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Primary Cardiac Pheochromocytoma (Paraganglioma)

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1. Introduction

Pheochromocytomas are catecholamine-producing neuroendocrine tumors arise from primitive neural crest cells. About 90% of these tumors occur as solitary benign tumors of the adrenal medulla, where majority of chromaffin cells are concentrated. Only ten percent originates from extra-adrenal sites with the organ of Zukerkandal (paraganglia along abdominal aorta) being the most common. Chromaffin cells can also be found in the wall of blood vessels, along the aorta, prostate, urinary bladder and ovaries. Primary cardiac pheochromocytomas are extremely rare, occurring in only 0.001% to 0.03% of several reported autopsy series. Most of these tumors are found in the left atrium, possibly explained by close proximity of paraganglionic cell nest to the left atrium. Primary cardiac pheochromocytoms produce large amount of catecholamine, primarily norepinephrine and less frequently epinephrine.

2. Pathophysiology and pathology

Cardiac pheochromocytomas occur at any age, but mostly in the fourth and fifth decades of live. They are extremely vascular, their blood supply exclusively derived from coronary circulation. These tumors are usually functional, producing excessive amount of catecholamines, primarily secrete norepinephrine, which is a potent vasoconstrictor and raises the peripheral vascular resistance. Therefore, systolic blood pressure rises, but diastolic blood pressure may fall. It has very minimal, if any, direct effect on the heart, actually, cardiac output may fall reflexly as a result of an increase in the blood pressure. Cardiac pheochromocytoma arise from branchiomeric (coronary, pulmonary or aortopulmonary) paraganglia or visceral autonomic (atrium or interatrial septum) paraganglia. Often, they are dark red-brownish, soft, fleshy and highly vascular non encapsulated tumors that found under the aorta and pulmonary artery in association with left atrium (Figure 1). They can extend into the atrio-ventricular groove and the coronary arteries. Malignant changes are present in 10% of catecholamine-secreting tumors as defined by presence of metastasis or local tissue invasion. Cardiac pheochromocytomas appear more invasive and difficult to “shell out”, unlike benign adrenal pheochromocytomas.



Fig. 1. Resected cardiac pheochromocytoma.

3. Clinical manifestations

The clinical manifestation of cardiac pheochromocytoma can be related to catecholamine secretion, size of the tumor and competition for blood supply with normal myocardium. Headache, excess sweating, flushing and palpitation are the usual symptoms of cardiac pheochromocytomas. Sustained, labile, or paroxysmal hypertension is typically present in almost all patients with cardiac pheochromocytomas as a result of excess circulating catecholamines. These tumors can cause angina chest pain as it competes with normal myocardium for coronary blood supply. Occasionally seizures occur and very rarely intense mesenteric artery vasoconstriction may cause ischemic enterocolitis with severe abdominal pain. However clinical manifestations of cardiac pheochromocytomas are less dramatic compared to adrenal pheochromocytoma. We reported a case presenting with angina chest pain and the tumor received dual blood supply from both right and left coronary arteries and had undergone complete surgical excision utilizing cardiopulmonary bypass (Figure 3, 4).

4. Diagnosis

Clinical suspicion is raised in cases of resistant hypertension particularly in patients with family history of pheochromocytoma. Quantification of the plasma or urinary catecholamine metabolite metanephrine is the most sensitive test for the diagnosis (sensitivity 99%, specificity 81%) in one study that compared biochemical markers in detection of catecholamine-secreting tumors. In addition, contrasted enhanced computed tomography of the abdomen is indicated to rule out adrenal gland involvement. Anatomic localization of cardiac pheochromocytomas can be made with high speed dynamic computed tomography with intravenous contrast bolus administration (Figure 2). Magnetic resonance imaging has been found to be more sensitive in the localization of extra-adrenal pheochromocytoma based on greater resolution and sensitivity for soft tissue. Pheochromocytomas demonstrate a hyper-intense signal on T2-weighted images and typically iso-hypo-intense relative to myocardium on T1-weighted images. Presence of peripheral rim enhancement on late gadolinium enhancement (LGE) indicates vascularity of cardiac pheochromocytomas distinguishing them from a vascular cardiac mass such as cardiac thrombi and lipoma. Total body 123-iodine-iodobenzylguanidine (MIBG) scintigraphy scan is well established for preoperative localization of cardiac pheochromocytoma and has also been found to be of use in locating other neural-endocrine tumors and search for metastatic disease. A recent study, Indium-Octreotide uptake scan has been described in cardiac pheochromocytoma, but its clinical significance is not well established. Coronary angiography is useful in judging local excision of the disease, coronary artery involvement and screening for atherosclerotic disease in these patients with hypertension.

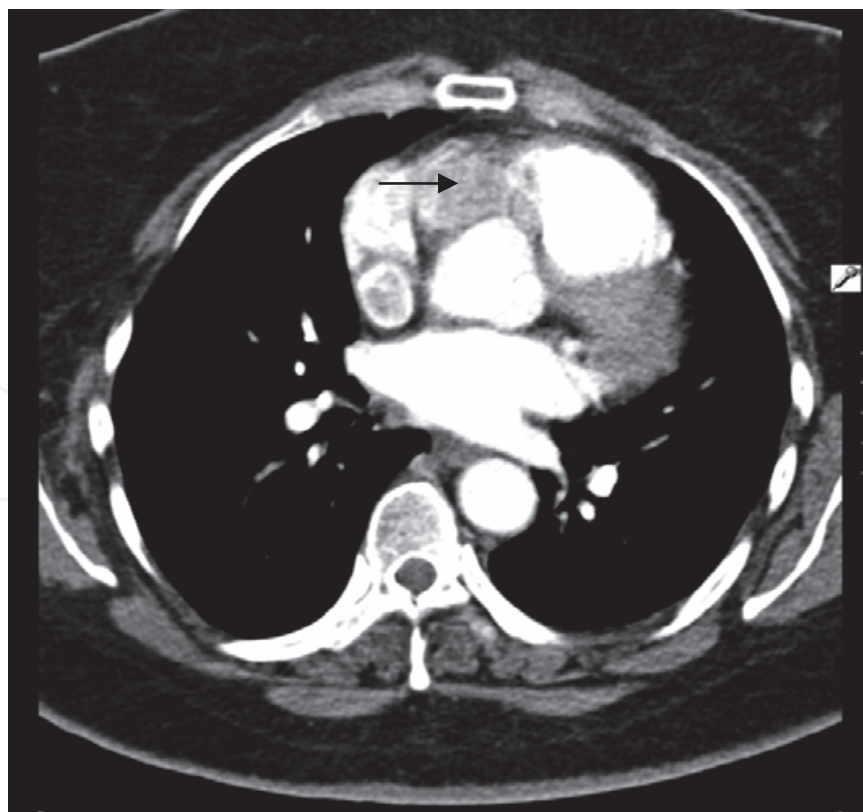


Fig. 2. Computed tomography scan of chest demonstrating a mass overlay the aortic root and extended through right ventricular muscle fibers (arrow).

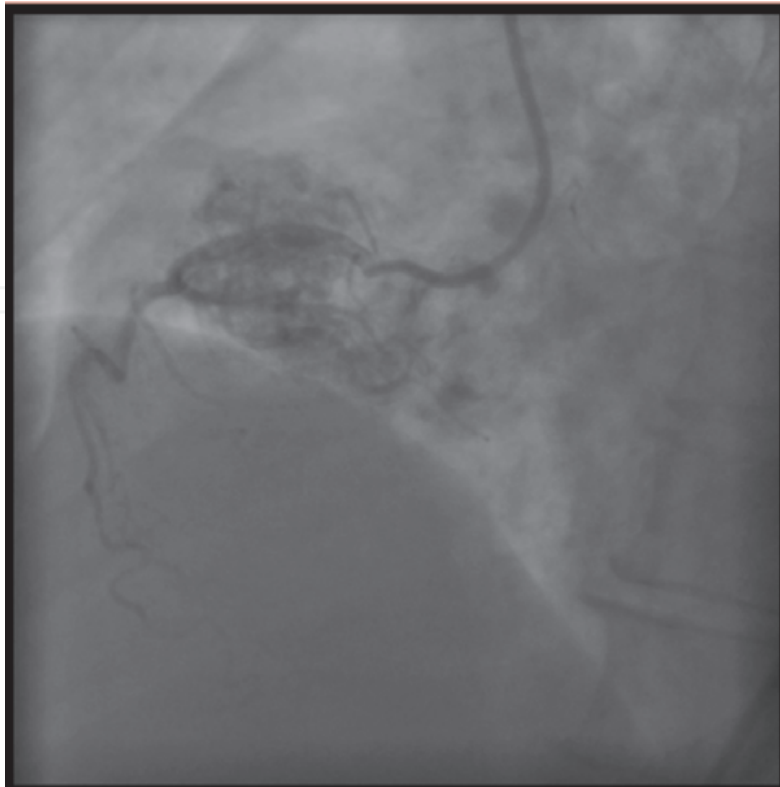


Fig. 3. Coronary angiogram: showing right coronary artery tumor blush.

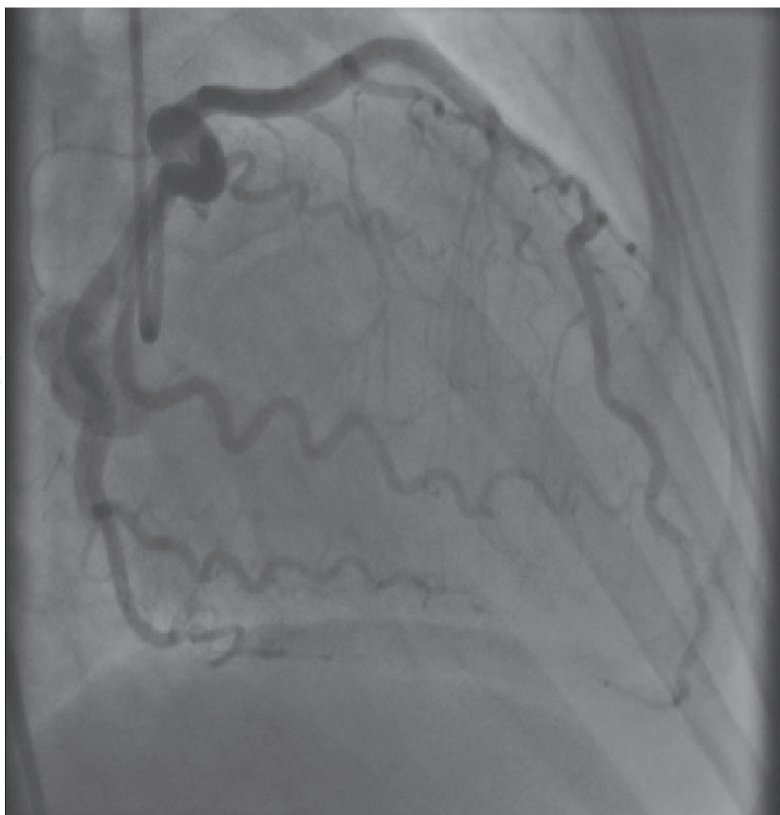


Fig. 4. Coronary angiogram: showing left circumflex coronary arteries tumor blush.

5. Management

Complete Surgical resection with adequate disease-free margins and reconstruction with pericardial or synthetic patch is the gold standard treatment for primary cardiac pheochromocytomas, but this can be technically difficult and is complicated by the position and extension of the tumor. These include pheochromocytoma extension into atrioventricular groove, direct coronary artery involvement and extension into the left ventricle. In these patients, resection with adequate margin carries high mortality and morbidity as a result of fatal hemorrhage and myocardial infarction. In this group of patients cardiac transplantation is the best treatment option provided distant metastasis has been excluded. Adequate preoperative preparation with alpha and beta adrenergic blockers and total cardiopulmonary bypass with cardioplegic arrest should be instituted to isolate the heart from systemic circulation before manipulation of the tumor. However, in all patients, life-long surveillance for recurrence should be performed with regular follow-up employing biochemical testing for fractionated metanephrines and imaging techniques such as 123-iodine-iodobenzylguanidine (MIBG) and computed tomography scans when appropriate.

6. Conclusion

Primary cardiac pheochromocytomas are very rare tumor of the heart. The multimodality imaging studies in assessing cardiac tumor are important in planning surgical strategy. Complete resection of cardiac pheochromocytoma with adequate disease free margin is the standard treatment which safely performed using cardiopulmonary bypass. Long-term surveillance is warranted for tumor recurrence.

7. Acknowledgment

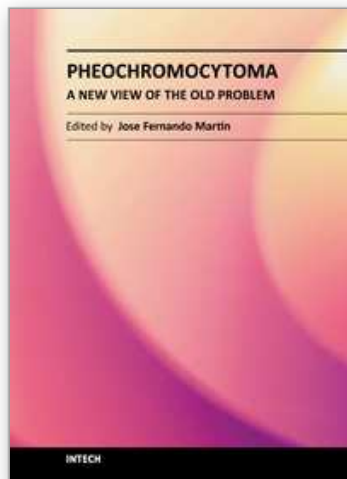
The author expresses his gratitude to his wife Dr. Nadia Batawil for her outstanding support.

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Pheochromocytoma - A New View of the Old Problem

Edited by Dr. Jose Fernando Martin

ISBN 978-953-307-822-9

Hard cover, 164 pages

Publisher InTech

Published online 16, December, 2011

Published in print edition December, 2011

The book is divided into six sections. The first three sections focus on the pathophysiology of the disease, showing anatomo- and histopathological aspects, experimental models and signaling pathways and programmed cell death related to pheochromocytoma. The fourth discusses some specific aspects of clinical presentation, with emphasis on clinical manifestations of headache and heart. The fifth section focuses on clinical diagnosis, laboratory and imaging, including differential diagnosis. Finally, the last section discusses the treatment of pheochromocytoma showing clinical cases, a case about undiagnosed pheochromocytoma complicated with multiple organ failure and other cases about catecholamine-secreting hereditary tumors. Thus, this book shows the disease "pheochromocytoma" in a different perspective from the traditional approach. Enjoy your reading.

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Iskander Al-Githmi (2011). Primary Cardiac Pheochromocytoma (Paraganglioma), Pheochromocytoma - A New View of the Old Problem, Dr. Jose Fernando Martin (Ed.), ISBN: 978-953-307-822-9, InTech, Available from: <http://www.intechopen.com/books/pheochromocytoma-a-new-view-of-the-old-problem/primary-cardiac-pheochromocytoma-paraganglioma->

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