

Diagnosis and treatment of ATTR cardiomyopathy during cancer staging work up

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A 91-year-old male with no specific medical underlying disease, visited a primary care clinic for epigastric discomfort that had persisted for several years. Esophagogastroduodenoscopy revealed an ulcerofungating mass in the antrum of the stomach and he was referred to a tertiary hospital after tubular adenocarcinoma was confirmed by biopsy. During cancer staging work up, there were no evidence of bone metastasis in 99mTc-hydroxymethylene diphosphonate bone scintigraphy but it showed prominent radiotracer uptake in the heart region. Echocardiography showed hypertrophy in left ventricle (LV) with highly reflective myocardium and diastolic dysfunction with enlarged left atrium. Speckle tracking imaging showed an 'apical sparing' pattern on the bull's eye plot of global longitudinal strain. Cardiac magnetic resonance imaging revealed septal LV hypertrophy, late gadolinium enhancement base-apex gradient, pericardial effusion, and pleural effusion. 99mTc-3, 3-diphosphono-1, 2-propanodicarboxylic acid scan and single-photon emission computed tomography showed intense myocardial uptake suggestive of transthyretin amyloid cardiomyopathy (ATTR-CM). Endomyocardial biopsy showed pericellular interstitial deposition of amorphous and pale pink material, positive birefringence on Congo red staining and positive TTR immunohistochemical staining. AL amyloid cardiomyopathy was ruled out based on serum free light chain concentration and serum, urine immunofixation electrophoresis. Genetic testing for TTR showed no mutation. On multidisciplinary discussion with the patient and family members, we decided to perform stomach cancer surgery because his functional capacity has been well preserved. After curative gastrectomy, the patient has been doing well for more than a year with tafamidis treatment for ATTR-CM. With growing awareness, incidental detection of ATTR-CM on bone scintigraphy during cancer staging work-up may increase, and further investigation for optimizing treatment strategy in these patients are required.

