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Neuroendocrinal Tumor-Inducing Severe Aortic Regurgitation With Low-Ejection Fraction Stable Heart Failure and Prosthetic Valve Management

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Case Report

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Abstract

Rationale: *Neuroendocrine tumors are rare distinct neoplasms with varying incidence. Neuroendocrine tumors are mostly affecting the gastrointestinal tract. Carcinoid tumors are considered a synonymous name for neuroendocrine tumors. Rheumatic heart disease is a major differential for carcinoid disease. Patient concerns: A young-aged housewife female patient presented to physician outpatient clinic for cardiovascular follow-up for abdominal tumors with aortic valve replacement. Diagnosis: Neuroendocrinal tumor-inducing severe aortic regurgitation with low-ejection fraction stable heart failure. Interventions: Echocardiography, aortic valve replacement, and electrocardiography. Outcomes: Diverse outcome post-aortic valve replacement had happened. Lessons: Valvular heart disease may be occurring with carcinoid tumors but the wrong diagnosis for the rheumatic heart is common. The fixation of low myocardial ejection fraction with absent heart failure is an innovative interesting issue. The absence of the above symptoms and symptoms of heart failure after aortic valve replacement despite low ejection fraction and the still present tumor may carry diverse outcomes.*

Keywords: Neuroendocrinal tumor, Carcinoid, Aortic regurgitation with low-ejection fraction, Stable heart failure, Valvular heart disease.

Abbreviations

AR : Aortic regurgitation
ECG : Electrocardiography
EF : Ejection fraction
LA : Left atrium
LV : Left ventricle
MR : Mitral regurgitation
NETs : Neuroendocrine tumors
POC : Physician outpatient clinic
VR : Ventricular rate

Introduction

Neuroendocrine tumors (NETs) are infrequent diverse neoplasms with varying incidence from 2.5 to 5 cases per 100,000 population [1]. These tumors can affect anybody site but commonly arise in the gastrointestinal tract (GIT). Gastrointestinal NETs were previously described as carcinoids. If these developed from the distal small intestine and proximal colon, NETs will be named midgut carcinoids [2]. Bu if arise from the stomach, duodenum, and bronchus NETs will be named foregut carcinoids. Rarely, NETs arise from the distal colorectal tract and are named hindgut carcinoids. Carcinoids are commonly slowing expanding tumors that may be over years. Few or asymptomatic presentations are usual until they are large developing or general body metastasizing [3]. Nearly, 30% to 40% of cases of fundamentally midgut carcinoids present with symptoms of carcinoid syndrome. Carcinoid syndrome is often presented with episodes of flushing and hypotension but less frequently; hypertension, diarrhea, and bronchospasm [4]. Most carcinoid syndrome is associated with liver metastasis, producing vasoactive substances, which reach the systemic circulation via the hepatic vein. Approximately, 5% of patients of primary ovarian or pulmonary sites and midgut carcinoids with retroperitoneal metastases can be present with carcinoid syndrome without liver metastases [5]. Carcinoid heart disease represents plaque-like, fibrous endocardial thickening involving heart valves (essentially in the right heart) and usually manifested as carcinoid syndrome but with considerable morbidity and mortality. The pathophysiology of carcinoid heart disease is still unknown. Chronic exposure to excessive circulating serotoninergic 5-hydroxytryptamine [5-HT]) substance is the main contributing factor. Symptoms of right heart failure (RHF) may occur but subsequently causing considerable disability of life [6]. Due to carcinoid, heart disease represents an independent negative prognostic factor in patients with advanced 5-HT–secreting neuroendocrine tumors; there is a strong need to improve early diagnosis and management of carcinoid syndrome or carcinoid heart disease. International consensus guidelines regarding the

diagnosis and management of carcinoid heart disease are still defective [7].

Case presentation

A 39-year-old married housewife Egyptian female patient presented to the physician outpatient clinic (POC) for cardiovascular follow-up. She gave a history of abdominal tumors since two years ago. Facial flushing, hypotension, headache, and recurrent nonspecific dull aching abdominal pain were the main past complaints. One year of these symptoms, she began to complaining of palpitation, diarrhea, chest pain, wheeze, and recurrent episodes of syncope. The abdominal tumor was diagnosed as a neuroendocrine tumor and severe aortic regurgitation due to probable rheumatic heart disease. Tumor biopsy showing well-differentiated neuroendocrinal carcinoma. There was no history of rheumatic fever. Two months after the diagnosis of severe aortic regurgitation she has undergone mechanical aortic valve replacement. After prosthesis replacement, the patient continued on warfarin (2.5 mg PO, nightly, once daily), torsemide (10 mg PO, fasting, once daily), bisoprolol (5 mg PO, after breakfasting, once daily), and candesartan cilexetil (8 mg PO, after breakfasting, once daily). Informed consent was taken. Upon general physical examination; generally, currently, the patient appeared thin appearance with a regular pulse rate; 86 bpm), blood pressure (BP) of 110/70 mmHg, respiratory rate of 16 bpm, the temperature of 36.7 °C, and pulse oximeter of oxygen (O2) saturation of 98%. The current ECG tracing showing a normal sinus rhythm of VR; 88 (Figure 1). The initial echocardiography was done within 2 days of preoperative aortic valve replacement showing thicken aortic cusps with severe aortic regurgitation (grade III/IV AR, peak/systolic gradient 26/13 mmHg, and aortic annulus 2.3cm), thicken both mitral leaflets, especially at tips with characteristic doming with mild MR (grade I-II/IV MR), dilated left atrium (LA), dilated left ventricle (LV) with EF of 39%, and diastolic dysfunction with reversed E/A%. (Figure 2). The echocardiography was repeated within 2 months of postoperative aortic valve replacement showing well-suited well-functioned aortic valve metallic prosthesis with trivial aortic regurgitation (peak/systolic gradient 20/10 mmHg), thicken both mitral leaflets, especially at tips with characteristic doming with mild MR (grade I-II/IV MR), dilated LA, dilated LV with EF of 36%, and diastolic dysfunction with reversed E/A%. (Figure 3). Carcinoids tumor complicated with aortic regurgitation and low-EF stable heart failure was the most probable diagnosis. The patient is advised to be still on the above medications. Further recommended cardiac, oncological, and endocrinal follow-up was recommended.

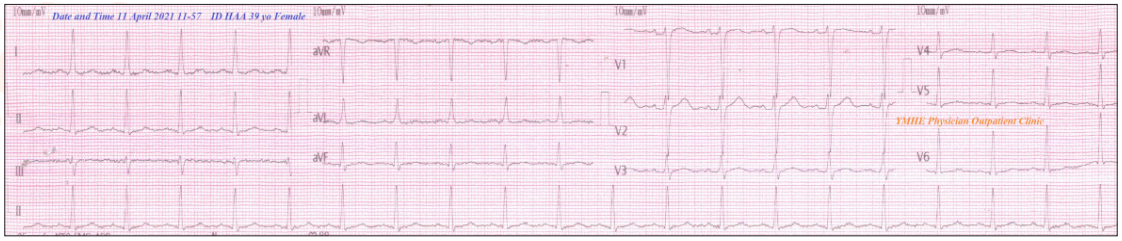

Figure 1: ECG tracing; was done within one year of aortic valve replacement showing normal sinus rhythm of VR of 88 bpm.

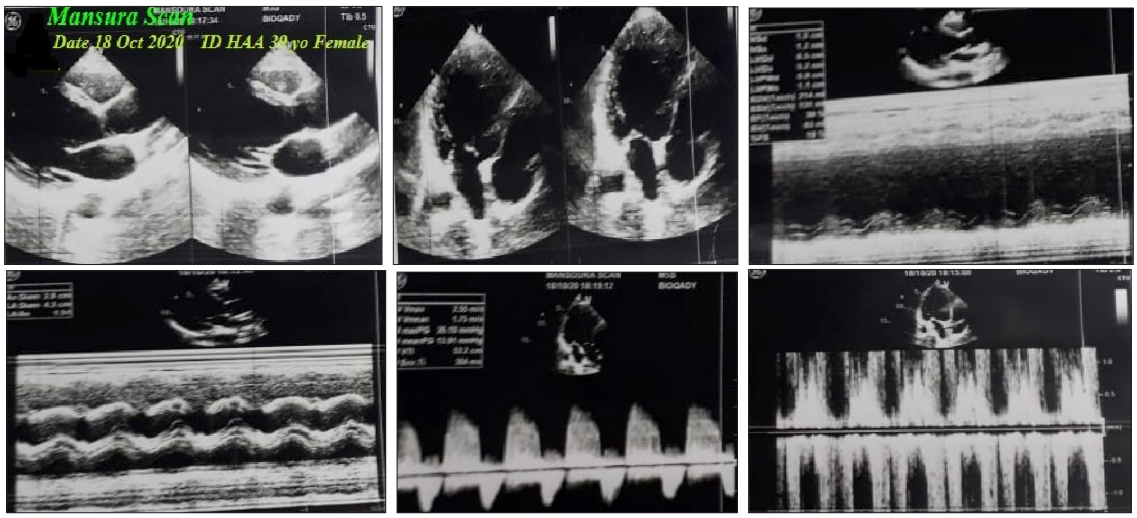

Figure 2: The initial echocardiography was done within 2 days of preoperative aortic valve replacement showing thicken aortic cusps with severe aortic regurgitation (grade III/IV AR, peak/systolic gradient 26/13 mmHg, and aortic annulus 2.3cm), thicken both mitral leaflets, especially at tips with characteristic doming with mild MR (grade I-II/IV MR), dilated left atrium, dilated left ventricle with EF of 39%, and diastolic dysfunction with reversed E/A%.

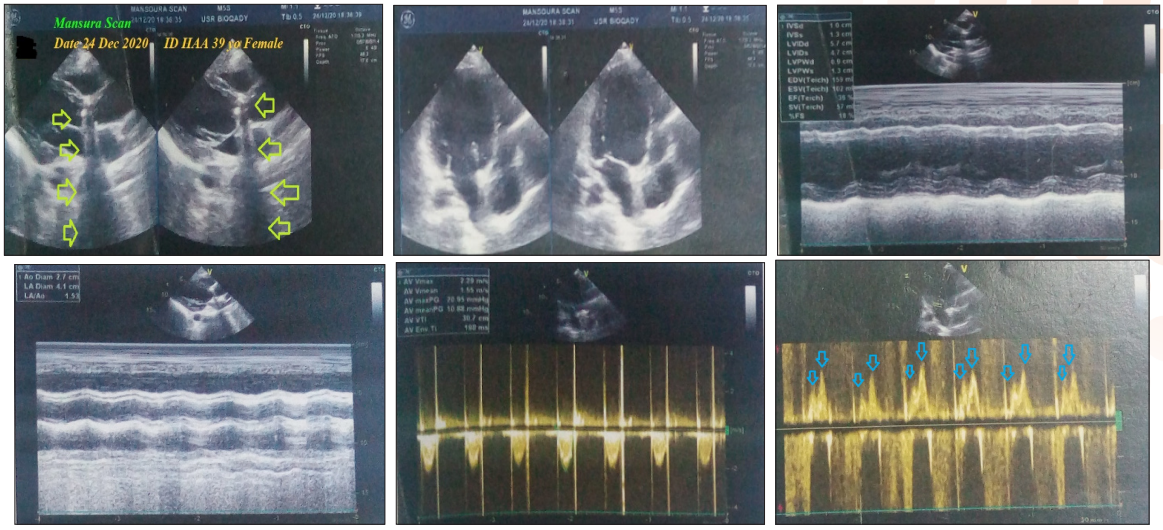

Figure 3: The echocardiography was repeated within 2 months of postoperative aortic valve replacement showing well-suited well-functioned aortic valve metallic prosthesis (lime arrows) with trivial aortic regurgitation (peak/systolic gradient 20/10 mmHg), thicken both mitral leaflets, especially at tips with characteristic doming with mild MR (grade I-II/IV MR), dilated left atrium, dilated left ventricle with EF of 36%, and diastolic dysfunction with reversed E/A% (blue arrows).

Discussion

Overview

- A young-aged housewife female patient presented to physician outpatient clinic for cardiovascular follow-up for abdominal tumors with aortic valve replacement.
- The primary objective for my case study was the presence of a patient with neuroendocrine tumors (NETs) with aortic valve replacement in POC.
- The secondary objective for my case study was the question of; how did you manage the case at home?
- The presence of episodes of facial flushing, hypotension, headache, abdominal pain, palpitations, diarrhea, recurrent syncope, chest pain, and wheeze, maybe point to possible carcinoid syndrome and gut affection.
- Synchrony of aortic regurgitation with the existence of an abdominal tumor, episodes of vasomotor changes, and biopsy are suggested for neuroendocrine or carcinoid tumors.
- Valvular heart diseases were excluded from the patient's history.
- The absence of the above symptoms and symptoms of heart failure after aortic valve replacement despite low EF and the still present tumor may carry diverse outcomes.
- Valvular heart disease and congenital heart disease were the possible differential diagnosis for the current case study.
- I cannot compare the current case with similar conditions. There are no similar or known cases with the same management for near comparison.
- The only limitation of the current study was the unavailability of interventional catheterization.

Conclusion and Recommendations

- Valvular heart disease may be occurring with carcinoid tumors but the wrong diagnosis for the rheumatic heart is common.
- The fixation of low myocardial ejection fraction with absent heart failure is an innovative interesting issue.
- The absence of the above symptoms and symptoms of heart failure after aortic valve replacement despite low EF and the still present tumor may carry diverse outcomes.

Conflicts of interest

There are no conflicts of interest.

Acknowledgment

I wish to thanks my wife to save time and improving the conditions for supporting me.

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