Journal of Case Reports in Practice (JCRP) 2018; 6(2): 23-28

ORIGINAL ARTICLE

Common causes of emergency visits in patients with systemic lupus erythematosus: a study focusing on the role of SLE-induced vasculitis

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ABSTRACT

Backgrounds: Systematic lupus erythematosus (SLE) is a chronic and autoimmune disease that involvement of several target organs, at emergency department (ER) it is important to consider SLE as a cause for acute organ involvement. The aims of this study were to determine the cause for emergency visits in SLE Patients, focuses of Vasculitis.

Methods: Eighty one known SLE patients based on "1982 Revised American College Of Rheumatology SLE Criteria "who had visited the emergency department of Imam Reza hospital from June 2012-December 2014 having undergone a thorough physical examination by the emergency service and classified based on demographic and SLE related sign and symptom. We focused on vasculitis and non-vacuities groups.

Results: In total 17.3% were diagnosed as vasculitis. In the vasculitis group cutaneous vasculitis (50%), thromoembolic events (7.1%) and cerebral vasculitis (7.1%) retinal vasculitis (7.1%) and aortic involvement was seen. the first three most common causes for ER visit in SLE patients were 1) SLE flare episode along with polyarthritis, mucosal ulcer and photosensivity. 2) Vasculitis 3) Renal involvement. Mean SLEDAI and SLICC scores were 22.79 (±13.29) and 1.95 (±1.46), respectively. Regarding SLICC parameters and their relevance to vasculitis ocular, renal, gastrointestinal and neuropsychiatric damage and diabetes mellitus were the most prevalent complications among the vasculitis group (p-value = 0.03, 0.01,<0.001, <0.03, <0.003, respectively).

Conclusion: Emergency medicine doctors have to consider autoimmune disease among the top of their differential diagnoses list. The majority of these patients, especially in developing countries, may be seen in ER with no previous diagnosis of lupus and they may be presented with severe organ damage as initial presentation.

Key words: systemic lupus nephritis, vasculitis, emergency visit

INTRODUCTION

Systemic lupus erythematosus (SLE), as a chronic autoimmune disease, is associated with involvement of several target organs. The onset of severe complications of SLE which has been observed relatively earlier in developing countries, most probably is due to poor medication adherence and irregular clinical follow ups.[1]

As a result of such acute manifestations and major complications during the disease course, SLE patients may require more emergency visits at emergency room (ER). Previous studies have indicated that patients with older age and a lower socioeconomic status have been more likely visited at ER whenever necessary and additionally they intend to have more scheduled outpatient visits with their rheumatologist or primary care physician.[2-3]

Therefore, for an emergency medicine specialist, it is important to consider SLE as a possibility when encountering a patient with symptoms and/or signs suggestive of multisystem organ involvement. In order to better prevention from the urgent and serious complications of SLE such as stroke, acute myocardial infarction, hemoptysis, respiratory distress and pulmonary emboli, early management and close observation of these high risk patients should be taken into consideration.[4-5]

Distinguishing an acute SLE flare episode from sep-

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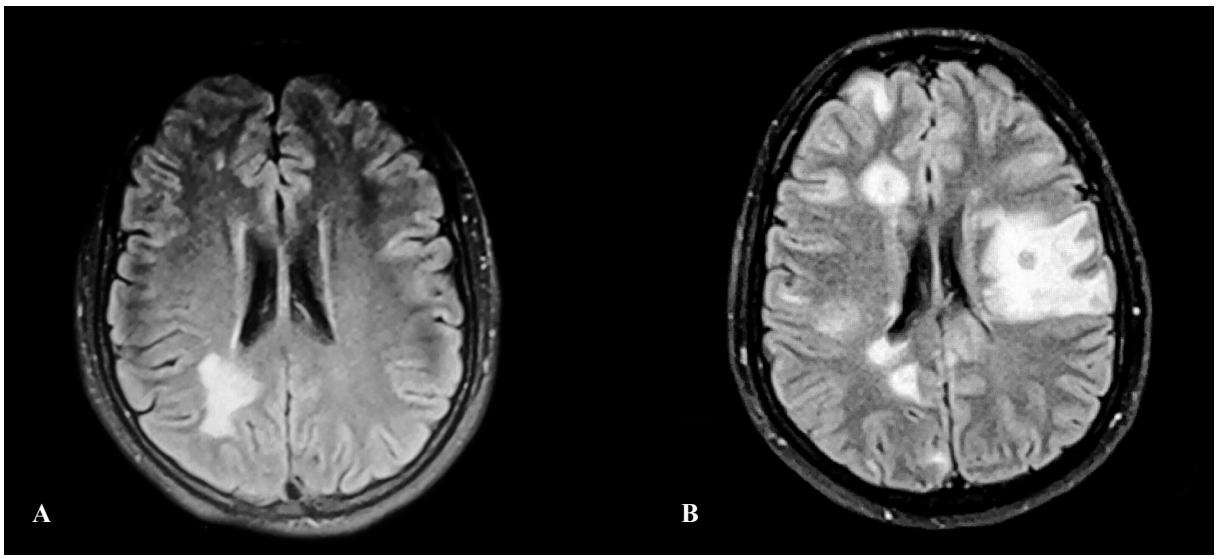

Figure 1, Axial inversion recovery sequence MRI scan of the brain demonstrates numerous confluent hyperintense lesions in the deep as well as subcortical white matter with discrete involvement of the cortex, compatible with vasculitis or viral infection. **A)** An MRI scan of 19-years old girl, newly diagnosed with lupus and **B)** shows a 60-year-old man who had developed acute extensive myocardial infarction.

sis, in the emergency setting can be the most challenging issue in rheumatology. Nevertheless, acute life-threatening and multisystem organ involvement in the ER should raise suspicion for any autoimmune diseases, particularly SLE.[1]

The aims of this study were to determine the causes for emergency visits and the outcomes of lupus patients who had been visited the ER of Imam Reza Hospital in Mashhad, Iran; in a retrospective evaluation; and also to identify the variety factors leading to hospitalization in such immune compromised patients.

MATERIALS AND METHODS

Eighty one known cases of SLE patients based on 1982 Revised American College Of Rheumatology SLE Criteria, who had visited the emergency department of Imam Reza hospital from June 2012-December 2014, underwent a thorough physical examination by the emergency medicine resident under supervision of two well-experienced rheumatologist, were enrolled.

The demographic data including age, gender, disease duration, the reason for referring to Emergency Department (ER), initial sign and symptoms at ER and SLEDI-2K and SLICC Scores were collected in a specifically-designed questionnaire. Inclusion Criteria comprise Lupus patients with or without vasculitis. Therefore we recruited and evaluated SLE patients based on 1982 Revised American College Of Rheumatology SLE Criteria. Exclusion criteria consist of all non Lupus Patients with other causes of vasculitis like as infectious vasculitis, idiopathic autoimmune vasculitis, polyarteritis nodosa, neoplasia associated vasculitis and other vasculitis rather than SLE-related ones were excluded from the study. Definite diagnosis and Imression of vasculitis confirmed based on clinical vasculitic manifestation like as skin vasculitis, laboratory finding consist of P/C ANCA antibodies and tissue biopsy .

The most common emergency presentations with respect to organ involvement included:

1) Constitutional symptoms followed by;
2) gastrointestinal 3) skin 4) cardiovascular
5) CNS 6) renal and 7) hematological symptoms.

Lupus patients were categorized into two groups with or without vasculitis.

Statistical analyses

For data analyses T-test, Wilcoxon's test, Chi-square test, Spearman's correlation test, Man-Whitney and Kruskal-Wallis tests were used. SPSS version 16 in statistical analysis was also applied. A $p<0.05$ was considered as statistically significant.

RESULTS

In total, eighty one lupus patients were considered eligible for this study. 73 (90.9%) cases were female and 8 (9.9%) were male. Their mean age was 28.7±11.2 years and the mean disease duration was 2.9± 3.3 years. The social and educational backgrounds of our patients were fully evaluated and divided into three groups (high school, undergraduate, postgraduate). The first three most common causes for ER visit in SLE patients were:

1. SLE flare episode along with polyarthritis, mucosal ulcer and photosensivity.
2. Vasculitis/ Vasculitis- like lesions such as palpable purpura, rush, venous thrombosis, cerebrovascu-

lar accidents, retinal vasculitis, mesenteric ischemia. 3. Renal involvement such as proteinuria, pyuria, and hematuria.

In total, fourteen lupus patients out of eighty one (17.3%) were diagnosed as vasculitis whereas the rest of patients did not have any vascular involvement. In the vasculitis group, consisting of one male (7/1%) and 13 (92.9%) females, seven patients had cutaneous vasculitis while seven others suffered from thromboembolic events. Five individuals had cerebral vasculitis, one had retinal vasculitis and one other patient had abdominal aortic vessels involvement and mesenteric insufficiency. Two patients were admitted in very critical condition and passed away during the time of hospitalization; a 19-years old girl, newly diagnosed lupus case who had been referred to the ER with status epilepticus (due to CNS lupus vasculitis) as her MRI show in Figure 1 (A), and a 60-year-old man who had developed acute extensive myocardial infarction after 5 days of hospitalization which is shown in Figure 1 (B).

The prevalence of the most common clinical findings which have been demonstrated in table 1 were as follows: kidney involvement (hematuria: 74%, proteinuria: 80.2%), arthritis (70.4%), anemia (63%), rash (56.6%), photosensitivity (49.4%), alopecia (35.8%) and mucosal ulcers (27.2%).

Interestingly, the current study showed a significant higher prevalence of organic brain syndrome, retinal vasculitis, psychosis, pericarditis and malar rash in lupus patients with vasculitis compared to non-vasculitis cases ($p<0.001$). Mean SLEDAI and SLICC scores were 22.79 (±13.29) and 1.95 (±1.46), respectively. Regarding SLICC parameters and their relevance to vasculitis (Table 1), ocular, renal, gastrointestinal and neuropsychiatric damage and diabetes mellitus were the most prevalent complications among the vasculitis group ($p=0.03$, 0.01, <0.001, <0.03, <0.003, respectively). However, disease duration and level of anti-ds DNA did not indicate a statistically significant difference between lupus patients with and without vasculitis ($p=0.64$, 0.89, respectively).

DISCUSSION

Systemic lupus erythematosus is an important autoimmune disorder characterized by multiple organ damages. It presents with a variety of clinical manifestations which are not easy to distinguish initially as SLE, particularly in the emergency department.

Certainly, there is a high probability for a SLE patient comes to ER during his/her life-time due to acute or severe clinical manifestations either SLE-related/SLE-unrelated or SLE flare.[6-7] Complications resulting from target organs' involvement are not uncommon. The possibility of missing in diagnosis in such patients in not uncommon; as on-call emergency medicine doctors may not consider connective tissue disorders e.g. vasculitis and SLE at the top of their differential diagnosis list.[3-13]

In developing countries like Iran, SLE patients are usually not diagnosed in the early stages and do not have a regular follow up due to poor socioeconomic conditions. Hence, the risk of infection, disease-related complications, disease progression, flare up and deterioration leading to ER visits would probably significant.[1, 6, 8-9] Therefore the vigilance of emergency doctors in early diagnosis of lupus patients seems to be merely essential in everyday practice. It is obvious that the awareness and training of front-line physicians in the ER who are directly involved with the management of emergent conditions is very crucial and contribute strong impact on the prognosis and outcome in such critical patients.[10-11, 14]

In our study, most patients were females in their childbearing age (90.9%). The most frequent cause of hospitalization was lupus flare presenting mostly as arthritis, kidney involvement, constitutional symptoms like fever, anemia and vasculitis (Table 2). As demonstrated in Table 2, different kinds of vasculitis manifestations were followed by kidney involvement (pyuria, hematuria, and proteinuria). In a Chinese studythe most frequent presentations were fever and cardiopulmonary manifestations.[3, 7, 15]

In a study conducted in Portugal, dyspena and tachycardia were most common 2. In the current study with respect to focusing on vasculitis at the ER, in 17.3% of our patients vasculitis manifested as cutaneous vasculitis (50%) whereas in 35.7% central nervous system vasculitis presented as seizure and hemiplegia (Table 2). In 7.1% retinal vasculitis manifested as sudden visual loss whereas in another 7.1% vasculitis with mesenteric insufficiency presented as abdominal pain. In the vasculitis group, prevalence of visual disturbance, organic brain syndrome, psychosis and pericarditis were significantly higher compared to the non-vasculitis group ($p=0.007$, $p=0.001$ and $p=0.000$, respectively) (Table 2).

Moreover, we observed a higher disease activity in the vasculitis group compared to the non-vasculitis group. In another study by Shariati-Sarabi et al. in 2013 on 71 SLE patients, SDI was significantly associated with SLEDAI-2K ($r=0.742$, $p<0.001$) and a higher SDI was obtained with a longer disease duration.[10] They also found a similar frequency (16%) of thrombotic events in a Cohort of 500 lupus patients in 2005.[11] Tomes et al. also studied 168 lupus patients and found venous thrombosis in 16% of the cases.[7] Therefore, it is very important for ER residents to be aware of and recognize the signs and symptoms of vasculitis in autoimmune diseases; this is of greater value in developing countries which do not have a referral system and most of the patients are not visited by their family physician on a regular basis. Patients visiting the ER with signs and symptoms of blurred vision, cutaneous lesions, thrombotic events, recurrent abdominal pain, dyspena and bloody sputum as

an emergent condition should be carefully evaluated for vasculitis and autoimmune diseases after primary and emergent care. Acute conditions in these patients can be a manifestation of vasculitis related to an autoimmune disease such as lupus erythematosus. Similar conditions can be prevented and controlled with immunosuppressive therapy (corticosteroids plus cytotoxic drugs), preventing mismanagement and over workup or invasive treatment approaches such as surgery. Additionally, a correct diagnosis will result in lower morbidity and mortality in such patients, particularly in child bearing women.

CONCLUSION

In ER, emergency medicine doctors have to consider autoimmune disease and vasculitis among the top of their differential diagnoses list particularly in life threatening conditions. The majority of these patients, especially in developing countries, may be seen in ER with no previous diagnosis of lupus and they may be presented with severe organ damage as initial presentation. This awareness will hopefully lead to correct and precise diagnosis. Surely, ER doctors' vigilance may reduce morbidity and mortality of these patients; subsequently, we can come to the final diagnosis with less time-wasting and unnecessary workup in such patients.

CONFLICT OF INTEREST

None.

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Table 1, SLICC parameters and their relevance to vasculitis

COMPLICATION		VASCULITIS		P-value
		Positive	Negative	
Musculoskeletal damage	**Negative**	13(19.3%)	67(83.8%)	**0.17**
	Positive	1(100%)	0(0%)	
Thromboembolic event	**Negative**	13(16.3%)	67(83.3%)	**0.17**
	Positive	1(100%)	0(0%)	
Gastrointestinal damage	**Negative**	7(9.7%)	65(90.3%)	**<0.001**
	Positive	7(77.8%)	2(22.2%)	
Diabetes	**Negative**	10(13.3%)	65(86.7%)	**0.007**
	Positive	4(66.7%)	2(33.3%)	
Neuropsychiatric damage	**Negative**	11(14.5%)	65(85.5%)	**0.56**
	Positive	3(60%)	2(40%)	
Ocular damage	**Negative**	10(13.5%)	63(87.5%)	**0.06**
	Positive	4(57.1%)	4(44.4%)	
Neuropsychiatric damage	**Negative**	10(13.5%)	68(86.5%)	**0.58**
	Positive	4(57.1%)	3(42.9%)	
Peripheral vascular damage	**Negative**	9(12.5%)	63(87.5%)	**0.06**
	Positive	5(55.6%)	4(44.4%)	
Renal damage	**Negative**	6(9.7%)	56(90.3%)	**0.01**
	Positive	8(42.1%)	11(57.9%)	
Pulmonary damage	**Negative**	9(16.7%)	45(83.3%)	**0.53**
	Positive	5(18.5%)	22(81.5%)	
Skin damage	**Negative**	10(16.7%)	50(83.3%)	**0.52**
	Positive	4(19%)	17(81%)	
Cardiovascular damage	**Negative**	14(17.9%)	64(82.1%)	**0.03**
	Positive	0(0%)	3(100%)	

Table 2, SLEDAI parameters and their relevance to vasculitis

Complication		Vasculitis		P-value
		Positive	Negative	
Visual Disturbance	**Negative**	10(13.3%)	65(86.6%)	**0.007**
	Positive	4(66.7%)	2(33.3%)	
Organic brain syndrome	**Negative**	8(8.3%)	64(91.7%)	**0.001**
	Positive	6(66.7%)	3(33.3%)	
Seizure	**Negative**	12(15.4%)	66(86.4%)	**0.08**
	Positive	2(66.7%)	1(33.3%)	
Pericarditis	**Negative**	10(13.5%)	64(86.5%)	**0.01**
	Positive	4(57.1%)	3(42.9%)	
Psychosis	**Negative**	10(13.5%)	64(86.5%)	**0.01**
	Positive	4(57.1%)	3(42.9%)	
Myositis	**Negative**	11(14.7%)	64(85.3%)	**0.06**
	Positive	3(50%)	3(50%)	
CVA	**Negative**	13(16.5%)	66(83.5%)	**0.32**
	Positive	1(50%)	1(50%)	
Cranial nerve palsy	**Negative**	13(16.7%)	2(33.3%)	**0.007**
	Positive	1(33.3%)	65(83.3%)	
Lupus headache	**Negative**	12(16.4%)	61(83.6%)	**0.42**
	Positive	2(25%)	6(75%)	
Alopecia	**Negative**	7(13.5%)	45(86.5%)	**0.18**
	Positive	7(24.1%)	22(75.9%)	
Pleurisies	**Negative**	12(16.7%)	60(83.3%)	**0.48**
	Positive	2(22.2%)	7(77.8%)	
Urinary cast	**Negative**	10(16.1%)	52(83.9%)	**0.42**
	Positive	4(21.1%)	15(78.9%)	
Proteinuria	**Negative**	1(6.2%)	15(93.8%)	**0.17**
	Positive	13(20%)	52(80%)	
Arthritis	**Negative**	3(12.5%)	21(87.5%)	**0.35**
	Positive	11(19.3%)	46(80.7%)	
Pyuria	**Negative**	6(15.4%)	33(84.6%)	**0.44**
	Positive	8(19%)	34(81%)	
Increased Anti-ds DNA	**Negative**	4(14.3%)	24(85.7%)	**0.76**
	Positive	10(18.9%)	43(81.1%)	
Mucosal ulcer	**Negative**	10(16.9%)	49(83.1%)	**0.56**
	Positive	4(18.2%)	18(81.8%)	
Fever	**Negative**	13(17.6%)	61(82.4%)	**0.65**
	Positive	1(14.3%)	6(85.7%)	
Hematuria	**Negative**	6(28.6%)	15(71.4%)	**0.11**
	Positive	8(13.3%)	52(86.7%)	
Low com.	**Negative**	10(20.8%)	38(79.2%)	**0.23**
	Positive	4(12.1%)	29(87.9%)	
Malar rash	**Negative**	10(27.8%)	26(72.2%)	**0.02**
	Positive	4(8.9%)	41(91.1%)	
Thrombocytopenia	**Negative**	14(19.7)	57(80.3%)	**0.12**
	Positive	0(0%)	10(100%)	
Leukopenia	**Negative**	14(19.4)	58(80.6%)	**0.15**
	Positive	**0(0%)**	**9(100%)**	