

## Improvement of Dilated Cardiomyopathy with Methylprednisolone in a Patient with Multiple Fibrosclerosis

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### ABSTRACT

Multifocal fibrosclerosis is a rare syndrome of unknown cause that is characterized by fibrosis involving multiple organ systems. Definitive diagnosis can only be made based on biopsy findings. In this case, the biopsy specimen of the patient demonstrates pulmonary hyalinated granuloma or sclerosing mediastinitis. There are few reports of multiple fibrosclerosis with heart failure. Here, we reported a case of retroperitoneal fibrosis with massive mediastinal involvement extending to pleura and pericardium causing pleuro- pericardial effusion with dilated cardiomyopathy. Systolic dysfunction was improved and pericardial effusion disappeared with methylprednisolone treatment.

### ► Implication for health policy/practice/research/medical education:

This case report helps us remember the etiology of the dilate cardiomyopathy. Fibrosclerosis is a rare reason for dilate cardiomyopathy which can be easily treated with steroids. In case a patient refers with idiopathic dilate cardiomyopathies, physicians must research for the reasons. Cardiac MRI can be used to detect the fibrosis. Early administration of steroid therapy for heart failure symptoms due to left ventricular systolic dysfunction and/or pericardial involvement may be beneficial in the patients with multifocal fibrosclerosis.

### 1. Introduction

Retroperitoneal Fibrosis (RPF) is a rare disease with peak incidence in the fifth to seventh decades of life. The symptoms of RPF may be general/nonspecific or localized (due to replacement or compression of organs). Yet, definitive diagnosis can only be made based on biopsy findings. Besides, Magnetic Resonance Imaging (MRI) is essential for evaluating the extent of the disease process (1). However, some patients require ureteral, intestinal, or paravascular surgery due to obstruction in spite of medical treatment. There are few reports of retroperitoneal and mediastinal fibrosis with pleural and pericardial involvement

described as multifocal fibrosclerosis (2). Here, we present a case of retroperitoneal fibrosis with massive mediastinal involvement extending to pleura and pericardium causing pleural, pericardial effusion, and heart failure, with improvement after methylprednisolone administration.

### 2. Case Presentation

The case was a 47-year-old man who was diagnosed with mediastinal fibrosis 5 months ago with thoracoscopic biopsy after presenting emphysematous changes, retroperitoneal-paravertebral pleural thickening, and pleural effusion on thorax CT. He was admitted to internal medicine with complaints of easy fatigue, vomiting, and abdominal pain. The patient was thin. On physical examination, thyroid was palpated as stiff. Besides, bilateral bronchovesicular rales were heard on the lungs and pleural frotman was absent. His

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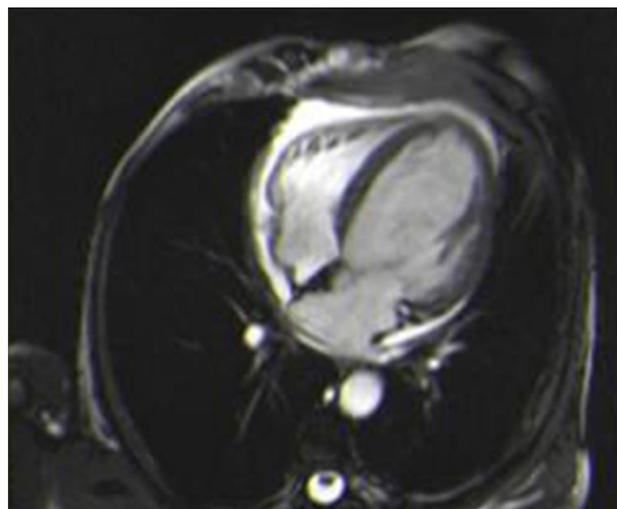
Blood pressure was 140/80 mmHg on either arms, his heart rate was 88 beats per minute, s1 and s2 were rhythmic, s3 and s4 were absent, and 2/6 pansystolic murmur was heard from the apex. Epigastric sensitivity was observed with deep palpation and 5 cm of hepatomegaly was palpated. ECG revealed sinus rhythm and incomplete right bundle branch block. In addition, cardiomegaly with mediastinal widening was observed on chest roentgenogram. Reticulonodular consolidation areas were also detected in the left side of the lung. There was a marked increase in C-reactive protein and the Erythrocyte Sedimentation Rate (ESR), but the serology for connective tissue disease and perinuclear antineutrophil cytoplasmic antibodies was negative. Thyroid function tests were normal. Nevertheless, creatinine level was elevated to 3.7 mg/dL. Using abdominal ultrasound, grade II hydronephrosis was determined and MR urography was performed; soft tissue from basal pole of both kidneys extending to minor pelvis, on paraaortic area and on iliac chains, was like retroperitoneal fibrosis surrounding both ureters at iliac crossing level. After consultation with the urology department, bilateral pigtail catheter was implanted. Enlarged cardiac chambers, severe Left Ventricular (LV) systolic dysfunction (Ejection fraction: 24%), severe mitral regurgitation, and moderate pericardial effusion were revealed by echocardiogram and conventional treatment for heart failure was started, except for inhibitors of the renin-angiotensin-aldosterone system. During the follow up, creatinine level was elevated up to 9 mg/dL in conjunction with hyperkalemia, fever, nausea, and vomiting. However, the symptoms were relieved by hemodialysis and administration of intravenous ciprofloxacin. Creatinine level was also fixed at 4 - 4.5 mg/dL. Additionally, nephrostomy cannula was affixed and urinary output was increased up to 6 liters per day. Then, the creatinine level decreased to 1.6 mg/dL and remained stable thereafter.

Ureterolysis was planned after 3 months. We were concerned about the malignancy and performed thoracoscopic biopsy from the most involved site on Positron

Emission Tomography (PET) (anterolateral mediastinum on the level of left 6 - 7 the intercostal space). Pathological evaluations revealed cross-sections of thick walled vessels, hyalinated connective tissue with lymphoid cells on some areas, pulmonary hyalinated granuloma, and sclerosing mediastinitis. According to cardiac MRI, we considered myocarditis as a cause of dilated cardiomyopathy (Figure 1), but did not perform biopsy. Initially, the patient received methylprednisolone 32 mg per day, tapering 4 mg every 2 weeks. After initiation of corticosteroid treatment, the patient felt better and denied heart failure symptoms. Control echocardiogram was performed two weeks later. Accordingly, ejection fraction was increased to 35% and pericardial effusion was minimal. The patient was discharged with desirable clinical conditions. During outpatient follow-up 1.5 months later, the control echocardiogram showed ejection fraction to be 45% (Figure 2).

### 3. Discussion

RPF is a rare disease with peak incidence in the fifth



**Figure 1.** Cardiac MRI Revealing Low Ejection Fraction with Dilated Cardiac Chambers and Diffuse Enhancement of Myocardium Compatible with Myocarditis



**Figure 2.** Transthoracic Echocardiogram Showing the Left Ventricle. Ejection Fraction Was Calculated as 45%

to seventh decades of life. In the present study patient who was a 47-year-old man, the etiology was to a great extent unclear. However, its occasional association with autoimmune diseases and its response to corticosteroids and immunosuppressive therapy suggested that it was probably immunologically mediated.

Definitive diagnosis of RPF can only be made based on biopsy findings. MRI is also essential for evaluating the extent of the disease process (1). Besides, PET can be used to guide biopsy (3). In the present study, PET was performed and biopsy was taken from the most involved area. Pathology reported pulmonary hyalinated granuloma or sclerosing mediastinitis. These concepts are no established terms in the literature. After restoring the function of the involved (hollow) organs, medical therapy with prednisone, immunosuppressive drugs, or tamoxifen was started aiming at converting the active disease to stable disease. Multifocal fibrosclerosis is a rare syndrome that is characterized by fibrosis involving multiple organ systems. However, presentation of mediastinal-retroperitoneal fibrosis is rare. In one series of 491 patients, mediastinal involvement was found in only 3.3% of the idiopathic RPF cases (2). Nonetheless, there were few reported cases of pericardial involvement in this combined disorder (4). In the case reported here, the patient had retroperitoneal fibrosis and massive mediastinal fibrosis with pleuro-pericardial involvement.

There were some reports of multifocal fibrosclerosis with dilated cardiomyopathy as case reports in the literature (5). The patient reported here had these features, as well. To the best of our knowledge, no report of myocarditis and improvement of dilated cardiomyopathy with methylprednisolone treatment is available in the patients with multifocal fibrosclerosis.

Omura et al. in 2006 reported a case of multifocal fibrosclerosis combined with idiopathic retro- peritoneal and pericardial fibrosis who had massive pericardial effusion

and died with clinical signs of cardiovascular failure despite pericardiostomy and aggressive treatment (6). Early administration of steroid therapy may be beneficial for heart failure symptoms due to LV systolic dysfunction and/or pericardial involvement in the patients with multifocal fibrosclerosis. However, there is no agreement on the dose and duration of steroid therapy.

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#### **Authors' Contribution**

All the authors contributed to all stages of performance of the case report.

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