Case Report

Carcinoid syndrome associated severe tricuspid regurgitation

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Abstract

Background: Carcinoid heart disease is a serious complication of metastatic neuroendocrine tumors caused by prolonged exposure to circulating serotonin and other vasoactive substances. It predominantly affects right-sided cardiac valves and commonly presents severe tricuspid regurgitation. We report a 65-year-old female with Grade II ileal neuroendocrine tumor with liver metastasis evaluated for preoperative cardiac fitness. Echocardiography revealed severe tricuspid regurgitation, severe pulmonary arterial hypertension (PASP 67 mmHg), dilated right-sided chambers, and mild left ventricular dysfunction (EF 45%). Coronary angiography demonstrated normal epicardial coronary arteries. Histopathology confirmed a well-differentiated Grade II neuroendocrine tumor with chromogranin and synaptophysin positivity and Ki-67 index of 5–6%. The findings were consistent with carcinoid heart disease. Early recognition is essential for optimal perioperative and subsequent post-operative management of TR.

Keywords: Carcinoid heart disease; Neuroendocrine tumor; Tricuspid regurgitation; Pulmonary hypertension; Ki-67

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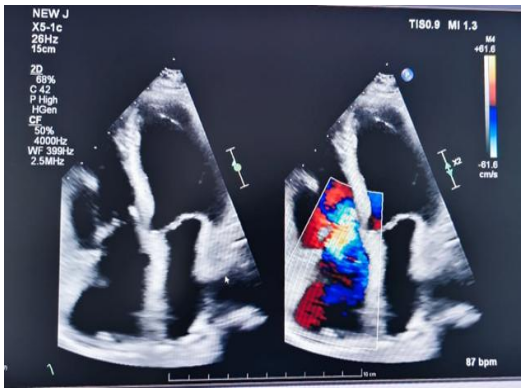
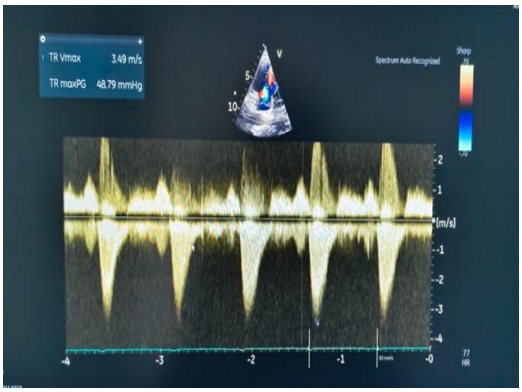
1. Case Presentation

A 65-year-old female with well-differentiated neuroendocrine tumor of the ileum extending to the caecum with liver metastasis was referred for cardiac evaluation prior to ileal resection and liver metastatectomy. She presented with progressive exertional dyspnea (NYHA Class III). She was a known hypertensive and had history of bronchial asthma.

Electrocardiogram showed normal sinus rhythm with left bundle branch block. Transthoracic echocardiography revealed mild concentric left ventricular hypertrophy with global hypokinesia and ejection fraction of 45%. The right atrium and right ventricle were dilated. The tricuspid valve appeared thickened and non-coating with severe tricuspid regurgitation (broad jet). Severe pulmonary arterial hypertension was present

with pulmonary artery systolic pressure of 67 mmHg. Trivial mitral regurgitation was noted.





For perioperative risk stratification, coronary angiography was performed and showed normal left main, left anterior descending, left circumflex, and right coronary arteries.

2. Histopathological Findings

Microscopic examination showed tumor cells arranged in nests and trabeculae composed of uniform round to polygonal cells with eosinophilic cytoplasm and finely stippled "salt and pepper" chromatin (Fig 1).

Immunohistochemistry demonstrated strong cytoplasmic positivity for chromogranin A and synaptophysin (Fig 2). Ki-67 immunostaining revealed a proliferative index of approximately 5–6% (Fig 3), consistent with Grade II (intermediate grade) neuroendocrine tumor.

Based on clinical, echocardiographic, and pathological findings, a diagnosis of carcinoid heart disease was made. The patient was initiated on diuretic therapy and cleared up for surgery with moderate cardiac risk.

3. Discussion

Carcinoid heart disease develops in approximately 20–50% of patients with metastatic carcinoid syndrome [1,2]. Persistent exposure to serotonin results in fibrotic plaque deposition on endocardial surfaces, predominantly affecting the tricuspid and pulmonary valves [3].

Severe tricuspid regurgitation is the most common manifestation and may lead to progressive right heart failure [4]. Echocardiography remains the diagnostic modality of choice, demonstrating thickened, retracted, non-coapting leaflets with severe regurgitation [5]. Pulmonary hypertension may occur secondary to chronic volume overload and right ventricular dysfunction [6].

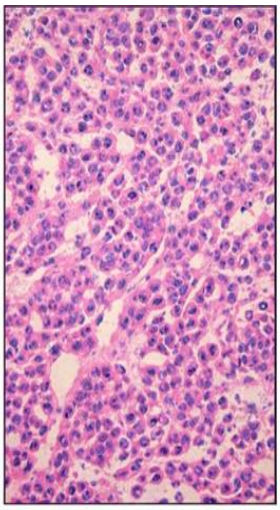


Figure 1
Well-differentiated Grade II tumor cells arranged in nests and trabeculae with H&E; 40×).

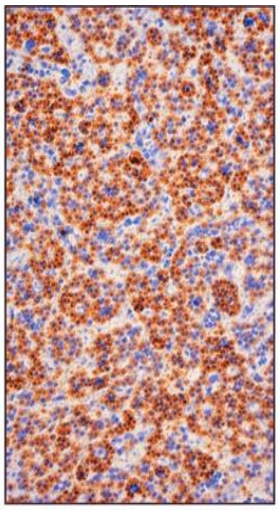


Figure 2
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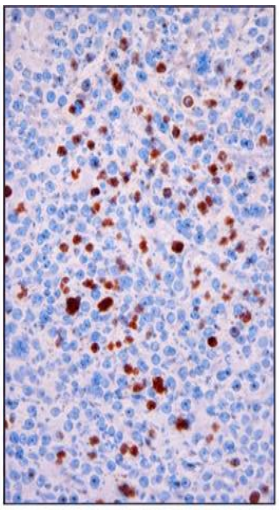


Figure 3
Ki-67 immunostaining showing proliferative index of approx 5-6% (40×).

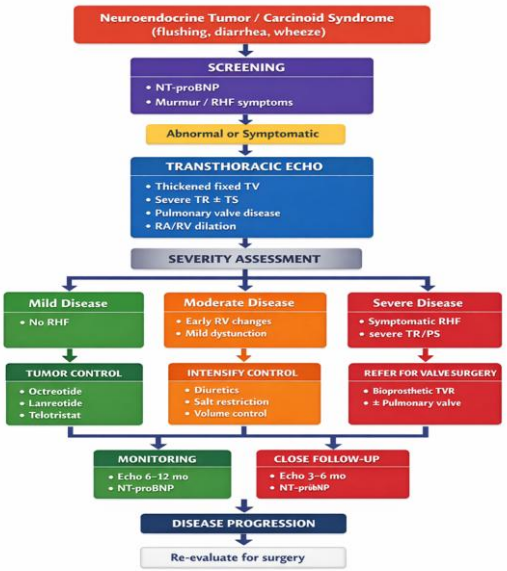
In the present case, normal coronary angiography excluded ischemic etiology of left ventricular dysfunction, reinforcing the diagnosis of serotonin-mediated valvular disease.

Histopathological confirmation of Grade II neuroendocrine tumor with chromogranin and synaptophysin positivity and Ki-67 index of 5–6% supported the diagnosis of metastatic carcinoid syndrome, the underlying cause of cardiac involvement.

Early identification is crucial, as severe tricuspid regurgitation significantly increases perioperative risk [7].

4. Conclusion

Carcinoid heart disease should be suspected in patients with metastatic neuroendocrine tumors presenting with right-sided valvular dysfunction. Severe tricuspid regurgitation with pulmonary hypertension and normal coronary arteries strongly suggests serotonin-induced valvular pathology. Comprehensive cardiac evaluation plays a vital role in surgical planning and prognosis.



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