

Introduction

Carcinoid heart disease is a rare disease affecting around 20% of patients with carcinoid syndrome. It is considered a result of high circulating serotonin levels depositing heart valves. Serotonin is normally cleared by the liver, lung and brain but the higher burden causes deposition into the heart. The breakdown of serotonin (5-HIAA) can be detected in the urine

Case Presentation

A 48 year old woman presented to the ED for shortness of breath. She recently visited her outpatient doctor's office and her entire body was red, especially her face and right arm. She was tested in the outpatient for Antistreptolysin A which was positive and completed a 10 day course of cephalexin. Unfortunately, the redness did not go away. She also had some abdominal distress with nonbloody diarrhea and tachycardia in the 110's.

Other relevant labs include:

- 5-HIAA value of **40mg/24h**
(Reference <2 to 9 mg/24hr)
- Anti IgA Tissue Transglutaminase
Negative
- Epinephrine **54**
(Reference Range: 10 to 200)
- Norepinephrine **1070**
(Reference Range: 80 to 520)
- Dopamine **162**
(Reference Range: 0 to 20)

She lost approximately 30 lbs in the last 2 months because of this chronic diarrhea. Recent celiac serology testing has been negative.

An echocardiogram was performed revealing ejection fraction of 60-65% with Moderate Tricuspid Regurgitation.

CT Abdomen/ Pelvis with Contrast showed a prominent, spiculated nodal mass at the root of the mesentery. There is focal nodular thickening of small intestine wall.

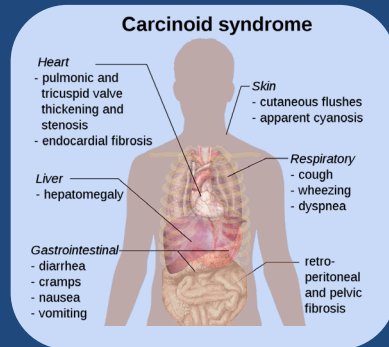


Figure 2: Common Symptoms seen in Carcinoid Syndrome ** Cureus

Discussion

Carcinoid syndrome refers to various symptoms mediated by well-differentiated neuroendocrine tumors (NETs) of the digestive tract and lungs. NETs produce, store, and release polypeptides, biogenic amines, and prostaglandins.

NETs present in the GI tract (55%), lungs (30%), and less commonly other locations such as the ovaries and kidneys. Within the GI tract, most NETs are in the small intestine (55%), most commonly in the ileum. Other locations include the rectum (20%), appendix (16%), colon (11%), and stomach (7%).

Clinical features:

- Cutaneous flushing: Episodic flushing is the clinical hallmark and occurs in 85% of patients. Associated with midgut NETs.
- Venous telangiectasias appear in late course carcinoid syndrome.
- Secretory diarrhea in 80% of patients.

The characterized symptoms of carcinoid syndrome are vasomotor flushing, gastrointestinal hypermotility and cardiac involvement. Right heart valve disorders increase morbidity and mortality due to right heart failure complications of fluid overload.

Carcinoid heart disease is caused by the deposition of pearly fibrotic plaques into the endocardium of right sided valves leading to multiple patterns of severe valve dysfunction. Plaque formation causes leaflet thickening, and fusion of the valves. Echocardiography remains a reliable method for the definitive diagnosis. Valve replacement is currently the gold standard treatment for symptomatic carcinoid valve disease.

Conclusion

The surgical indications of carcinoid heart disease are symptomatic right ventricular failure, severe valvular dysfunction and systemic venous pressure elevation. Contraindications are end-stage metastatic disease poorly controlled carcinoid symptoms despite octreotide therapy medical management. This case highlights the rare syndrome of carcinoid heart disease with follow up surgery in the outpatient.