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Commentary

The Great Imitator: Lupus Myocarditis

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DESCRIPTION

Systemic Lupus Erythematosus (SLE) is a well-known entity in the medical field. Cardiac involvement can be seen in over 50% of lupus patients, affecting all structural components of the heart [1]. Only less than 9% of people with SLE have myocarditis [2]. We often see myocarditis described in the literature, but lupus myocarditis is less commonly reported, especially if it is co-existing with neurological complications. With varying presentations, the approach to diagnosis has remained broad.

Co-existing lupus myocarditis and neurological manifestations

Myocarditis can be a fatal manifestation of SLE, and the majority of the evidence is derived from case reports, case-control, and cohort studies. It is rare, but post-mortem analysis revealed the presence of myocarditis in 40% of cases [3]. Clinical manifestations of lupus myocarditis may vary from unexplained tachycardia to fulminant Congestive Cardiac Failure (CCF). Presenting symptoms frequently include unexplained dyspnea, palpitations, chest pain with or without increased troponin elevation, syncope, arrhythmia, acute or chronic congestive heart failure, sudden cardiac death, and fulminant cardiogenic shock. Pleuritic chest pain may occur in the presence of concomitant pericarditis. SLE associated with myocarditis has neurological manifestations such as transient ischemic attack, stroke, cerebral venous sinus thrombosis, posterior reversible encephalopathy syndrome, meningitis, meningoencephalitis, demyelinating syndrome, seizure disorder, transverse myelitis, and dysautonomia. However, the prevalence of myocarditis with each of these potential concomitant disorders is unknown [4]. No current guidelines exist for diagnosis of lupus myocarditis.

Multiple approaches to diagnosis

Currently, no single clinical feature or imaging technique is diagnostic of Light Microscopy (LM). Electrocardiogram (EKG) abnormalities are commonly non-specific in LM but EKG remains important in the identification of other causes of myocardial dysfunction. Although the prevalence of Antinuclear Antibody (ANA), anti-dsDNA, and anti-Smith (anti-Sm) in LM is similar to that of SLE seen in the general population; anti-Ro/SSA was reported in up to 69% of LM patients compared with 40% in other SLE patients [5]. Anti-Ro/SSA also predicts the presence of Late Gadolinium Enhancement (LGE), representing myocardial fibrosis and/or necrosis on Cardiac Magnetic Resonance Imaging (CMRI) [6]. Troponin is a more sensitive indicator of myocyte injury than creatine kinase [7], however, can be elevated in multiple other conditions. The histological findings of LM on light microscopy are non-specific, mimicking viral myocarditis [8]. Therefore, Endomyocardial Biopsy (EMB) is nonspecific. Due to the limitation of EMB, Imaging including Echocardiography, CMRI, and Positron Emission Tomography/Computed Tomography (PET/CT) are used in the detection of both clinical as well as subclinical myocardial involvement in SLE. Due to its non-invasive nature, Cardiac MRI is the diagnostic imaging of choice for myocarditis. In a 21-year-old woman with LM, the rest 13N-ammonia perfusion images and 18F-FDG PET were effective in following up the treatment outcome [9].

Treatment strategies

Treatment strategies in patients with lupus myocarditis and heart failure continue to be supportive and empirical immunosuppression [10]. Immunosuppression for 12 months was associated with improvement in SLE disease activity and functional CMRI parameters without significant improvement in morphological evidence of injury [11]. Immunosuppression, usually with high-dose steroids is the cornerstone of treatment, although cyclophosphamide, azathioprine, mycophenolate, and intravenous immunoglobulin can be alternative options to control disease activity. In a multicentric retrospective study comparing patients receiving cyclophosphamide therapy to those who did not, there was no statistically significant difference between the 2 groups regarding ICU stay, length of hospital stay, and median Left Ventricular Ejection Fraction (LVEF) at the onset, at 1 month, and the last visit [12]. A retrospective case-control study reported that 12 out of the 25 patients diagnosed with LM were treated with Intravenous Immunoglobulins (IVIg), with results similar to those of other immunosuppressive therapies [13]. IVIg has also been suggested as an effective therapy

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therapy for myocarditis secondary to other autoimmune diseases such as dermato-polymyositis, adult-onset Still disease, and Kawasaki disease [14]. The specific mechanism of immunomodulation in SLE is not completely understood but it involves the effect of IVIg on T and B cell intracellular signaling, the interferon signaling pathway, and the disrupted elimination of immune complexes and other cellular debris. A patient with severe SLE with LM and proliferative glomerulonephritis was successfully treated with Immunoadsorption (IA) and IVIG in addition to cyclophosphamide [15]. Immunoadsorption (IA) is an extracorporeal treatment modality that removes circulating autoantibodies and immune complexes. A 20-year-old woman with fulminant LM fully recovered with normal cardiac function after treatment with plasma exchange [16]. A 10-year monocentric retrospective cohort study evaluated the use of rituximab in refractory lupus myocarditis in three SLE patients and showed improved outcomes in all [17]. Additionally, azathioprine can also be used as steroid-sparing maintenance therapy [18]. In a suitable patient, orthotropic heart transplant can be a last resort and a case report presented a patient with orthotopic heart transplantation with no evidence of recurrence of lupus myocarditis on subsequent endomyocardial biopsies and imaging during a follow-up of 24 months [19-22].

CONCLUSION

SLE is a multisystem inflammatory disease and one of its feared complications is LM. Cerebral dysfunction can co-exist either due to direct manifestation or secondary to complications of myocarditis. Cardiac MRI is still the non-invasive diagnostic test of choice for LM as Endocardial biopsy is non-specific and invasive. Various treatment modalities can be utilized; however, the main approach is still immunosuppression. Few case reports have described the role of IVIg, but concrete evidence and guidelines are lacking in medical literature. Clinicians should be aware of early presenting symptoms and signs of myocarditis especially in patients with preexisting SLE. There are only a few studies on LM, and there is an imminent need for guidelines to establish recommendations for screening, diagnosing, and treating this condition.

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