Carcinoid Syndrome Arising from the Descending Colon causing Carcinoid Heart Disease

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Abstract

Carcinoid heart disease is a rare but impactful sequelae for patients with carcinoid syndrome, associated with a poor long-term prognosis (Ram, Yuan). Patients with classic features of flushing, diarrhea, and bronchospasm require attention and workup for carcinoid syndrome. Patients with florid carcinoid syndrome have a 50% likelihood of cardiac involvement [5]. Carcinoid heart disease occurs when large amounts of vasoactive substances reach the right heart, cause endocardial injury, and resultant plaque is deposited at the endocardial surface of the right sided heart valves, papillary muscles, and chordae tendineae [5,11,12]. Symptoms vary depending on the affected structures, but most often are consistent with right heart failure. Diagnosis can be difficult and requires a high degree of clinical suspicion to obtain serologic studies, imaging, and often right heart catheterization. Treatment requires a multidisciplinary approach, involving medical and surgical management. Appropriate treatment allows for improved quality of life and survival in these cases [5]. Here we will discuss the case of a 74 year old gentleman with carcinoid disease, with metastasis to the liver, and known carcinoid heart disease, requiring tricuspid and pulmonic valve replacement.

Introduction

Carcinoid tumors are a rare entity, arising in about 1.2 to 2.1 per 100,000 individuals in the general population every year [1]. However carcinoid heart disease only arises in about 50% of these individuals with about 20% of these individuals initially presenting due to cardiac complaints [2,3]. Carcinoid tumors most commonly arise from the gastrointestinal tract, specifically at either the midgut and occasionally also arise from the bronchopulmonary system.

Once there is cardiac involvement it is associated with a poor long term prognosis with an estimated 3 year survival rate of only 31% [4,5]. The most commonly affected areas of the heart include the pulmonic valve and the tricuspid valve. The left side of the heart is usually not affected unless there is a patent foramen ovale or there are very high levels of vasoactive substances which include: serotonin, histamine, bradykinin, tachykinins, prostaglandins and 5-hydroxytryptophan which overwhelm the pulmonary arterial circulation.

Carcinoid tumors classically arise from the midgut which includes the jejunum, ileum, appendix and ascending colon. However on rare occasions it can arise from the hindgut which includes the transverse, descending colon, sigmoid colon, rectum and genitourinary system. Approximately only 11% of carcinoid tumors arise from the colon [1]. Carcinoid tumors of the colon only represent 1% of colonic neoplasms [1]. Patients with colonic carcinoid tumors at diagnosis usually have local nodal or distant metastases with an overall 5 year survival rate of 25%-41% [6].

Case Presentation

Patient is a 74 year old male with a past medical history of carcinoid disease with metastasis to the liver, which he receives monthly Octreotide injections at Moffitt, that presented to our hospital for an elective pulmonic valve and tricuspid valve replacement on 11/4/1029 with our cardiovascular surgery team. The surgery had a successful placement of a 27mm freestyle bio-prothesis PV and 31mm bioprosthetic valve TV, without any complication.

He required multiple pressors after surgery including various intervals of milrinone, levophed, epinephrine, and vasopressin while being on an octreotide drip. He was initially sent to the ICU on the ventilator with epicardial leads and 2 chest tubes, which were eventually removed before discharge. He was successfully extubated shortly after surgery on 11/4, but on 11/7 a rapid response was called due to altered mentation, decreased responsiveness, and dyspnea, and the patient was subsequently intubated again.

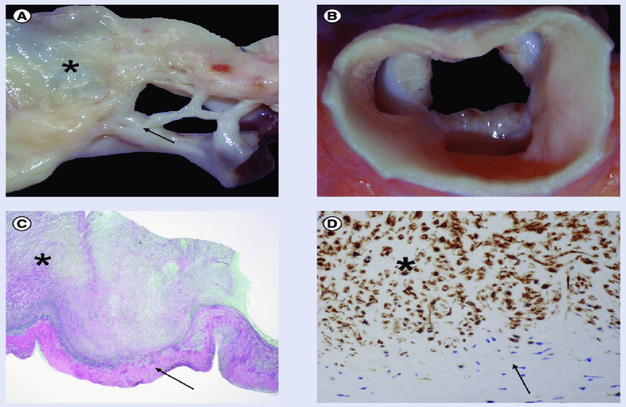

Figure A showing Tricuspid Valve Carcinoid Heart disease, Figure B showing Pulmonic Valve Carcinoid Heart disease. Figure C shows florid myofibroblastic cell proliferation and Figure D shows myxoid stroma with myofibroblastic cell proliferation [13].

He was placed on epinephrine and vasopressin at that time and underwent an emergent TTE, which wasn't adequate so a TEE was done by cardiology. TEE showed an LVEF 55-60%, normal RV cavity size/function, mild-moderate AI, mild MR, tricial TR, normal PVR, no intracardiac shunt on the bubble study, and well seated bioprothetic valves. A weaning trial was performed and he was extubated on 11/9/2019. His hospital course was also complicated by a stent exchange from extrinsic compression of the carcinoid tumor which he has changed every 3 months and was started on Rocephin then Omnicef for an associated UTI. He also developed new onset atrial flutter which resolved with amiodarone, and was started on Eliquis.

Discussion

Gastrointestinal neuroendocrine tumors, formerly known as carcinoid tumors, are rare (incidence of 5 per 100,000 persons/year) and derived from the neuroendocrine producing cells of the gastrointestinal tract.

Despite most commonly originating from the bronchus, jejenoileum and the rectum, GI-NETs can arise from any part of the GI tract. Typically, patients do not present with the classical findings of intermittent flushing, intestinal hypermotility resulting in diarrhea and bronchospasm [7].

Carcinoid Heart Disease is a rare manifestation of the carcinoid syndrome thought to be a result of the action of the many vasoactive substances produced and released by the NETs. These substances result in the deposition of plaque on the endocardial surfaces of the cardiac chambers as well as onto the valve leaflets, chordae tendinae and papillary muscles.

In most cases, this plaque deposition occurs on the right side of the heart and may result in the simultaneous development of tricuspid and pulmonic valve disease which when occurring simultaneously can be considered as pathognomonic of carcinoid heart disease.

The mainstay of treatment for carcinoid heart disease is valve replacement of the tricuspid and pulmonic valves. Indications for valve replacement for a patient with carcinoid heart disease and associated valvular involvement do not differ from standard indications which include impaired exercise tolerance and fatigue, as well as a decline in ventricular function. Valve replacement surgery appears to improve survival in these patients. Surgical resection of cardiac metastases can be done at the time of valve surgery. Prior to valve surgery medical therapy should be optimized with somatostatin-analogs however alone, they do not reverse valvular disease or valvular lesions [8,9,10].

Conclusion

Carcinoid heart disease is a rare cause of right heart failure. Carcinoid heart disease can occur in nearly 50% of patients with carcinoid syndrome [5]. Key diagnostic factors include 24-hour urinary excretion of 5-HIAA, and TEE [5,11]. Medical and surgical treatment can significantly improve overall quality of life and long-term survival [5]. Valve replacement surgery is currently the gold standard treatment for symptomatic carcinoid valve disease [11]. Results of ongoing clinical trials will be important to achieve further improvements in early diagnosis, medical management, and surgical management, including alternative options for patients who are high-risk for interventions.

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