To understand the epidemiology and presentation of acromegalic cardiomyopathy with symptoms of diastolic heart failure with restrictive physiology

On admission: BP 193/111, HR of 63, RR 24, saturating 97% on NC

Elevated GH and IGF

Treat HF with preserved EF: control volume status, BP, pulse

To appreciate the interactions between growth hormone (GH) and insulin

An ECG (Figure 2) and CXR (Figure 3) performed: ECG demonstrated atrial

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Patients often experience increased # of valvular abnormalities and arrhythmias, such as:

Treat acromegaly, with hopes of cardiac benefit: somatostatin analog

Two years post procedure with close follow up by heart failure team, neurosurgery,

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Learning Objectives

• To understand the epidemiology and presentation of acromegalic cardiomyopathy with symptoms of diastolic heart failure with restrictive physiology

• To appreciate the interactions between growth hormone (GH) and insulin-like growth factor I (IGF-1) for the maintenance of normal cardiac function

• To elaborate on the status of GH/IGF-1 in relation to heart failure and the potential use of GH antagonists as a tool in the adjunctive treatment of heart failure

Case Presentation

• A 55-year-old African American man with a hx of HTN presented to the ED with chronic, acutely worsening dyspnea, chest pain, and fatigue.

• On admission: BP 193/111, HR of 63, RR 24, saturating 97% on NC

• On admission, in significant cardiopulmonary distress, speaking 3-word sentences. JVD present, diffuse crackles bilaterally. PMI laterally displaced and diffusely palpable in the 5th intercostal space. Rhythm was irregularly irregular heart without murmurs, ruts, or gallops. 3+ pitting edema below the knees bilaterally. Strength and sensation intact. Exam notable for macroglossia, and extremely large hands and feet.

• An ECG (Figure 2) and CXR (Figure 3) performed: ECG demonstrated atrial fibrillation, a rate of 61/min, intra-ventricular conduction block; and right axis deviation; no pathologic Q waves seen. CXR showed massive cardiomegaly with pulmonary vascular congestion and right lower lobe consolidation

• Patient was sent for to evaluate the hemodynamic status as diuretics had not been as brisk as previously. Figure 4 provides initial laboratory data

• Thorough physical exam demonstrated massively enlarged feet, macroglossia. In addition, CT scan showed massively enlarged cardiac chambers, leading to a focused differential diagnosis: acromegaly, chronic atrial fibrillation, valvular disease, cardiomyopathy (Figure 5)

• Echocardiogram showed LVEF = 60%, severe concentric left ventricular hypertrophy, massive left and right atrial enlargement at 11.6 x 15.9 cm (444.7ml/m2) and 11.5 x 5.54 cm, respectively (Figure 6, 7, 8)

• Patient was felt to be in decompensated heart failure with preserved EF

• After implementation of traditional HF therapy, coronary angiogram was performed, revealing non-flow limiting coronary artery disease

• Additional history obtained: recently incarcerated, was receiving “weekly injections for therapy”. He had grown four inches taller (now 6’ 8’’) and increased in height from a shoe size of 16 to 20. Was the star power forward for his college basketball team!

• Elevated GH and IGF-1 levels at 11.5ng/mL and 700ng/ml respectively (normal 81-225). No GH suppression after 75-g oral glucose tolerance test. Pituitary MRI demonstrated mild asymmetric prominence on left side of the pituitary gland, revealing microadenoma.

• With signs and symptoms of heart failure in a setting of preserved EF with a diagnosis of acromegaly, options included:
  - Treat HF with preserved EF; control volume status, BP, pulse
  - Treat acromegaly, with hopes of cardiac benefit: somatostatin analog
  - Treat heart failure and acromegaly concurrently

• We implemented dual therapy: diuretics, anti-hypertensives, pulse control, low salt diet, somatostatin analogs, and regular follow-up

• Surprisingly, our patient continued to have frequent decompensations of heart failure.

• Endonasal endoscopic transsphenoidal surgery to resect the pituitary microadenoma was performed; repeat echo showing improvement of LV hypertrophy

• Two years post procedure with close follow up by heart failure team, neurosurgery, and endocrinology, patient has had no further admissions for decompensated heart failure, has a normal GH level

• Patients with acromegaly have a 30% higher mortality rate with 60% of deaths due to cardiovascular complications – hence immediate diagnosis and management required for better control of acromegaly

• Acromegaly is a rare disease with an annual incidence of 4 cases/mil and a prevalence of 40 cases/mil, characterized by increased GH and IGF-1 levels

• “Acromegalic cardiomyopathy” is characterized by eccentric hypertrophic hypertrophy, diastolic dysfunction, progressive systolic impairment, and eventual heart; our star basketball power forward lost his ability to pump forward due to diastolic dysfunction

• Patients often experience increased # of valvular abnormalities and arrhythmias, such as:
  - ectopic beats, paroxysmal atrial fibrillation, paroxysmal supraventricular tachycardia, BBB
  - GH impacts structure and function of the normal adult heart by stimulating cardiac growth and contractility and regulating vascular tone and peripheral resistance.
  - Cardiovascular complications, including cardiomyopathy and arterial hypertension, should be evaluated and followed serially.

• Transsphenoidal surgery is far better for microadenomas than macroadenomas, which unfortunately represent the vast majority, >70%; all GH-secreting pituitary adenomas; hence, medical therapy is vital and is based on GH-lowering medications.