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SYSTEMIC SCLERODERMIA AND HYPERTROPHIC CARDIOMYOPATHY - CAUSAL OR STOCHASTIC AFFILIATION

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Introduction

Hypertrophic cardiomyopathy (HCM) is characterized by the presence of left ventricular hypertrophy which cannot be explained only by ventricular filling abnormalities. HCM has been previously described in a small number of patients with systemic scleroderma (SDS).

Keywords

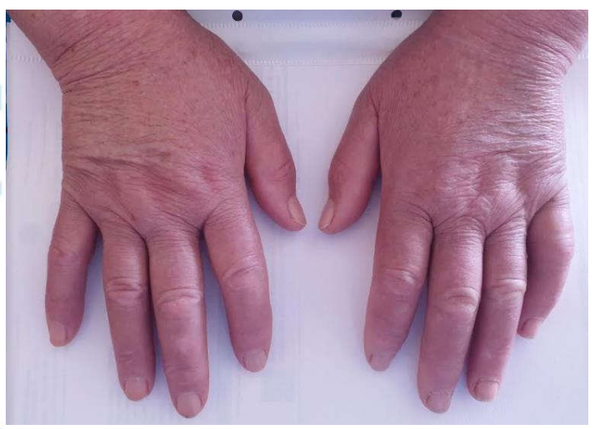
systemic sclerosis, hypertrophic cardiomyopathy.

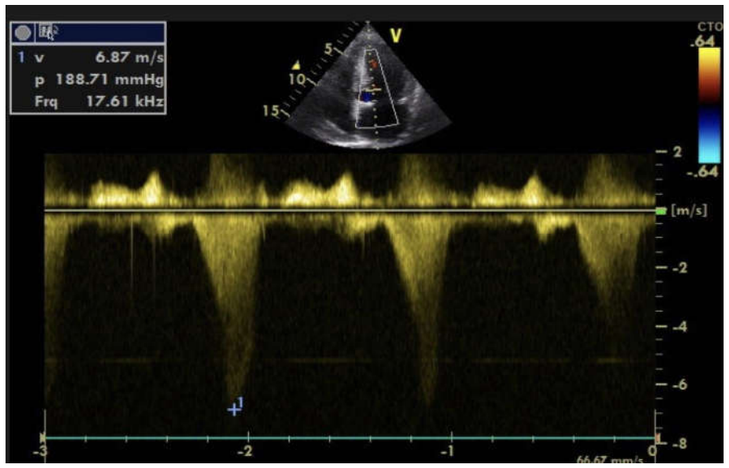
Purpose

To highlight the importance of the multidisciplinary approach to a patient with systemic sclerodermia.

Material and methods

Patient with paresthesia at low temperatures, discoloration of the fingers, dysphagia, arthralgias, thickening and stiffness of the skin, fatigue and dyspnea was examined clinically and paraclinical.





Results

Clinical and paraclinical parameters: BP-130/80mmHg, HR-74bpm; PCR-22.9 mg / L, ESR-21 mm / h, pro-BNP-2461 ng / ml, positive Scl-70, ANA-1/5120, HLA-DR3 was positive; ECG-sinus rhythm, LV myocardial hypertrophy. Trans-thoracic echocardiography: LV diastolic dysfunction, ejection fraction 61%, severe obstruction of the LV ejection tract. HCM is an autosomal dominant genetic disorder associated with HLA-DR3 genes, acting with genetic and non-genetic factors, in which the link to SDS is perceived. Diffuse connective tissue disease can be considered a "natural experiment" in the interaction between inflammation and heart disease, which could elucidate the fundamental mechanisms by which inflammation accelerates the development of cardiovascular disease.

Conclusions

This affiliation can be interpreted as two concomitant diseases or a causal association.