Cardiovascular System involvement in systemic lupus erythematosus (SLE)

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Abstract:
The systemic lupus erythematosus (SLE) is considered the most important autoimmune disease which affects most body organs and systems. SLE is mostly diagnosed in middle-aged females. However, it can affect any age and both genders. Lupus patients may present with minor skin and joint symptoms up to critical internal organ damage. Both clinical and laboratory findings are needed for sure diagnosis. Cardiac disease is frequent in about 50% of lupus patients, but usually asymptomatic. However, they are critical and life-threatening, mostly due to premature and accelerated atherosclerosis. Therefore, the American Heart Association considers female patients with SLE as a high-risk group for the development of cardiovascular (CV) disease. Lately, the rate of survival of lupus patients has improved, but the patient is still at high risk of CV morbidity and mortality. Cardiovascular System involvement in systemic lupus erythematosus includes pericardial involvement, myocardial involvement, valvular disease, and coronary artery disease.

Cardiac diseases are common in lupus patients (¹). Although most of them are clinically silent, cardiac diseases in lupus can cause significant morbidity and mortality (²). Nowadays, the survival rate of lupus patients has increased, however, the cardiovascular (CV) mortality and morbidity are high (³). Lupus patients have about 7 times higher CV risk and death rates when compared with the overall population (⁴). So, the American Heart Association includes Lupus patients, especially women, as a high-risk group for CV disease (⁵).

Keywords: SLE; cardiovascular; Heart disease

Pericardial involvement
Pericarditis occurs in 11-54% of Lupus patients. Infrequently, pericarditis may occur due to other causes as viral, uremia, tuberculosis (⁶).

Clinical picture
Retrosternal or precordial chest discomfort (aggravated by lying down), dyspnea, and palpitations. Tamponade and constrictive pericarditis are rare (⁷).

Investigation:
ECG may exhibit sinus tachycardia, PR depression, and widespread ST elevation.
Transthoracic echocardiogram (TTE) is important for diagnosis (⁷). Cardiac computed tomography (CT) and/or MRI are helpful in case of constrictive pericarditis (⁸).
Myocardial involvement
Myocarditis
Myocardial involvement can occur in 3-9% of SLE patients (1). Recently, its prevalence is 0-8% due to the widespread usage of corticosteroids in lupus patients (9).

Clinical picture
Fever, dyspnea, chest pain, palpitation resting tachycardia, gallop rhythms, jugular venous distention, and new cardiac murmurs (10).

Investigation
ECG exhibit sinus tachycardia, widespread ST and T wave changes, arrhythmias, and conduction defects.

TTE measures the degree of ventricular dysfunction and low ejection fraction has been observed in most patients (11). Cardiac MRI is the best choice in the diagnosis of non-ischemic inflammation of the myocardium and so lupus myocarditis (12).

Cardiomyopathy
Lupus can cause cardiomyopathy directly due to disease activity or secondary due to CAD and/or hypertension. Also, cardiomyopathy secondary to hydroxychloroquine has been rarely reported (13). Sure diagnosis requires an endomyocardial biopsy to exclude Fabry’s disease (14).

Valvular Disease
In lupus, it is believed that valvular affection is the most common CV disease. It comprises valvular masses (Libman-Sacks vegetations), valvular thickening, valve regurge, and stenosis (15). It was reported that valvular disease cannot be prevented or cured by using steroids and/or immunosuppressives in the treatment of lupus patients (7).

Clinical picture
Verrucous endocarditis is usually asymptomatic or may lead to cardiac murmur (16). Mild regurge of the aortic and mitral valves are frequently noticed, but valvular stenosis is uncommon (1).

Investigation
Echocardiography is a valuable non-invasive technique to distinguish the valvular structure, vegetations, and degree of valvular dysfunction (17). A transesophageal echocardiogram is the modality of choice for detecting valvular lesions in lupus patients (18).

Coronary artery disease (CAD)
It is believed that 30% of the total SLE patients’ deaths are due to CAD (19). Traditional CV risk factors do not clarify the advanced rates of CVEs reported in lupus patients, (20). SLE - associated factors comprise global disease activity, disease duration, cumulative damage, aPL, and renal disease (21). These factors lead to accelerated atherosclerosis and myocardial inflammation (22).

SLE-specific risk factors of Atherosclerosis:
1-Disease activity & duration:
It was reported that an extended disease period is associated with carotid plaque (23) and coronary calcium scores. (24).

2-Renal disease:
Chronic kidney disease (CKD) and ESRD are factors that increase CV morbidity and mortality. Also, hypertension and dyslipidemia may add to this high risk. (25)

3-The role of SLE medications in relation to cardiovascular risk:
A- Glucocorticoid therapy
It is believed that a longer duration of steroid therapy and high steroid dosage is associated with a higher risk of atherosclerosis in lupus patients. (26) However, Roman et al. reported that previous or recent use of prednisone may be athero-protective due to its anti-inflammatory effects (27).

B-Antimalarial
It is believed that antimalarials are protective against CVEs and linked with a better metabolic profile regarding lipid and glucose (28).
The clinical picture of CAD
Clinical manifestations are angina pectoris and myocardial infarction. The danger of MI is augmented 50-fold in young females with SLE (29). Even after the correction of the traditional CV risk factors, the threat of MI is still higher 8-fold (30).

Diagnostic approach
Resting and exercise ECG are the standard techniques to detect CAD in lupus (31). Also, it is believed that the imaging of atherosclerotic plaques is helpful in diagnosis (32). There are invasive and non-invasive methods of imaging. Angiography is the best invasive imaging technique (33). While, B-mode ultrasound (34), CT (35), and MRI (36) comprise the non-invasive procedures.

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