HYPERTROPHIC OBSTRUCTIVE CARDIOMYOPATHY IN THE SETTING OF SYSTEMIC SCLERODERMA

R Sogomonian,1 H Alkhawam,1 S Lee,1 D Chang,1 EA Moradoghli Haftevani2.1 Internal Medicine, Icahn School of Medicine at Mount Sinai Elmhurst Hospital Center, Elmhurst, New York, United States;2 Ross University, New Jersey, New Jersey, United States

Restrictive cardiomyopathy has been a common variant seen in systemic sclerosis (SS) with myocardial fibrosis. The association of SS with restrictive cardiomyopathy has well been established, but that with HOCM is not clearly understood. Herein, we report a case of a patient with SS, identified to have both HOCM and myocardial fibrosis.

A 54-year-old woman with systemic sclerosis, idiopathic lung disease with moderate pulmonary hypertension, presented with fatigue, decreased appetite and shortness of breath. Vital signs were significant for oxygen saturation of 86% on room air, tachycardia of 117 bpm, and blood pressure of 110/53 mm Hg. Physical examination revealed diffuse rhonchi in all lung fields, malar rash and skin excoriation in bilateral lower extremities without edema. Laboratory studies were significant for elevated brain natriuretic peptide (BNP) of 858 pg/mL. Transthoracic echocardiography revealed left ventricular hypertrophy (LVH) with ejection fraction of 78%. Electrocardiography illustrated LVH. Cardiac magnetic resonance imaging (cMRI) was significant for severe left ventricular cardiac asymmetric septal hypertrophy with outflow obstruction caused by anterior motion of the mitral valve. Cardiac biopsy revealed evidence of diffuse fibrosis, but did not show iron, glycogen, or amyloid depositions.

Patient was maintained on mycophenolate mofetil, low dose of methylprednisolone, morphine, clonazepam and transferred to hospice care.

Hypertrophic obstructive cardiomyopathy (HOCM) is the most common genetic cardiac disorder with an autosomal dominant transmission. It is characterized by asymmetric LVH out of proportion of systemic after load. The most common cardiac involvement in SS is myocardial fibrosis in a restrictive pattern, while HOCM is rarely seen in SS.