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Dermatomyositis Following Heart Transplantation: A Case Report and Literature Review

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Abstract

Background and Objectives: Cardiac involvement is well recognized in cases of dermatomyositis (DM) and polymyositis (PM), with a variable frequency ranging from 9% to 72%. However, clinically significant heart involvement in DM/PM is relatively uncommon, and there are few reports of cardiac transplantation in DM. Our objectives were to describe a case of severe cardiac involvement in DM necessitating heart transplantation and to review the literature on cardiac involvement in DM and PM.

Methods: We describe a case of dermatomyositis in a patient with severe heart failure who underwent heart transplantation at our institution. Pathological examination of both the patient's explanted heart and skeletal muscle is reviewed. Additionally, a search of the MEDLINE database was conducted to identify reports of cardiac involvement in DM and PM.

Results: A 36-year-old man with DM presented with severe heart failure to our institution and underwent evaluation for heart transplantation. After a three-month hospitalization, he underwent successful cardiac transplantation. Pathological examination of his explanted heart revealed a pattern of inflammation and damage similar to that observed in DM in skeletal muscle. The patient is currently doing well, 20 months post-transplant, and is receiving maintenance therapy with tacrolimus, mycophenolate mofetil, rituximab, and low-dose prednisone. To our knowledge, this is the first case report of heart transplantation in dermatomyositis with analogous pathology observed in both cardiac and skeletal muscle.

Conclusions: Severe cardiac involvement necessitating transplantation is rare in dermatomyositis but can occur and appears to be related to a similar inflammatory process as observed in skeletal muscle.

Keywords: Dermatomyositis; Seditious myopathy; Cardiomyopathy; Cardiac transplantation; Orthotopic heart transplant

Introduction

Dermatomyositis (DM) and polymyositis (PM) are both idiopathic inflammatory myopathies (IIM) characterized by proximal muscle weakness and inflammatory cell infiltrates within the skeletal muscle. Cardiac involvement, including conduction abnormalities, arrhythmias, congestive heart failure, valvular/pericardial/coronary artery disease, and left ventricular dysfunction, has been reported as a common cause of death in these conditions. Severe cardiac involvement in IIM is rare, with only two reported cases of cardiac transplantation in IIM patients, one involving PM and the other showing giant cell myocarditis on cardiac muscle pathology. In this report, we present a case of severe cardiac involvement in DM necessitating heart transplantation and review the existing literature on cardiac involvement in DM and PM [1].

Material and Methods

A 36-year-old African American male, previously in good health, presented to an external medical facility with widespread muscle pain and weakness primarily affecting the proximal muscles. He reported difficulty in raising his arms above his head and climbing stairs. Additionally, he exhibited a pruritic, erythematous rash on his upper back and chest, along with itching and swelling around his eyes, a hoarse voice, and swelling and stiffness of his hands. Laboratory tests revealed a significantly elevated creatine phosphokinase (CPK) level of 12,006, and MRI of bilateral femurs showed extensive muscle edema [2]. He was initiated on prednisone at a dose of 80 mg daily for suspected myositis. Subsequently, he developed dysphagia, and a muscle biopsy of his left thigh revealed severe inflammatory myopathy characterized by perivascular inflammation and areas of facial and perifascicular atrophy consistent with dermatomyositis or its variant.

Two months after starting prednisone therapy, methotrexate was added at a dose of 15 mg daily, and the prednisone was gradually tapered. However, due to ongoing muscle weakness and elevated CPK levels after six weeks on methotrexate, rituximab was introduced. Within six months of diagnosis, the patient developed severe fatigue and dyspnea, leading to the diagnosis of cardiomyopathy with reduced ejection fraction (10-15%) and normal coronary arteries [3]. Over the subsequent four months, he experienced multiple hospital admissions at an external facility for heart failure complicated by atrial fibrillation, ventricular tachycardia, gastrointestinal bleeding with hemoptysis, and deep venous thrombosis in the lower extremity.

Given his complicated clinical course, the patient was transferred to our institution for evaluation for orthotopic heart transplantation (OHT). On admission, he exhibited residual lower extremity proximal muscle weakness and a mild hyperpigmented rash on his upper chest and back. He was receiving prednisone 10 mg daily, methotrexate 25 mg subcutaneously daily, and rituximab, which had been discontinued

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Received: 30-Jan-2024, Manuscript No troa-24-127731; **Editor assigned:** 02-Feb-2024, PreQC No. troa-24-127731(PQ); **Reviewed:** 16-Feb-2024, QC No. troa-24-127731; **Revised:** 23-Feb-2024, Manuscript No. troa-24-127731(R); **Published:** 29- Feb-2024, DOI: 10.4172/troa.1000217

Citation: Thakur A (2024) Dermatomyositis Following Heart Transplantation: A Case Report and Literature Review Transplant Rep 9: 217.

seven months prior to admission. Serologic testing revealed the presence of anti-Ku antibody. During hospitalization, he developed cardiogenic shock necessitating placement of an intra-aortic balloon pump followed by bi-ventricular assist device (BiVAD) placement. Immunosuppressive medications were not increased due to concerns about BiVAD infections as advised by the Cardiology Transplant service, which could jeopardize OHT [4].

A month after his initial admission, the patient experienced bleeding and purulent discharge from his BiVAD sites, leading to discontinuation of methotrexate and reduction of prednisone to 7 mg daily. However, his lower extremity weakness worsened, and hoarseness of voice recurred. Methotrexate was reintroduced at a dose of 10 mg weekly, but his CPK levels continued to rise, reaching 1400 IU/L. Subsequently, he was treated with moderate-dose prednisone (40 mg daily) and intravenous immunoglobulin (IVIG). Within two weeks of this flare of dermatomyositis, the patient underwent successful orthotopic heart transplantation [5].

Examination of the explanted heart revealed mildly thickened valvular cusps, with mitral chordae attached to both papillary muscles. Histological examination showed multifocal and severe fibrosing myocarditis with active myocardiocyte injury, perifascicular myofiber injury, and microvascular immunoreactivity with antibodies to membrane-attack complex (C5b9). Active mononuclear inflammation, primarily composed of T-lymphocytes, was noted, with no apparent dominance of CD4+ helper or CD8+ cytotoxic T cells.

Discussion

Cardiac involvement in dermatomyositis (DM) and polymyositis (PM) has been extensively documented in the literature, with reported prevalence rates ranging from 9 to 72%, depending on patient selection and diagnostic methods. However, clinically significant cardiac involvement is much less common [6].

A systematic review reported electrocardiogram (EKG) and Holter monitor abnormalities, such as conduction defects, ST-T changes, and frequent atrial or ventricular premature beats, in over 85% of patients with inflammatory myopathies (IIM). Other modalities for cardiac evaluation include echocardiography showing wall motion abnormalities, valvular and pericardial abnormalities in over 62% of cases, Technetium99m-pyrophosphate (99mTc-PYP) scintigraphy [7], and cardiac MRI showing abnormal enhancement in a study of 4 cases which improved after corticosteroid treatment. The most commonly reported clinical symptom of cardiac involvement is congestive heart failure, although coronary artery disease, including acute myocardial infarction, is also increased in patients with inflammatory myopathies. Patients with DM/PM have increased mortality compared to the general population, with cardiopulmonary diseases being the leading cause of death [8].

Only two case reports have been published regarding cardiac transplantation in myositis. The most recent report described a case of fulminant giant-cell myocarditis in a patient with possible DM diagnosed two weeks prior to transplantation. The patient received high doses of methylprednisolone and intravenous immunoglobulin (IVIG) but rapidly progressed to cardiogenic shock and ultimately underwent successful orthotopic heart transplantation. Histological examination of the explanted heart revealed foci of mixed inflammatory cells admixed with multinucleated giant cells/sinking myocardial filaments, consistent with fulminant giant-cell myocarditis. The other case report described a patient with polymyositis who developed severe heart failure over 7 days following diagnosis and eventually underwent successful heart transplantation. The pathology of the explanted heart was not extensively described in this report [9].

In our current report, we detail the cardiac pathology and observe parallels between the myocardium and the skeletal muscle, particularly those related to characteristic patterns of DM injury. The most notable resemblance was the perifascicular distribution of atrophy and damage in the skeletal muscle, which was matched by zones of additional fiber damage in the cardiac muscle. Intense perifascicular alkaline phosphatase reactivity, specific to DM, was also noted in both cardiac and skeletal muscle. The multifocal and variable nature of the disease from region to region was similar to the pattern of injury typically seen in dermatomyositis affecting skeletal muscle. Additionally, overexpression of the membrane attack complex (MAC) in the vasculature, a hallmark sign of DM pathology, was observed abundantly in both skeletal and cardiac muscle microvasculature in our case. These histological parallels suggest that dermatomyositis is indeed a systemic disease that can cause muscular inflammation and damage beyond the skeletal muscle [10]. However, myocardial findings were somewhat limited compared to the features observed in skeletal muscle biopsy. Nonetheless, the biopsy was performed some time prior to transplantation and may not fully represent the concurrent skeletal muscle pathology at the time of orthotopic heart transplantation.

Conclusion

While most cases of cardiac involvement in dermatomyositis (DM) may go unnoticed clinically, it is crucial to recognize that severe cases of cardiac involvement leading to transplantation do occur, and in such cases, transplantation can be life-saving. The histopathologic examination suggests a pattern of similar inflammatory damage in the heart as observed in the skeletal muscle.

Acknowledgment

None

Conflicts of interest

None

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