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Cardiac tamponade in a patient with mixed connective tissue disease

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Introduction: Mixed connective tissue disease (MCTD) is an overlap syndrome with clinical features of systemic lupus erythematosus (SLE), polymyositis and scleroderma typically with U1 RNP antibodies. The presence of pulmonary arterial hypertension is a common complication of MCTD. Cardiac involvement is frequent and the disease can affect not only the myocardium but also the pericardium at any stage of the disease.

Clinical case: We present the case of a 50-year-old woman followed-up at our lupus clinic for MCTD with interstitial lung disease, myocarditis and pulmonary hypertension, ANA title 1/640 and RNP positive. The patient was on prednisolone (PDN) 5mg, mycophenolate mofetil 2.5g, hydroxychloroquine 400mg, bosentan 250mg, riociguat 6mg and treprostinil 0.046mg/h.

The patient was admitted to hospital due to progressive worsening of exertional dyspnea, paroxysmal nocturnal dyspnea and peripheral edema over the last month. At physical examination, she presented muffled heart sounds, bilateral lung crackles and symmetrical lower limb edema. The electrocardiogram showed low QRS voltage in all limb lead. CRP <0.5mg/dL, pro-BNP 6948pg/mL and urinalysis with no proteinuria. Transthoracic echocardiogram depicted a pericardial effusion that measured 2 to 5.1 cm surrounding the heart without evidence of diastolic collapse of the right chambers, with pulmonary hypertension (PSAP 82mmHg). The chest x-ray showed an increased cardiac silhouette with "water bottle heart" and bilateral small pleural effusion. The patient was started on i.v. diuretics and pulsed with 1g of methylprednisolone (mPDN) for 3 days with progressive signs of cardiogenic shock after 3 days. The patient underwent pericardiostomy with a large-bore subxiphoid surgical drainage and 2000mL of serous pericardial fluid was removed over 3 days. Culture of pericardial fluid was negative. A serositis related to MCTD was admitted. There was clinical improvement the day after the procedure, she followed a tapering PDN scheme and the patient was discharged one week after and remained stable in class II NYHA at 6 months follow-up.

Conclusion: Pericardial disease is the most common cardiac manifestation in patients with MCTD, affecting up to 40% of patients. Usually presents as pericarditis with or without effusion. Large effusions and cardiac tamponade are rarely seen (<2% of patients). Treatment with nonsteroidal anti-inflammatory and high dose of steroids is usually sufficient. Rarely it is needed the drainage of the fluid. Since our patient had severe pulmonary hypertension the pericardial effusion reached important dimensions and was clinically significant without signs of haemodynamic compromise. The presence of impeding cardiogenic shock without response to high dose of mPDN was the indication for drainage with large bore subxiphoid surgical approach, with good results. The following scheme of PDN and immunosuppressive therapy is important to avoid relapse of pericardial disease.