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Etiology of Enlarged Heart: A Rare Pancreatic Tumor

Divey Manocha, Nidhi Bansal, Hani Kozman

SUNY Upstate Medical University, Syracuse, NY, USA

Background: Pancreatic endocrine tumors account for < 1% of all pancreatic cancers.

Methods: We report an exceedingly rare and unique case of a 70–year-old woman with past history of hypertension and arthritis who presented with worsening pedal edema, dyspnea and upper abdominal discomfort. Physical exam revealed jugular venous distension, normal breath sounds, systolic murmurs over pericardium, hepatomegaly and B/L 3+ pedal edema. Metabolic profile and TSH were within normal limits. Chest X-ray revealed moderate cardiomegaly. Echocardiogram showed aortic, tricuspid and pulmonary regurgitation with left ventricular hypertrophy. A diagnosis of diastolic heart failure was made and appropriate medical therapy was instituted. Lack of significant symptomatic improvement created a management dilemma for the primary team. Her hepatomegaly previously thought to be secondary to heart failure also persisted. This resulted in sonogram of the upper abdomen which surprisingly showed 2 large masses (> 5 cm) in the right lobe of liver. Hepatitis viral serology and AFP tests were normal. CT scan of abdomen and pelvis detected a new 3 cm mass in tail of pancreas and multiple hepatic lesions to further add to the mystery. A core biopsy of the hepatic mass lesion showed complete replacement of liver by tumor cells with round nuclei and abundant cytoplasm containing eosinophilic granules. Immunostains show marked positivity for synaptophysin and chromogranin, confirming their neuroendocrine origin. She subsequently underwent a somatostatin receptor scintigraphy to reveal increased uptake in pancreas and multiple sites in liver. EUS of pancreas was also done to delineate the tumor extent. Blood chromogranin levels were extremely high at 2885 ng/ml (normal, ≤ 50 ng/ml). Urine 5-HIAA was 67 mg/g creatinine (normal, ≤ 14 mg/g).

Results: A diagnosis of primary pancreatic carcinoid with hepatic metastases with carcinoid syndrome was made. The patient refused any aggressive interventions like radiofrequency ablation or chemoembolization. She was initiated on Sandostatin therapy with partial relief of her symptoms.

Conclusions: Pancreas is an exceedingly rare location for carcinoid tumors, accounting for < 0.6% cases. Prevalence of symptomatic pancreatic carcinoids is even rarer (estimated to be 1:10). Most frequent symptoms associated are abdominal pain (66%) and diarrhea (52%). Our patient presented with abdominal pain and carcinoid heart disease which is secondary to infiltration of cardiac chambers and valves. Pancreatic carcinoids are difficult to diagnose early due to subtle presentation. High index of suspicion is needed for timely diagnosis as metastatic disease carries a poor prognosis.