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Young Stroke Following Libman-Sacks Endocarditis associated with Systemic Lupus Erythematosus and Secondary Antiphospholipid Antibody Syndrome

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Abstract

Libman-Sacks Endocarditis (LSE) is a form of nonbacterial thrombotic endocarditis (NBTE) occurring in the setting of hypercoagulable states like solid organ malignancies, systemic lupus erythematosus (SLE) and primary or secondary antiphospholipid antibody syndrome (APLS).LSE is the most characteristic cardiac manifestation of SLE. Even though the incidence of clinically significant valve dysfunction and embolic phenomenon is low when it is associated with secondary APLS, risk for embolic cerebrovascular events is high.

We present a case of a 39-year-old female who admitted with a left sided ischemic stroke ultimately turned out to be having LSE secondary to SLE and APLS. This will demonstrate the importance of having a high degree of suspicion for the diagnosis of LSE and its etiology when evaluating a young patient with stroke.

Key words: Libman -Sacks Endocarditis, Systemic lupus erythematosus, Antiphospholipid antibody syndrome

Introduction

Libman -Sacks Endocarditis(LSE) is a form of nonbacterial thrombotic endocarditis (NBTE) where the non-infective verrucous vegetations develop in the valves- mainly the mitral valve followed by aortic valve, chordae tendinea and endocardial surface.[1] In hypercoagulable states, where there is formation of fibrin-platelet thrombi, gradually organizes and leads to fibrosis and scarring with subsequent valve dysfunction.[2] NBTE was discovered in 80% of patients with certain solid organ malignancies involving the pancreas, ovary, lung in a postmortem study and in 46% of patients with autoimmune diseases like Lupus and APLS.[3]

SLE is a complex multisystemic connective tissue disorder. Cardiac involvement is a recognized complication of SLE in 30% of the cases and virtually can affect all the cardiac components including valves, endocardium, pericardium, conduction system and coronary arteries. LSE usually has an asymptomatic course in the majority but reported complications are superimposed bacterial endocarditis, thromboembolic events, and severe valvular dysfunction.[4] In the presence of lupus and secondary APLS, they can have more symptomatic disease leading to embolic phenomenon. Moyssaki et al demonstrated in his study that Libman-Sacks lesions have prognostication value and associated with lupus duration, disease activity, anticardiolipin antibodies, and APLS manifestations.[1]

Case presentation

39-year-old female admitted to Neurology unit of National hospital, Sri Lanka with sudden onset slurring of speech associated with right sided upper limb numbness and weaknesses. This was not associated with weakness of the ipsilateral lower limb, vertigo, visual obscurations, sphincter dysfunction, seizures or altered sensorium. On further questioning, she had a similar episode three months back which resolved spontaneously over three hours.

She had no fever with headache, photophobia or phonophobia. She is married for ten years with no children. She had one first trimester miscarriage and thereafter secondary subfertility. She has no preceding history of arterial or venous thrombosis or personal or family history of haemoglobinopathies. She did not use oral contraceptive pills. She is not a known patient with congenital heart disease or childhood history of rheumatic fever. She has significant alopecia with fatigue and but no weight loss, skin ulcers, joint pains, photosensitivity, renal problems, seizures, pleurisy, pericarditis, or anemia.

Her BMI was 28kg/mm^2. Her pulse was regular with a rate of 84 bpm. She was normotensive. Precordial examination was unremarkable with normally positioned apical impulse, normal heart sounds, no murmurs or pericardial friction rub or carotid bruit. Pertinent neurological examination revealed monoparesis of the right upper limb with MRC power 3, with exaggerated reflexes in addition to left sided upper motor neuron type facial nerve palsy. No sensory impairment. Rest of the system examination was normal.

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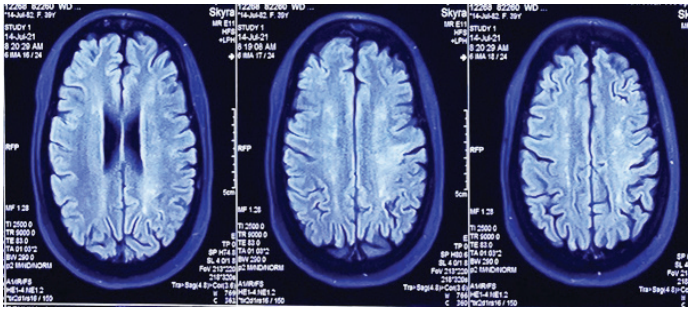

Figure 1: MRI brain showing bilateral centrum semiovale ischaemic changes

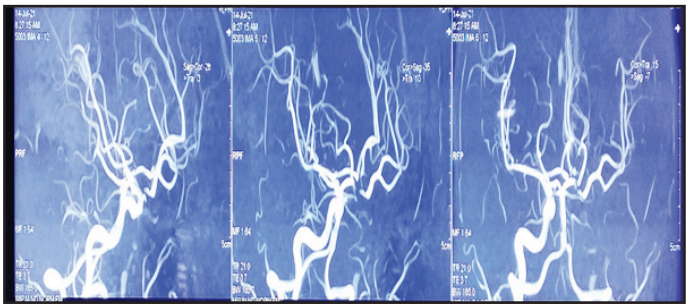

Figure 2 :MRA showing severely diseased left internal carotid artery (ICA)

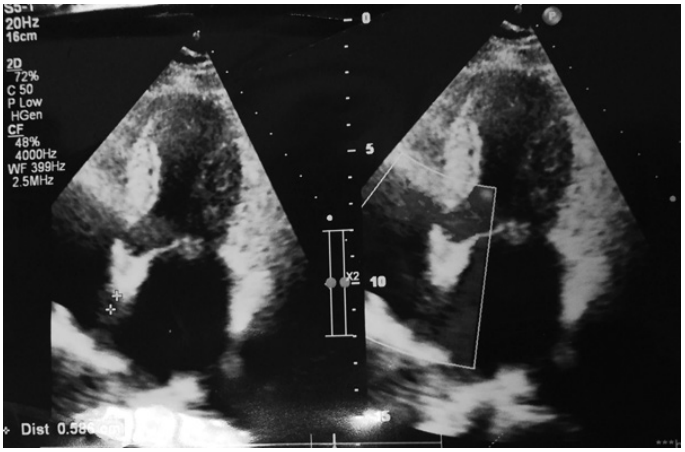

Figure 3 : Transthoracic two-chamber view demonstrating small sessile, loosely organized, fixed mitral vegetation

Her NCCT brain taken at that time was normal. MRI brain was suggestive of bilateral centrum semiovale ischaemic changes **(Figure 1)** and MRA revealed severely diseased left internal carotid artery (ICA) possibly due to vasculitis. **(Figure 2)**

For further evaluation CT carotid angiogram was done and it revealed total occlusion of the left internal carotid artery from its origin possibly due to thrombosis. Both trans-thoracic and trans-oesophageal cardiac assessment revealed good biventricular function with small atrial septal defect (ASD) with left to right shunting. There was grade 3 mitral regurgitation (MR) with eccentric regurgitant jet with coaptation failure of posterior and anterior mitral valve leaflets (MVL). Most importantly there was a 7×9 mm vegetation attached to PMVL in the atrial surface. **(Figure 3)**

Inflammatory parameters were not suggestive of infection, white blood cell count 5.87×109/L (NR: 4.0-10.0 ×109/L), C-reactive protein 1.3 mg/dl (NR: <6 mg/dl) and erythrocyte sedimentation rate (ESR) 81mm/hr (NR: <20 mm/hr). blood cultures showed no growth. C3- 112 (NR: 87-200 mg/dl) and C4 - 52 (NR: 19-52 mg/dl). Rheumatological evaluation revealed positive ANA titre >1/100 with homogenous pattern and elevated double stranded DNA. Anticardiolipin antibody IgG, IgM was positive with strongly detected lupus anticoagulant and elevated beta 2 glycoprotein Ab. All were suggestive Libmann sack endocarditis in SLE with secondary APLS.

She was started on anticoagulation therapy for APLS along with short course of high dose steroids to mitigate vegetation growth followed by weight adjusted prednisolone and hydroxychloroquine.

Discussion

This is a case vignette of a young female presented with an ischemic stroke. Careful evaluation revealed that she was having SLE associated with APLS and LSE. The cause for the ischemic stroke in this setting is multifactorial. This presentation can be a constellation of embolic phenomenon resulted from LSE and organ manifestation of SLE associated CNS vasculitis and/or neuropsychiatric SLE.

Manifestations of neuropsychiatric systemic lupus erythematosus (NPSLE) include cerebrovascular disease (CVD)in the form of stroke, transient ischemic attacks (TIA), and seizures orcognitive dysfunction or focal brain lesions on magnetic resonance imaging (MRI). CVD in SLE usually re-

sults due to cerebritis, vasculitis, hypercoagulability, and atherosclerosis.[5] However, cardio-embolic phenomenon tends to occur in the presence of anti-phospholipid antibodies. Krawczyk et al has demonstrated that presence of LSE in SLE was associated with longer disease duration, pericarditis, nephritis, recurrent stroke and APLS.[6]

LSE can present as a range from asymptomatic valvular thickening to heart failure from valvular dysfunction. It is very important to distinguish between infectious and noninfectious vegetations as treatment differs significantly. We can use the location of vegetation and clinical and laboratory markers for infective process to differentiate these two.[7]

There is no clear role regarding administration of glucocorticoid and/or cytotoxic therapy for valvular lesions, due to scarcity of clinical evidence.[8, 9] Some valvular abnormalities respond to anticoagulation therapy. Anticoagulation therapy is similar in patients without valvular lesions in the presence of APLS. Surgical approach with valve replacement or valve repair is considered in patients with severe valvular dysfunction.[10]

As we have shown in our case once a young patient coming with cryptogenic stroke, an underlying 'silent' autoimmune disorder is a likely possibility. Once autoimmune conditions like lupus and APLS is diagnosed, coexistent valvular heart disease like LSE should be excluded ideally with trans-oesophageal echocardiogram. While addressing the risk factors, it is paramount to identify cardiac manifestations and start on timely management as it will improve the patient's quality of life.

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