Pathogenesis-based classification of established susceptibility genes
  - Pheochromocyte group
  - Neuroendocrine tumor group
  - Syndromic tumors
• Pathogenesis-based knowledge helps design new therapeutic approaches that target specific gene pathways
  - Example: Hypoxia-inducible factor 2α antagonist (IPI-994) which promote HIF hydroxylation and degradation of stabilization that promote tumor growth at a specific pathogenetic point
• Additional clinical presentations and genetic implication may offer opportunities to further understand the pathophysiology of pheochromocytoma and paraganglioma (Kantarcioglu & Pacak, 2019)

Signs & Symptoms
• Main signs and symptoms (catecholamines)
  - Hypertension
  - Palpitations
  - Headache
  - Paresthesia
  - Less common signs and symptoms (Fatigue, Anxiety, Myalgia, Vomiting, Abdominal pain, Fatigue, Headache, Hypertension, and hypertension)

Implications for Nursing Care
• Class monitoring of BP, HR, pulse, urinary output
• Signs can occur once a month or multiple times per day ranging from minutes to hours
• 24-hour urine collection of metanephrine and catecholamines or measurement of plasma free metanephrines
• Telemetry monitoring for abnormalities
• Elevated or depression of ST segment
• Flattening or inversion of T wave
• Prolongation of QT interval
• High or peaked T waves
• Emphasis on BP segment (Hilens & Marshall, 2018)

Genetic Implications
• Alpha blockade before beta blockade
• Blocking vasodilatory 
  - Juxtaglomerular
  - Hypertensive crisis
  - Autonomic crisis
• Proportive goals (Hilens & Marshall, 2018)
  - Tetrofuram, Anyango, Van Sell, & Miller-Anderen, 2009
  - Eroxolinet
  - Normanetin, Below 160/90 mmHg for at least 24 hours before surgery

Removal
• Surgical resection is the primary treatment strategy
• High risk for morbidity and mortality in the perioperative period (Bai et al, 2018)
  - Can be as high as 30%
• Improvements in perioperative medical preparation, anesthesia, and surgical techniques have reduced risk to 2-10%
• 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
• Perivascular vasoconstriction induced by catecholamines probably leads to possible use of vasopressin agonists (noradrenalin) and volume expansion with crystalloid or colloid products
• Prolonged exposure of catecholamines to the myocardium and coronary arteries could presumably lead to collagen deposition and fibrosis in the myocardium

Conclusion
Pheochromocytoma and paraganglioma are rare neuroendocrine tumors associated with catecholamine production
• Surgical resection is the primary treatment
• Genetic research has provided additional information regarding pathogenesis and targeted therapy
• The classic triad of diaphoresis, palpitations, and headache has a reported sensitivity of 89% and specificity of 67% for pheochromocytoma specifically
  - In the presence of hypertension, it raises to 91% and 94%, respectively
• Rarity and variability of these tumors make them difficult to manage and many are discovered incidentally during radiologic examinations or at autopsy
• The key to diagnosing is to first think of it (Bai et al, 2018; Faraggi et al, 2017)

References
Tsegay, E., Anyango, Van Sell, & Miller-Anderen, 2009

What are your symptoms?

Retrieved from https://artsugar.co/products/pheochromocytoma

Retrieved from https://www.medicalnewstoday.com/articles/313203.php