Pulseless Systemic Lupus Erythematosus: A Rare Presentation

Abstract

Vascular disease is frequent in patients with systemic lupus erythematosus, which can be related to the disease process, or can develop as an accompanying co-morbidity and represents the most frequent cause of death in established disease. However, at times the presentations can be uncommon and subtle, and warrants a thorough examination both clinically and radiologically.We report a case of a young female with photosensitive malar rash, oral ulcers, intermittent fever with joint pains, history of two abortions, and unilateral absent radial and brachial artery pulses on clinical examination. The evaluation revealed positive antinuclear antibody (4+), anti-Smith antibody (2+), direct Coomb's test (2+), and antiphospholipid antibody panel was negative. Color doppler flow imaging of right upper limb (arterial) revealed irregular wall thickening with a narrow lumen and mildly reduced peak systolic volume. Computed tomography aortogram revealed wall thickening and luminal narrowing involving the entire length of the right brachial and radial artery. We report this case for its rarity and unique presentation of medium vessel vasculopathy.

Keywords: *Absent pulses, lupus erythematosus, medium vessel vasculopathy*

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Introduction

Systemic lupus erythematosus (SLE) is characterized by protean manifestations. It is an autoimmune disease in which organs and cells undergo damage initially mediated by tissue binding autoantibodies and immune complexes. Clinical manifestations are heterogenous, and ninety percent of patients at diagnosis are women of childbearing age.[1] It is characterized by the production of antibodies against a variety of nuclear antigens.

Asymptomatic medium vessel vasculopathy in systemic lupus erythematosus patients is a rare form of vascular involvement.[2] We report a case wherein clinico-radiological evaluation suggested medium vessel vasculopathy involving the right brachial and radial artery.

Case Report

A 23-year-old female, farmer by occupation, married since last seven years with a history of two spontaneous abortions (first in 1st trimester and second in 2nd trimester of pregnancy; no treatment taken and no cause elicited) without any living children, presented with a photosensitive rash over the face, trunk and upper extremities and recurrent painless oral ulceration of four years duration. She also had history of intermittent fever with multiple joint pains with morning stiffness (lasting for an hour) of two years duration.

She also gave a history of diffuse thinning of scalp hair associated with significant hair fall. There was no history suggestive of Raynaud phenomenon, skin thickening, dyspeptic symptoms, muscle weakness, seizures, or internal organ involvement.

General examination revealed pallor, absent pulses on the right side (radial and brachial artery), rest of the pulses were normal with a blood pressure of 100/64 mm Hg (left arm) and 108/68 mm Hg (right arm; maybe falsely more due to increased pressure in the narrowed lumen). The systemic examination was essentially normal.

Dermatological examination revealed lupus hair in the form of diffuse nonscarring alopecia with lustreless, coarse, dry, and rough hair, an erythematous malar rash with few scaly plaques over the cheeks and mandibular region showing atrophic scarring in the center at places [Figure 1]. Involvement of the upper and lower back, extensor surface of upper limbs, V-region

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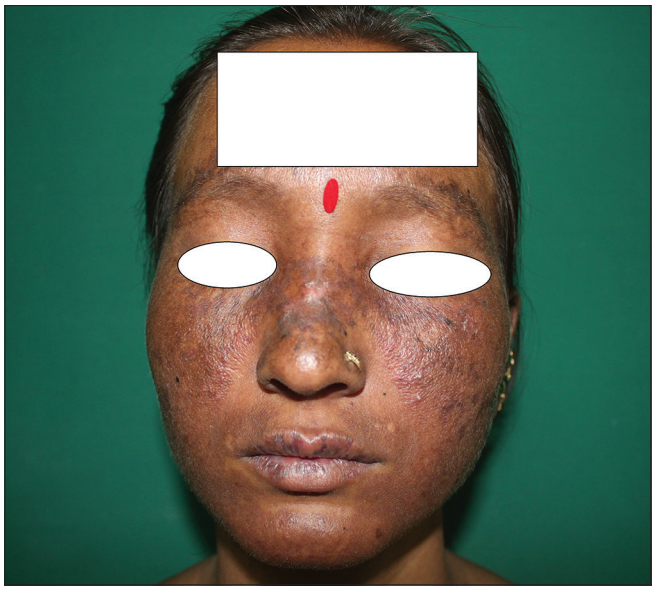

Figure 1: Erythematous to hyperpigmented malar rash over the face with characteristic sparing of the nasolabial folds

of neck in the form of multiple well-defined discrete to confluent hyperpigmented to erythematous scaly plaques showing atrophic scarring in the center. Oral mucosa revealed multiple ulcers over the hard palate [Figure 2]. Positive investigations revealed Hb of 8 gm% (Peripheral Blood Smear: occasional schistocytes, normocytic normochromic anemia), Platelets: 2.89 lacs/cumm, direct Coomb's test was positive (2+), antinuclear antibodies by indirect immunofluorescence was positive (speckled pattern with titers 1:80), anti-Smith antibody was positive (2+), and rest of the antibodies including anti-ds-DNA were negative. Liver function tests, renal function tests, erythrocyte sedimentation rate, C reactive protein, 24-h urinary proteins, 2D-echocardiography, and ultrasound abdomen were within normal limits. Antiphospholipid antibody panel (Lupus anticoagulant, IgG/IgM anticardiolipin antibodies, IgG/IgM ß-2 glycoprotein) was negative, although a high clinical suspicion of antiphospholipid syndrome was kept. Pulmonary function tests (PFT) showed a mild restrictive pattern. High resolution computed tomography chest done to rule out interstitial lung disease because of a restrictive pattern on PFT was within normal limits. Color Doppler flow imaging of bilateral upper limbs (arterial) showed irregular wall thickening with a narrow lumen and mildly reduced peak systolic volume, triphasic flow. Computed tomography (CT) aortogram showed wall thickening and luminal narrowing involving the entire length of the right brachial and radial artery without enhancement of the vessel wall [Figures 3 and 4]. The aorta and the subclavian arteries bilaterally did not show any evidence of mural thickening, luminal narrowing, or features of inflammation in the form of postcontrast mural enhancement.

Based on clinical, laboratory, and radiological findings, she was diagnosed as a case of systemic lupus erythematosus

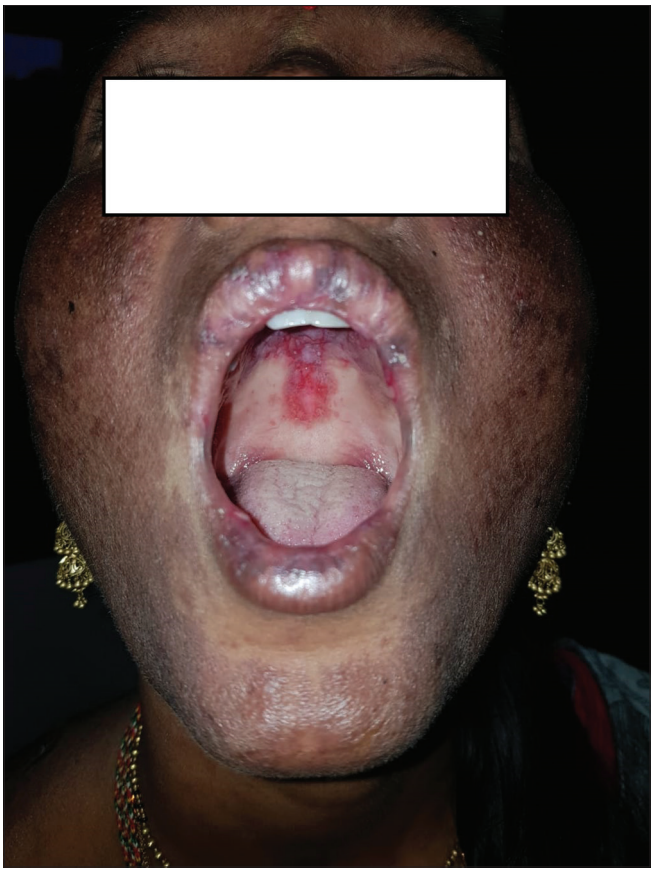

Figure 2: Multiple oral ulcers over the hard palate

with medium vessel vasculopathy as she satisfied the EULAR/ACR (European League against Rheumatism/ American College of Rheumatology) 2019 criteria. The patient refused for radial artery biopsy. She was treated with low dose oral steroids and tab hydroxychloroquine along with sun protection. As there was no evidence of thrombosis on imaging, she was not started on anticoagulants; however, she was counselled for starting the same in her next pregnancy (as she was planning to conceive). She was also started on aspirin and tablet azathioprine. Azathioprine was added because of severe cutaneous lupus not responding to topical steroids and secondly because of the remote possibility of autoimmune hemolytic anemia as direct Coomb's test (DCT) can be positive in SLE without hemolysis. Her skin and oral lesions responded well to treatment, however, the pulses remained absent till the last follow up.

Discussion

Vascular disease is frequent in patients with systemic lupus erythematosus (SLE) and represents the most frequent cause of death in established disease. It can be a part of the disease process or can be an accompanied comorbidity like steroid-related atherosclerotic disease, or represent the synergistic pathogenic outcome of accelerated atherosclerosis.[3]

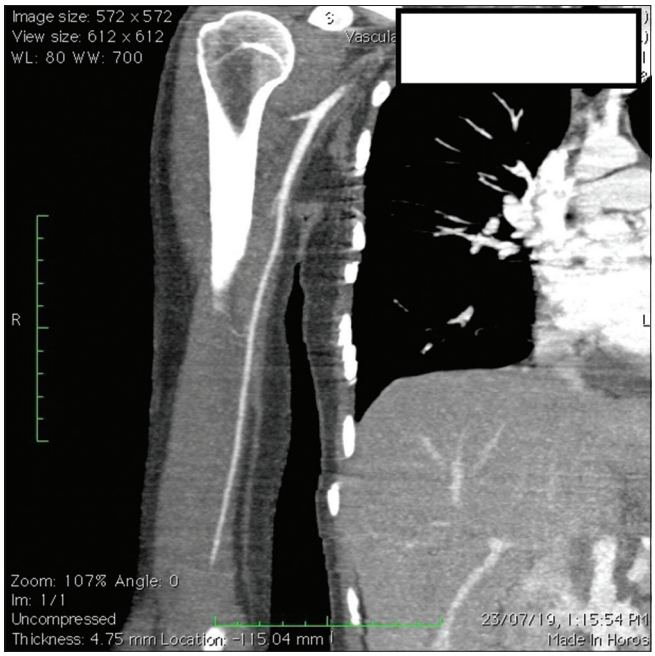

Figure 3: CT aortogram showing wall thickening and luminal narrowing involving the entire length of the right brachial artery without enhancement of the vessel wall

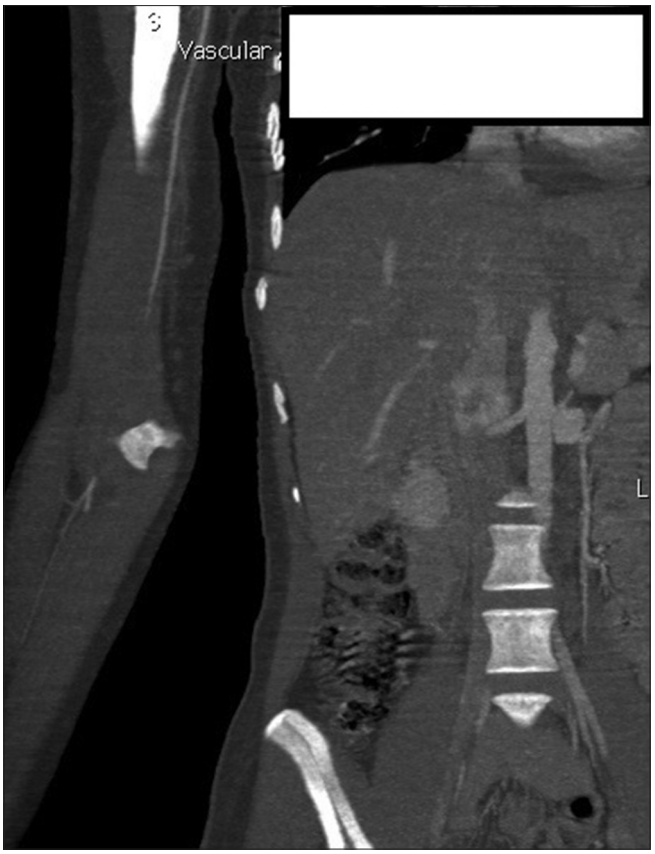

Figure 4: CT aortogram showing wall thickening and luminal narrowing involving the entire length of the right radial artery without enhancement of the vessel wall

Vascular disease in SLE can be classified as (a) lupus vasculopathy, (b) necrotizing vasculitis, (c) thrombotic microangiopathies, and (d) accelerated atherosclerosis.[4]

Vasculopathy in SLE is a rare form of vascular involvement characterized by arterial stenosis or occlusion, with the accumulation of immunoglobulins and complement in the arterial intima and no inflammatory change.[2] The pathogenesis of lupus vasculopathy is not known. Accumulation of immunoglobulins and complements in the vessel wall suggests an immune-mediated vascular injury, however, lack of inflammatory cell infiltration into the vessel wall in lupus vasculopathy remains an enigma.[5-7] Lupus vasculopathy is commonly found in the kidneys and the pathological findings in renal lupus vasculopathy are mainly present in small vessels, such as the preglomerular arterioles and small arteries.[8] Although documented predominantly in the kidney, lupus vasculopathy can involve the heart, joints, spleen, skin, retina, and lungs[8] with most of them involving the small vessels.[2,9,10] Previous cases of lupus vasculopathy were mainly treated with immunosuppressive therapy because of coincidental active lupus involvement or its hypothesized mechanism, however, it has not been established whether conventional immunosuppressive agents are effective.[2]

The reported incidence of lupus vasculitis ranges from 11% to 36%.[11,12] The majority of cases have small vessel vasculitis (90%) with cutaneous lesions being the most common presentation. Medium vessel vasculitis is less frequent and has predominantly visceral vasculitis with mononeuritis multiplex as the most frequent form followed by abdominal vasculitis.[12] Antibodies against endothelial cells have been implicated in the pathogenesis of several connective tissue diseases, predominantly vasculitides.[13] Lupus vasculitis requires exclusion of antiphospholipid associated vascular occlusion or embolic vascular occlusion. Differentiation is often difficult as both can occur simultaneously.

Our patient tested negative for antiphospholipid antibody panel. Her colour doppler flow imaging (CDFI) (arterial) right upper limb revealed irregular wall thickening with a narrow lumen and mildly reduced peak systolic volume, following which a CT aortogram was done, which revealed mild narrowing of the entire length of right brachial artery and significant luminal narrowing of the right radial artery starting from its origin along with wall thickening with no vessel wall enhancement.

Hence based on clinico-radiological analysis, a likely possibility of medium vessel vasculopathy involving the right brachial and the radial artery was made, which is a rare form of vascular involvement in SLE. She refused for biopsy of the radial artery for definite diagnosis and was managed on low dose steroids and immunosuppressants following which the skin lesions and the oral ulcers regressed; however, the absent pulse was status quo till the last visit.

We report this case for its rare clinical presentation and also to stress the relevance of a good detailed clinical examination so as to timely diagnose the life-threatening presentations of an uncommon condition.

Conclusion

Vasculopathy is a rare vascular involvement in systemic lupus erythematosus and at times it can have very subtle presentations. A good detailed clinical and radiological evaluation can timely diagnose the serious manifestations of this uncommon condition.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

References

1. Rose NR, Mackay IR. The Autoimmune Diseases.Vol 2. Academic Press, INC; 1992.p. 279-300.
2. Waki D, Onishi A, Morinobu A. Large vessel vasculopathy in a patient with systemic lupus erythematosus: A case report. J Med Case Rep 2019;13:189.
3. Pyrpasopoulou A, Chatzimichailidou S, Aslanidis S. Vascular disease in systemic lupus erythematosus. Autoimmune Dis 2012;2012:876456.
4. Seshan SV. Lupus vasculopathy and vasculitis: What is the difference and when do they occur? AJSP RevRep 2007;12:214-21.
5. Appel GB, Pirani CL, D'Agati V. Renal vascular complications of systemic lupus erythematosus. J Am Soc Nephrol 1994;4:1499-515.
6. Seshan S. Lupus vasculopathy and vasculitis-what is the difference and when do they occur? Pathol Case Rev 2007;12:214-22.
7. Descombes E, Droz D, Drouet L, Grünfeld JP, Lesavre P. Renal vascular lesions in lupus nephritis. Medicine (Baltimore) 1997;76:355-68.
8. Gonzalez-Suarez ML, Waheed AA, Andrews DM, Ascherman DP, Zeng X, Nayer A. Lupus vasculopathy: Diagnostic, pathogenetic, and therapeutic considerations. Lupus 2014;23:421-7.
9. Sung JM, Hsu SC, Chen FF, Huang JJ. Systemic lupus erythematosus presented as non-inflammatory necrotizing vasculopathy-induced ischemic glomerulopathy and small vessels-related ischemic cardiomyopathy. Lupus 2002;11:458-62.
10. Hughson MD, He Z, Henegar J, McMurray R. Alveolar hemorrhage and renal microangiopathy in systemic lupus erythematosus: immune complex small vascular injury with apoptosis. Arch Pathol Lab Med 2001;125:475-83.
11. Drenkard C, Villa AR, Reyes E, Abello M, Alarcón-Segovia D. Vasculitis in systemic lupus erythematosus. Lupus 1997;6:235-42.
12. Ramos-Casals M, Nardi N, Lagrutta M, Brito-Zerón P, Bové A, Delgado G, *et al*. Vasculitis in systemic lupus erythematosus: Prevalence and clinical characteristics in 670 patients. Medicine (Baltimore) 2006;85:95-104.
13. Guilpain P, Mouthon L. Antiendothelial cells autoantibodies in vasculitis-associated systemic diseases. Clinic Rev Allerg Immunol 2008;35:59-65.