

Sudden Cardiac Death in Systemic Lupus Erythematosus: A Case Report

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Abstract

Background: Systemic lupus erythematosus (SLE) is a chronic inflammatory, autoimmune disorder, which causes multi-organ damage. It is predominantly seen in the 15 to 40 years of age group. SLE leads to comparatively higher complications in young individuals than the elderly, particularly of a young female of childbearing age group. The Cardiovascular system is most commonly affected; thus, cardiovascular disease is now the third leading cause of death in SLE patients, behind renal disease and infection. The literature on autopsy findings in SLE is few. This case report highlights various autopsy findings in SLE and the importance of cardiac pathology findings in sudden death cases of SLE. **Case presentation:** A 30 years old female presented with an alleged history of fall from a motor cycle due to giddiness while riding as a pillion rider. She got hospitalized and a known diabetes mellitus and hypertension case for four years and was diagnosed with SLE for one year. On admission, her blood glucose was 528 mg/dL; potassium was 6.8 mEq/L and sodium 148mEq/L; later, she expired one day after the incident. The heart weighed 345 grams and left ventricular thickness measuring 2.8 cm. Further, the heart on histopathological examination revealed left ventricular hypertrophy, left anterior descending artery showed 60% narrowing of the lumen, right coronary artery showed 50% narrowing of the lumen, left circumflex coronary artery showed 50% narrowing of the lumen. **Conclusion:** Coronary artery disease accounts for 30% of the overall deaths of SLE patients. A recent study shows that compared to post-menopausal women, the risk of myocardial infarction is 50-fold high in pre-menopausal women with SLE. Henceforth sudden death in SLE patients should be focused on cardiac pathology, and also, this case highlights the importance of meticulous autopsy.

Key words: Systemic lupus erythematosus, Cardiovascular disease, Sudden death, Myocardial infarction, Left anterior descending artery

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Introduction:

Systemic lupus erythematosus (SLE) is an autoimmune disorder, which causes significant organ damage, the etiology of

which is still unclear. Though it affects all age groups, it is predominant in the age group 15 to 40 years.¹ The incidence is 1 to 10 cases in the 100000 population. SLE can manifest as myocardial infarction and is the important cause of sudden cardiac death in the young age group. Compared to post-menopausal women, the risk of myocardial infarction is 50-fold high in pre-menopausal women with SLE. Coronary artery disease accounts for 30% of the overall deaths of SLE patients.² It is associated with numerous systemic manifestations like peritonitis, pancreatitis, lupus pneumonitis,

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etc. At the same time, other cardiovascular manifestations include pericarditis, endocarditis, myocarditis, and myocardial infarction. In the respiratory system, pleuritis can be seen in 60% of SLE patients. Only in 13% of SLE patients, lung parenchyma is affected in the form of interstitial lung disease and lupus pneumonitis.³ SLE patients with lupus nephritis have a higher chance of developing CVD and myocardial infarction.⁴ Systemic lupus erythematosus leads to comparatively higher complications in young individuals than elderly, particularly of the young female gender of childbearing age group.⁵ Cardiovascular diseases started increasing amongst SLE patients.⁶ The cause of sudden or unexpected death in 5-10% of SLE patients is myocardial infarction and arrhythmias followed by cerebrovascular accidents, epilepsy, and pulmonary hypertension.^{1,7} There is sparse literature available on the autopsy findings in SLE patients. Hence this article will highlight the importance of autopsy findings.

Case Report:

A thirty-year-old female presented with an alleged history of fall from a motor cycle, as a pillion rider, due to giddiness. She was a known case of type-II diabetes mellitus and hypertension for the past four years and was diagnosed with SLE for one year. On admission, her blood glucose was 528 mg/dL; potassium was 6.8 mEq/L and sodium 148mEq/L. The blood pressure and pulse were 220/120 mm/hg and 150/min, respectively. The Glasgow Coma Scale (GCS) score was E1V2M4, and the patient expired within 11 hours of admission. Height and weight of the individual was 156 cm and 55 kgs respectively. During the post-mortem examination, contusion and abrasions of varying size and shape were noted, consistent with fall from the bike in motion. On internal examination, the heart weighed 345 g with a left ventricular thickness was 2.8 cm (Fig: 1). The histopathological examination of the

coronary vessels revealed that the left anterior descending artery shows 60% luminal narrowing (Fig: 2). While the right coronary artery, left circumflex artery (Fig: 3), and left coronary artery (Fig: 4) showed 50%, 50%, and 10% luminal narrowing, respectively. Thus, significant lumen narrowing was seen in three important vessels. The Brain showed thin film of patchy sub-arachnoid hemorrhage, and the lungs showed patchy consolidation. The liver showed centrilobular necrosis with steatosis. The kidneys showed 20% sclerosed glomeruli, some glomeruli showed fibrin caps, and some showing wire loop lesions (Fig: 5), and large renal blood vessels showed mild circumferential hyaline arteriosclerosis. The cause of death, in this case, was acute coronary insufficiency and its complication in a case of SLE.

Figure 1: Cut section of heart showing left ventricular thickening

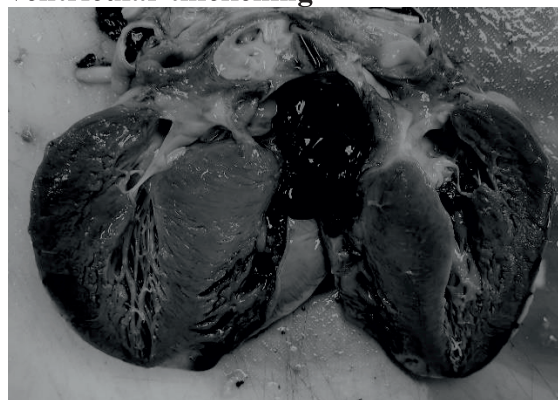


Figure 2: Left anterior descending artery showing 60% narrowing of the lumen. (H&E Stain,4X)

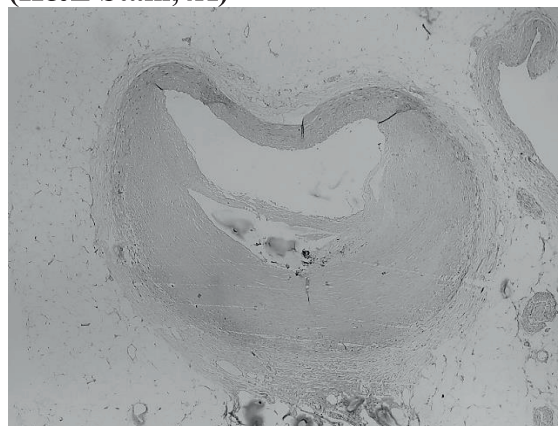


Figure 3: Left circumflex artery showing 50% narrowing of the lumen. (H&E Stain, 4X)



Figure 4: Left coronary artery showing 50% narrowing of the lumen. (H&E Stain, 10X)

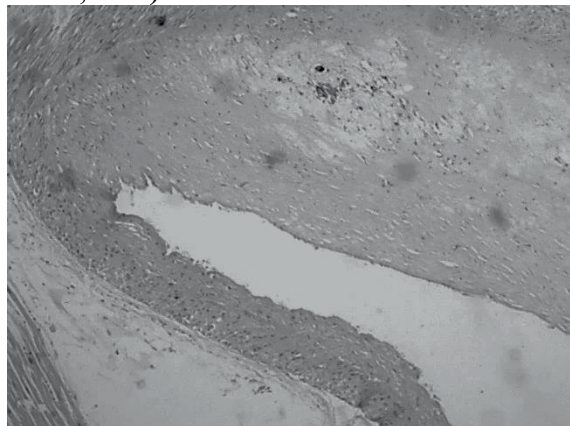
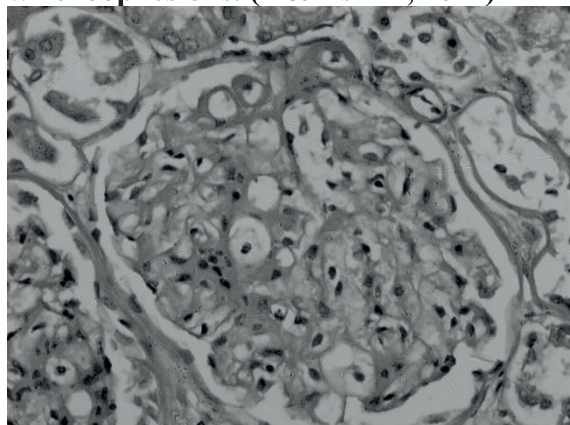


Figure 5: Glomeruli showing occasional wire loop lesions. (H&E stain, 10 X)



Discussion:

Cardiac complications play a significant role in the mortality rate among young age SLE

patients. In SLE patients, the third leading cause of death is cardiovascular disease. Pericarditis is a common cardiac manifestation in SLE patients. Around 11 to 54% and 3 to 9% of SLE patients had pericardial effusion and myocarditis, respectively, during the course of the disease. Recent studies show that 30% of death in SLE patients is due to atherosclerotic heart disease. Compared with post-menopausal women, there is a 50-fold increased risk of myocardial infarction in pre-menopausal women with SLE. The pathomechanism related to accelerated atherosclerosis is still not clear. Still, risk factors like arterial hypertension, diabetes, obesity, smoking, dyslipidemia, metabolic syndrome, and homocysteine, along with long-term medications for SLE treatment, have been shown to increase the occurrence of accelerated atherosclerosis. A recent study from Sweden showed that within one year after diagnosis or two years before diagnosis, the risk for MI in SLE patients increased by 5-fold. In atherosclerosis, the first step to happen is endothelial dysfunction, and its rate increases in the initial five years of diagnosis of SLE. This endothelial damage leads to the release of vascular cell adhesion molecules (VCAM), which helps to regulate inflammation-associated vascular adhesion and trans-endothelial migration of leukocytes like macrophages and T lymphocytes, and it gets accumulated in the walls of the arterial lumen, which then secretes pro-inflammatory cytokines and thus leading to the formation of atherosclerotic plaques.^{2,4,8} Urowitz MB et al. included 1848 patients in his cohort study and found the mean age at diagnosis of SLE to be 34.7 ± 13.3 years in 1640 females. Among 1848 patients, 31 had MI, of which patients who had myocardial infarction before SLE diagnosis or in the first two years diagnosis were 23, of those 14 were female. The mean age for experiencing MI was 49.3 ± 13.4 years.⁹ Panchal L et al. conducted an autopsy study on 27 SLE patients; of those, ten (37.03%)

had myocarditis, nine (33.3%) had endocarditis, seven (25.92%) had pericarditis and myocardial scarring. Only one patient (3.70%) had 90% atherosclerotic narrowing of the left anterior descending coronary artery. Left ventricular hypertrophy was seen in 4 patients. In renal involvement, 19 (70.37%) had lupus nephritis, and 5 (18.52%) had end-stage renal disease, and 2 (7.41%) had acute and chronic pyelonephritis. Pulmonary involvement was seen in 17 patients, with bronchopneumonia in 7 (25.92%) patients, 5 (18.52%) had interstitial lung disease, 3 (11.11%) had an intra-alveolar hemorrhage, and 2 (7.41%) patients had tuberculosis.¹⁰ De Matos Soeiro A et al. conducted a retrospective study on 11 patients, 10 were females, and their median age was 47 years. 64% of cases presented with acute myocardial infarction as a prominent feature. The Most commonly affected coronary artery in acute myocardial infarction is the anterior descending coronary artery, seen in 73% of cases, and the right coronary artery in 45% of cases. Korkmaz C et al. conducted a review of SLE literature during a period from 1975 to 2006 and found that 50 SLE patients had myocardial infarction before 35 years of age; of those, 41 were female, and 9 were male, and their mean age to suffer MI was 24 ± 6.4 years.^{5,11}

This case highlights the various autopsy findings in the SLE case and the importance of cardiac and renal pathology in such cases. As seen in some literature, sudden death in SLE can occur in young females of the childbearing age group, which was similar to this case as well.

Conclusion:

Coronary artery disease accounts for 30% of the overall deaths of SLE patients. A recent study shows that compared to post-menopausal women, the risk of myocardial infarction is 50-fold high in pre-menopausal women with SLE. Henceforth sudden death in SLE patients should be focused on cardiac

pathology, and also, this case highlights the importance of meticulous autopsy.

Conflict of Interest: None

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