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Management of Right Sided Carcinoid Heart Failure

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Management of Right Sided Carcinoid Heart Failure

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Abstract

Carcinoid heart disease is a rare complication of an already rare disease. This issue is theorized to be secondary to released proinflammatory molecules that deposit on the surfaces of heart valves. These molecules are released from the carcinoid tumor itself and manifest symptoms based on liver, lung and brain function. The detection of 5-HIAA, a breakdown source of serotonin, is useful for diagnosis. These deposits occur most commonly on the endocardium of valvular cusps and cardiac chambers. This case presents a 48 year old woman with echo findings of right heart failure in the setting of a carcinoid tumor. It is important for early recognition and treatment of cardiac lesions to prevent worsening heart failure.

Keywords

Carcinoid, Heart Failure, 5 HIAA

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Conflict of Interest Statement

The authors below certify that they have NO affiliations with or involvement in any organization or entity with any financial interest

CASE REPORT

Management of Right Sided Carcinoid Heart Failure

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Abstract

Carcinoid heart disease is a rare complication of an already rare disease. This issue is theorized to be secondary to released proinflammatory molecules that deposit on the surfaces of heart valves. These molecules are released from the carcinoid tumor itself and manifest symptoms based on liver, lung and brain function. The detection of 5-HIAA, a breakdown source of serotonin, is useful for diagnosis. These deposits occur most commonly on the endocardium of valvular cusps and cardiac chambers. This case presents a 48 year old woman with echo findings of right heart failure in the setting of a carcinoid tumor. It is important for early recognition and treatment of cardiac lesions to prevent worsening heart failure.

Keywords: Carcinoid, Heart failure, 5 HIAA

1. Introduction

Carcinoid heart disease is a rare disease affecting 50% of patients with systemic carcinoid syndrome.¹ The likely mechanism is multifaceted including release of vasoactive chemicals such as serotonin, prostaglandins, histamine, bradykinins, and tachykinins.¹ These substances deposit on the endocardial valve surfaces, causing dysfunction of the valves. Serotonin is normally cleared by the liver, lung, and brain but the overwhelmed system causes deposition into the heart valves.² This is particularly the case in cirrhotic and lung pathology patients. The breakdown of serotonin can be measured to estimate the overall burden.² This case presents a 48-year-old woman with new echo findings compatible with right heart failure secondary to carcinoid syndrome.

2. Case presentation

A 48-year-old woman with a past medical history of hypothyroidism presented with dyspnea for the last month. Denies any productive cough, sick contacts, fevers, or chills. She endorsed diffuse redness

on her face and right arm. Anti-streptolysin A returned positive and therefore completed a 10-day course of cephalexin. Unfortunately, no resolution of symptoms. Other related symptoms included abdominal distress, non-bloody diarrhea and tachycardia. She lost approximately 30 pounds in the past 2 months due to chronic diarrhea. Recent celiac serology testing returned negative. CT Abdomen and Pelvis with contrast showed a prominent and speculated nodal mass at the root of the mesentery with focal nodular thickening of the duodenal wall. Of note, the adrenals were unremarkable, and no other masses were seen.

Lab results

Urinary 5-HIAA returned at 40mg/24hr	(Reference <30 mg/24hr)
Urinary Epinephrine returned 540 mcg/24 hr	(Reference 10 to 200 mcg/24hr)
Urinary Norepinephrine 1070 mcg/24hr	(Reference 80 to 520 mcg/24hr)
Dopamine 162 mcg/24hr	(Reference 0 to 20 mcg/24 hr)

Echo revealed Ejection Fraction 60–65% grade 1 diastolic dysfunction with Moderate Tricuspid Regurgitation. Diagnosed with right heart failure

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secondary to carcinoid syndrome. For blood pressure and volume status control, furosemide was started. Octreotide decreases the systemic surge of serotonin. Her symptoms improved and followed in the outpatient for heart failure. She is scheduled for resection of a carcinoid tumor from the duodenal wall.

4. Discussion

Carcinoid syndrome is a tumor of well-differentiated endocrine and neuro cells that comprise the gastroenterology and pulmonary systems.² There is a mass production of vasodilatory molecules that cause systemic symptoms. These tumor products cause a constellation of symptoms known as carcinoid syndrome.³ Neuroendocrine tumors present in the digestive tract, lungs, and less commonly ovaries and kidneys. Other locations include the rectum, appendix, colon, and stomach.⁴ The differential includes alternative tumors secreting catecholamines. This is ruled out in our case because CT abdomen and pelvis with contrast did not reveal any mass on the adrenals. In addition, the cardiac manifestations would be unusual in pheochromocytoma.⁵

Clinical features⁶

Cutaneous flushing	Appears in late course carcinoid syndrome
Venous telangiectasias	Occurs in about 80% of patients
Secretory diarrhea	Occurs in 10–20% of patients
Bronchospasm	Occurs along with flushing episodes
Wheezing/Dyspnea	Clinical hallmark in about 85% of patients

The valvular lesions in the heart are represented by fibrous plaques deposited on the endocardial surface.⁶ The cardiac chambers and the intima of pulmonary arteries and aorta are the prime targets for inflammatory markers.⁶ The right sided valves are affected first due to the closest area of involvement. The left sided valves are unaffected due to inactivation of vasoactive substances through the pulmonary system.⁷ The other systemic symptoms include pellagra, flushing, and bronchospasm.⁸ The manifestations of these symptoms are based on quantitative amounts of vasoactive substances in the blood stream.⁹

Carcinoid tumors one to two cm in size, full-thickness surgical removal is optimal to prevent leftover of progressive symptoms.¹⁰ Lesions larger

than two cm also has full-thickness removal with addition of regional lymphadenectomy to decrease spread. These measures decrease the chance of carcinoid crisis.¹¹

5. Conclusion

Early treatment and diagnosis of heart lesions will decrease the vasoactivity of these molecules on the heart valves.² The overflow of biologically active compounds from the tumor increases the risk for blood pressure fluctuations and worsening heart failure symptoms.

Conflict of interest

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