

Heart Failure As Initial Presentation of a Devious Metastatic Carcinoid Tumor

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Introduction

- Carcinoid syndrome usually presents with flushing, palpitations, secretory diarrhea, bronchospasm and hypotension.
- Progressive exertional dyspnea indicating possible carcinoid heart disease may be the initial presentation in 20% of patients. Physicians should highly suspect carcinoid heart disease in patients presenting with vague gastrointestinal complaints, progressive dyspnea, lower extremity edema and/or holosystolic murmur. Identifying such patients with early carcinoid heart disease has a great health impact and can lead to better prognosis.
- We describe a patient presenting with heart failure and echocardiogram findings of carcinoid heart disease as an initial presentation of a devious metastatic carcinoid.

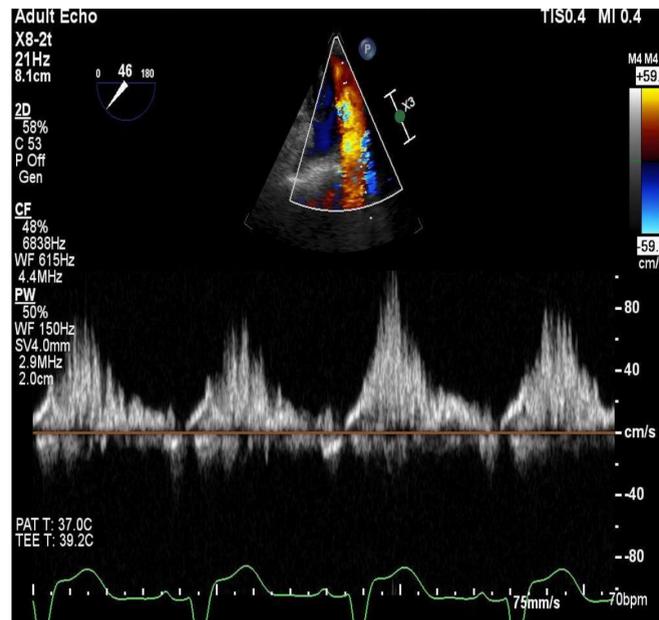


Figure 1. Transthoracic echocardiogram (TEE) color doppler showing severe tricuspid regurgitation jet.

Case Description

- A 74-year-old African American female known to have Type 2 diabetes, HTN, dyslipidemia, COPD, asthma, left breast cancer s/p mastectomy and hormone therapy, vitiligo and gout presented with progressive exertional dyspnea of 6-months duration and a 1-month history of diarrhea.
- Her physical exam was pertinent for T 36.2°C, BP 136/86, HR/80, RR/18 saturating 95%, visible vitiligo on face, hands and feet, JVD 12cm with prominent v wave, loud blowing holosystolic murmur in LLSB, upper and lower extremities with puffy pitting edema, abdominal ascites and bilateral crackles in mid lobe and bases.
- Laboratory tests were significant for hypernatremia at 147 mmol/L, hypokalemia at 2.9 mmol/L, elevated creatinine at 1.65 mg/dL, BUN at 23 mg/dL and elevated NT-proBNP at 4,866 pg/mL.
- A transthoracic echocardiogram (TTE) showed normal left ventricular systolic function with EF of 50-55% and severe right ventricle and atrial dilatation, severe tricuspid regurgitation and dyssynergia with a flattened septum suggesting RV pressure-volume overload. Transesophageal echocardiogram (TEE) for tricuspid valve assessment was made which showed severe flail motion of anterior, posterior and septal leaflet and wide open regurgitation which was very suspicious for carcinoid (Figure 1). She subsequently underwent right heart catheterization which showed elevated right-sided filling pressures and low cardiac output/ cardiac index. She was admitted to the hospital and was treated with aggressive diuresis and started on inotropic agent dobutamine.
- Further investigations looking for infiltrative diseases including ANA with reflex, rheumatoid factor, SPEP/UPEP, and Kappa/Lambda ratio were all normal. MRI of the abdomen without contrast showed multiple heterogenous solid masses throughout the liver consistent with metastatic disease (Figure 2).
- Serum serotonin, 24 hours 5-HIAA and chromogranin A were elevated (Table 1). Somatostatin analog octreotide was started and fine-needle aspiration of the right liver lesion was positive for a low-grade/well-differentiated neuroendocrine tumor (Figure 3).
- The patient was maintained on octreotide therapy for symptomatic relief and no further management was pursued due to critical health condition.

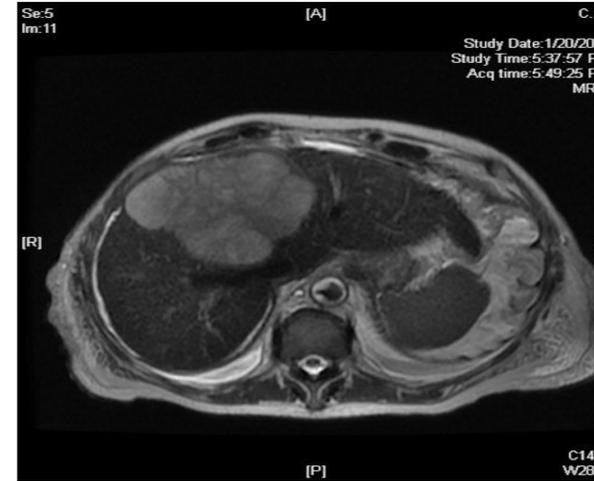


Figure 2. MRI without contrast demonstrate a mass within the central portion of the liver involving both the right and left lobes measures 11.2 x 8.5cm.

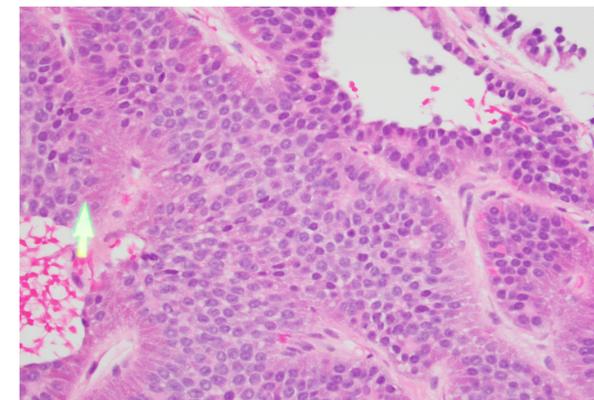


Figure 3. Right lobe liver lesion core biopsy, 400X (arrow). Positive for Neuroendocrine tumor, low-grade/well-differentiated.

	Patient laboratory values	Normal Values
Serum serotonin	930 ng/mL	50-200 ng/mL
Urinary 5-hydroxyindoleacetic acid (5-HIAA) urine per 24 hour	117 mg/d	0-15 mg/d
5-HIAA urine ratio to creatinine	225 mg/gCr	0-14 mg/gCr
Chromogranin A	43, 257 mg/ml	0-160 pg/mL

Table 1. Laboratory values

Discussion

- Carcinoid heart disease has an insidious presentation. Patients may be asymptomatic until the tumor metastasizes to the liver.
- Clinicians should be vigilant of signs and symptoms of carcinoid heart disease.
- A high suspicion coupled with serologic and imaging tests is crucial for earlier diagnosis.
- The left side of the heart is usually spared because the lungs metabolize the vasoactive substances before reaching the left-sided heart valves. Rarely, when the disease is found on the left side of the heart usually a right-to-left shunt is present.
- Typically echocardiography shows right ventricular volume overload with enlarged right ventricle and tricuspid valve leaflet thickening, retracted valve with impaired mobility of the valves leading to regurgitation and/or stenosis.
- Early detection of carcinoid heart disease can improve prognosis and symptoms in patients with heart valve involvement.
- Treatment is aimed for symptomatic relief with somatostatin-analogs, telotristat therapy and temporary diuretic therapy with definitive surgical valve replacement.

References

- Agha AM, Lopez-Mattei J, Donisan T, et al. Multimodality imaging in carcinoid heart disease. *Open Heart* 2019;6:e001060.doi:10.1136/openhrt-2019-00106
- Fox DJ, Khattar RS. Carcinoid heart disease: presentation, diagnosis, and management. *Heart*. 2004 Oct 1; 90(10): 1224-8. has been cited by the following article: Article. Right Heart Failure as "Sole" Presentation of Carcinoid Syndrome.
- Ram P, Penalver JL, Lo KBU, Rangaswami J, Pressman GS: Carcinoid heart disease: review of current knowledge. *Tex Heart Inst J*. 2019, 46:21-27. *Tex Heart Inst J*. 2019, 46:21-27. Ganeshan D, Bhosale P, Yang T, Kundra V: Imaging features of carcinoid tumors of the gastrointestinal tract .