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Pure right heart failure: A rare atypical presentation of carcinoid tumor

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Typically, carcinoid tumors present with signs and symptoms of abnormal hormone production, flushing being the most common sign. We report a case of a 61-year-old postmenopausal woman with history of uterine fibroid and hypertension, with an atypical carcinoid syndrome who presented with right heart failure. The patient presented to the emergency department with a 4-week history of progressive abdominal distention, bilateral lower extremity swelling and dyspnea of exertion. The abdominal pain was aggravated by food intake and associated with generalized colicky abdominal pain. Initially, she noticed shortness of breath with exertion which progressively got worse with minimal exertion and was associated with orthopnea and paroxysmal nocturnal dyspnea. She also noticed yellowish discoloration of the eyes. She does not have any prior medical history of hepatitis or alcohol consumption. On physical examination, she was afebrile, blood pressure of 190/109 mmHg, HR 104 bpm, respiratory rate 20. She had scleral icterus, abdominal distention and bilateral pitting grade II lower extremity edema. Lung auscultation revealed bilateral posterior-basal rales, and a holosystolic murmur was heard in tricuspid area. Diagnostic and therapeutic abdominal paracentesis was performed. Laboratory values revealed BUN 18, total albumin 3.2, total bilirubin 1.2, and direct bilirubin 0.6, serum albumin ascites gradient 2.2. CXR showed cardiomegaly with small pericardial and pleural effusion. EKG revealed low voltage QRS complexes. In addition to ascites, ultrasound abdomen identified enlarged liver with multiple heterogeneous and echogenic lesions consistent with hepatic metastases. These findings were further confirmed by CT abdomen that also revealed an irregularly shaped mesenteric mass of 4.4 x 4 x 3.4 cm, with no enlarged periaortic, iliac or inguinal lymph nodes. Echocardiography showed tricuspid valvular disease consistent with carcinoid tumor, severe tricuspid regurgitation, right atrial dilation and ventricular dilatation with EF of 58%, patent foramen ovale. Urinary 5 hydroxyindoleacetic acid (5-HIAA) and serum chromogranin-A were significantly elevated. On liver biopsy, nests and cords of tumor cells with pleomorphic hyperchromic nuclei and eosinophilic granular cytoplasm were noted. On immuno- histochemical staining, all the tumor markers were negative with the exception of synaptophysin, chromogranin, CD56, CDX2, MOC-31, monoclonal CEA and uroplakin. Based on histology and the immunohistochemical staining, the diagnosis was consistent with metastatic neuroendocrine carcinoma with a Ki 67 of 30%. Diffuse positivity of CDX-2 stain was suggestive of neuroendocrine tumor from gastrointestinal tract possibly pancreatic or hepatobiliary origin. Carcinoid tumors are relatively rare tumors with an incidence of 1 in 75,000 population. Most of the carcinoid tumors originate from the enterochromaffin cells of midgut. About 50% carcinoid tumors after a very indolent course, present with carcinoid syndrome. The onset of carcinoid syndrome usually follows metastases of midgut carcinoid tumor to the liver; following which carcinoid secretory products including serotonin (5-HT) are not completely inactivated. Higher concentration of carcinoid secretory products upon reaching the heart induce TGF- β mediated fibrous plaque formation. Other possible fibrinogenic mediators include tachykinins, neurokinins and substance P. Fibrous plaques are formed on the valvular leaflets, subvalvular apparatus, chordae tendineae, papillary muscles and subendocardium. The fibrous plaque formation in tricuspid valvular apparatus on the ventricular side and subsequent adhesion of the tricuspid valve to mural endocardium results in tricuspid insufficiency and hence regurgitation. Pulmonary stenosis is attributed to fibrous plaque involvement of the annulus. Pulmonary stenosis may worsen tricuspid regurgitation. There is direct correlation between degree of fibrous plaque formation, blood serotonin level and 5 hydroxyindoleacetic acid (5-HIAA) in urine. Up to 20% of the patients with carcinoid syndrome present with carcinoid heart disease, which includes tricuspid regurgitation and/or pulmonary stenosis along with the other typical manifestations of carcinoid syndrome. Our patient's symptoms and signs suggest a metastatic unresectable carcinoid tumor with longstanding high levels of serotonin which led to right-heart insufficiency due to tricuspid valve fibrosis. To the best of our knowledge this is the fourth reported case of carcinoid syndrome secondary to midgut carcinoid tumor that presented as isolated or pure right heart failure.

Biography

Pramod Theetha Kariyanna, MD, obtained his medical degree from Kempegowda Institute of Medical Sciences, India. Following which he served as a research scholar in Center for Arrhythmia research at the University of Michigan, Ann Arbor, U.S.A. He is presently a physician resident in Department of Internal Medicine at the Brookdale University Hospital and Medical Center, Brooklyn, NYC. He is a representative in the research committee at Brookdale University Hospital. He is active in academic writing and is a contributing author for the "First Aid for the USMLE Step 1" and he is chief image editor for the "Master the boards USMLE Step 2 CK". His interest lies in cardiovascular medicine and he plans to serve as an academic physician.

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