Non-coronary cardiac manifestations of systemic lupus erythematosus in adults: a comparative study

Sameh Sayhi1,6, Nour Gueddich1, Rym Dhahri1, Najeh Bousetta1, Bilel Arfaoui2, Nadia Ben Abdelhafid2, Faïda Ajili2, Bassem Louzir1

1Autoimmune Disease Research Unit (Ur17dn02), Internal Medicine Department, Military Hospital of Tunis, Tunis, Tunisia

6Corresponding author: Sameh Sayhi, Autoimmune Disease Research Unit (Ur17dn02), Internal Medicine Department, Military Hospital of Tunis, Tunis, Tunisia

Key words: Systemic lupus erythematosus, cardiac manifestations, pericarditis

Received: 18/03/2019 - Accepted: 23/06/2019 - Published: 02/07/2019

Abstract
Cardiac manifestations develop in the majority of patients with systemic lupus erythematosus (SLE) at some time during the course of their disease. This study was designed to assess cardiac abnormalities in patients with SLE by echocardiography and to compare the 2 groups of patients with and without cardiac manifestations. It was a transversal, descriptive study, conducted in the Internal Medicine Department at the Military Hospital of Tunis from January 2016 to June 2018. Eighty lupus patients, diagnosed on the basis of ACR (American college of rheumatology) criteria, were enrolled in the study and were evaluated by standard echocardiography with color Doppler. Out of 80 patients 42 (52%) had abnormal echocardiographic findings. Pericardial effusion was found in 55%, valvular abnormalities in 52% and 38% had pulmonary hypertension. Patients with pleural effusion (45 vs 15%) were more vulnerable to cardiac involvement as well as renal impairment (57 vs 44%). The difference, however, were not statistically significant (p>0.05) in the renal involvement. Active disease with low complement (80%) was associated with higher frequency of cardiac involvement than disease in remission (64%) but the result was not statistically significant (p=0.11). Cardiac abnormalities are very common in lupus patients even when clinically asymptomatic form. Echocardiography is an excellent non-invasive tool for cardiac evaluation. Their research must be systematic with echocardiography in order to reduce subsequent cardiac morbidity and mortality among the lupus patients.

The Pan African Medical Journal. 2019;33:156. doi:10.11604/pamj.2019.33.156.18697

This article is available online at: http://www.panafrican-med-journal.com/content/article/33/156/full/

© Sameh Sayhi et al. The Pan African Medical Journal - ISSN 1937-8688. This is an Open Access article distributed under the terms of the Creative Commons Attribution License (http://creativecommons.org/licenses/by/2.0), which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.
Introduction

Systemic lupus erythematosus (SLE) is a common chronic multi-system autoimmune disorder of unknown etiology causing injury to many organ systems. It predominantly affects young women. SLE affects 40-200 per 100 000 persons [1], with the higher values seen in black populations. Cardiac manifestations develop in the majority of patients with SLE at some time during the course of their disease [1]. Cardiac involvements in SLE are: valvular disease, most often mitral regurgitation and usually hemodynamically insignificant; pericardial disease, usually an asymptomatic effusion; myocardial dysfunction and coronary artery disease [2]. Current diagnostic methods in particular echocardiography allows early diagnosis of non-coronary cardiac manifestations of systemic lupus erythematosus.

Methods

The aim of our study was to assess the cardiac abnormalities in SLE patients, the contribution of echocardiography in this disease and to compare the 2 groups of patients with and without non-coronary cardiac manifestations. We have performed a transversal, descriptive study of SLE patients hospitalized in the Internal Medicine Department at the Military Hospital of Tunis between January 2016 and June 2018. Diagnosis of SLE was made according to the criteria of the American college of rheumatology (ACR). All patients underwent a cardiac ultrasound externally or during their stay in our department.

Results

The patients were 61 females and 19 males (sex-ratio=3) with a mean age of 38 years. We divided the workforce into two groups according to the presence or absence of cardiac manifestations: group 1 (patients with cardiac disease) and group 2 (patients without cardiac disease. The clinical features of the patients of group 1 are summarized in Table 1. Forty-two patients had cardiac involvement (Group 1). They were 33 females and 9 males with a mean age of the disease of 31.8 years (16-80 years) at the beginning of the disease and 41 years at the time of the study. In group 1, 83% were symptomatic. The symptoms were dominated by objectified chest pain (43%). In Doppler echocardiography, pericarditis was found in 23 patients (55%) with a single case of cardiac tamponade. Libman Saks endocarditis and lupus myocarditis were found in one case each. Pulmonary hypertension (HTP) was observed in 16 patients (38%) and valvular disease in 22 patients (52%). Cardiomegaly was observed in 9 patients (21%). Electrical abnormalities were dominated by micro-voltage found in 8 patients. The general symptoms (83%), skin lesions (76%) and musculoskeletal involvement (64%) were the most frequent events associated with the cardiac manifestations in group 1 (Table 1). ANA were positive in 97% of cases and antiphospholipid antibodies in 24%. Prednisone: 1mg/kg/day and immunosuppressive therapy were indicated respectively in 71% and 38% of patients of group 1. The comparative study of the two groups showed that patients with pleural effusion (45 vs 15%) were more vulnerable to cardiac involvement (P=0.004) as well as renal impairment (57 vs 44%) but the difference, however, was not statistically significant here (p>0.05). Rnp antibodies was higher in group 1 and statistically significant (p<0.05). Some antibodies, and antiphospholipid antibodies were higher in group 1 but the difference was not statistically significant here (p>0.05). Active disease with low complement (80%) was associated with higher frequency of cardiac involvement than disease in remission (62%) but the result was not statistically significant (p=0.07) (Table 2).

Discussion

Cardiovascular abnormalities are common in patients with systemic lupus erythematosus [3]. SLE is among systemic diseases most providers of heart disease [4, 5]. It probably develops in nearly all patients suffering from SLE at some time during the course of their disease. All components of the heart can be affected and venous thrombo-embolic complications may cause pulmonary hypertension and right heart failure [6]. Cardiac disease is observed in 30 to 62% by clinical, ultrasound or autopsy diagnostic tool [7, 8]. Its prevalence was 52% in our study, it was 70% in the French study of Badoui et al [9] and in 68% in the Polish study of Ostanek et al [10]. Pericardial involvement is the most common echocardiographic lesion in systemic lupus erythematosus (SLE) and is the most frequent cause of symptomatic cardiac disease. It is observed in 11 to 54% of the cases [7]. In our study, the frequency of pericarditis was 55% higher than those described in the recent studies. Tamponade occurs in less than 2% and constrictive pericarditis is extremely rare [1, 2]. Cardiac
involvement may precede the clinical signs of lupus and it is usually asymptomatic. It is generally diagnosed by echocardiography performed for some other reason, such as suggestive electrocardiographic abnormalities. Pericarditis, as with other types of serositis, most often occurs when SLE is active in other organs as well. In our experience, patients with pleural effusion (45 vs 15%) were more vulnerable to cardiac involvement (P=0.004). Cardiomyopathy is uncommon clinically but autopsy studies found myocardial involvement in 40-50% of patients [1]. Valvular disease is often underdiagnosed because most of the time they are asymptomatic. Pulmonary arterial hypertension (PAH) is a rare complication of systemic lupus erythematosus (SLE) [9]. Its prevalence depends on the method with which it is diagnosed and the tools used for that (echocardiography or heart catheterism). This prevalence varies from 4.2 to 17.5% in the literature when diagnosed by echocardiography and from 0.5 to 9.3% using the right heart catheterism. In our patients, where PAH was defined by a systolic pulmonary arterial hypertension ≥ 30 mmHg in echocardiography, its prevalence was of 38% higher than those of the literature.

**Conclusion**

Cardiac abnormalities are very common in lupus patients even when clinically asymptomatic. Echocardiography is an excellent non-invasive tool for cardiac evaluation. Their research must be systematic with echocardiography in order to reduce subsequent cardiac morbidity and mortality among the lupus patients.

**What is known about this topic**

- Cardiovascular abnormalities are common in patients with systemic lupus erythematosus;
- Cardiomyopathy is uncommon clinically but autopsy studies found myocardial involvement.

**What this study adds**

- This study was designed to assess cardiac abnormalities in patients with SLE by echocardiography and to compare the 2 groups of patients with and without cardiac manifestations;
- Echocardiography is an excellent non-invasive tool for cardiac evaluation even when clinically asymptomatic.

**Competing interests**

The authors declare no competing interests.

**Authors’ contributions**

All the authors have read and agreed to the final manuscript.

**Tables**

**Table 1**: clinical features of patients with cardiac manifestations

**Table 2**: comparison of patients with and without cardiac manifestations

**References**

1. Jain D, Halushka MK. Cardiac pathology of systemic lupus erythematosus. J Clin Pathol. 2009; 62(7): 584-92. [PubMed](https://pubmed.ncbi.nlm.nih.gov/19763429/) | [Google Scholar](https://www.ncbi.nlm.nih.gov/pubmed/19763429)

2. Brigden W, Bywaters EG, Less MH. The heart in systemic lupus erythematosus. Br Heart J. 1960; 22: 1-16. [PubMed](https://pubmed.ncbi.nlm.nih.gov/14168278/) | [Google Scholar](https://www.ncbi.nlm.nih.gov/pubmed/14168278)

3. Ong ML, Veerapen K, Chambers JB, Lim MN, Manivasagar M, Wang F. Cardiac abnormalities in systemic lupus erythematosus: prevalence and relationship to disease activity. International Journal of Cardiology. 1992; 34(1): 69-74. [PubMed](https://pubmed.ncbi.nlm.nih.gov/2492983/) | [Google Scholar](https://www.ncbi.nlm.nih.gov/pubmed/2492983)

4. Ruiz-Irastorza G, Khamashta MA, Castellino G, Hughes GRV. Systemic lupus erythematosus. Lancet. 2001; 357(9261): 1027-32. [PubMed](https://pubmed.ncbi.nlm.nih.gov/11398250/)

5. Schattner A, Liang MH. The cardiovascular burden of lupus: a complex challenge. Arch Intern Med. 2003; 163(13): 1507-10. [PubMed](https://pubmed.ncbi.nlm.nih.gov/14580382/) | [Google Scholar](https://www.ncbi.nlm.nih.gov/pubmed/14580382)
6. Oliveira GH, Seward JB, Tsang TS, Specks U. Echocardiographic findings in patients with Wegener granulomatosis. Mayo Clin Proc. 2005; 80(11): 1435-40. PubMed | Google Scholar

7. Smiti M, Ben Salem T, Larbi T, Braham Sfaxi A, Ben Ghorbel I, Lamloum M et al. Péridardites lupiques: prévalence, caractéristiques cliniques et immunologiques. Presse Med. 2009; 38(3): 362-5. Google Scholar

8. Louzir B, Othmani S, Ben Abdelhafidh N. Le lupus érythémateux systémique en Tunisie, Etude multicentrique nationale: a propos de 295 observations. Rev Med interne. 2003; 24(12): 768-74. Google Scholar

9. Badui E, Garcia-Rubi D, Robles E, Jimenez J, Juan L, Deleze M et al. Cardiovascular manifestations in systemic lupus erythematosus: prospective study of 100 patients. Angiology. 1985; 36(7): 431-41. PubMed | Google Scholar

10. Ostanek L, Płonska E, Peregud-Pogorzelska M, Mokrzycki K, Brzosko M, Fischer K et al. Cardiovascular abnormalities in systemic lupus erythematosus patients in echocardiographic assessment. Pol Merkur Lekarski. 2006; 20(117): 305-8. PubMed | Google Scholar

---

**Table 1**: clinical features of patients with cardiac manifestations

| Clinical features               | Patients (n=42) | n (%) |
|---------------------------------|----------------|-------|
| general signs                   |                |       |
| Reached mucocutaneous           |                |       |
| Photosensitivity                | 16 (38%)       |       |
| Nasal ulcerations               | 2 (5%)         |       |
| malar rash                      | 13 (31%)       |       |
| joint damage                    |                |       |
| arthralgia                      | 19 (45%)       |       |
| arthritis                       | 8 (19%)        |       |
| Deformation hand Jaccoud        | 1 (2%)         |       |
| renal impairment                |                |       |
| Glomerular nephropathy          | 18 (43%)       |       |
| Renal Insufficiency             | 10 (24%)       |       |
| Serositis                       |                |       |
| pleurisy                        | 13 (31%)       |       |
| ascites                         | 5 (12%)        |       |
| lung disease                    |                |       |
| diffuse infiltrative lung disease | 5 (12%)   |       |
| achieved hematologic            |                |       |
| neutropenia                     | 5 (12%)        |       |
| thrombopenia                    | 5 (12%)        |       |
| Heamolytic anemia               | 6 (14%)        |       |
| neurological impairment         |                |       |
| reached parenchymal             | 6 (14%)        |       |
| events                          |                |       |
| thromboembolic                  |                |       |
| Thrombosis of the lower limbs   | 2 (5%)         |       |
| pulmonary embolism              | 2 (5%)         |       |
| lupus myositis                  | 1 (2,3%)       |       |
| hepatitis lupus                 | 1 (2,3%)       |       |

*
|                          | Group 1 n (42) | Group 2 n (38) | P   |
|--------------------------|---------------|----------------|-----|
| Sex (female)             | 34            | 32             | 0.70|
| General signs            | 35            | 29             | 0.43|
| Skin involvement         | 33            | 29             | 0.81|
| Musculoskeletal involvement | 27         | 24             | 0.91|
| Achieved hematologic     | 27            | 25             | 0.88|
| Serositis: pleural effusion | 19          | 6              | 0.004|
| Renal impairment         | 24            | 17             | 0.26|
| Neurological involvement | 6             | 6              | 0.85|
| Ac anti DNA natifs       | 29            | 26             | 0.95|
| Low complement           | 34            | 24             | 0.07|
| Ac anti Sm               | 9             | 4              | 0.18|
| Ac anti Rnp              | 8             | 1              | 0.03|
| Ac antiphospholipid      | 10            | 4              | 0.11|
| Corticosteroids 1mg/kg/j | 30            | 28             | 0.82|
| Cyclophosphamide         | 16            | 15             | 0.89|