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Tongue Involvement in Systemic Lupus Erythematosus – A Case Report

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Abstract:

Lupus was first recognised as a systemic disease with visceral manifestations by Moriz Kaposi (1837–1902). Systemic lupus erythematosus (SLE) is a systemic, chronic inflammatory condition with diverse clinical manifestations, primarily affecting the joints, internal organs, and the skin. It commonly affects patients in the fourth decade of life, especially women with a ratio 7 to 10. Its cause is still unknown and genetic, immunologic, hormonal and environmental factors have been implicated in its pathogenesis. The prevalence of mucosal involvement in SLE patients is debatable. Oral lesions are present in 9–45% of patients with SLE.

A 45 year old female patient reported to Darshan Dental & Oral cancer center with complaint of reddish lesion on the tongue. Clinical examination revealed grayish white lesion on left anterior dorsal part of tongue.

Histopathology revealed basal cell degeneration and diffuse sub-epithelial lymphocytic infiltration. PAS staining revealed diffuse positivity along the basement membrane. Her ANA values were 345. Correlating the clinical, Histopathological and serological findings, she was diagnosed with SLE. The patient is under regular follow up.

Keywords:	*Systemic Lupus Erythematosus, Tongue, Anti-Nuclear Antibody*
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1. Introduction

The term ‘lupus’ (Latin for ‘wolf’) was first used during the middle Ages to describe erosive skin lesions evocative of a ‘wolf’s bite’. Lupus was first recognized as a systemic disease with visceral manifestations by Moriz Kaposi (1837–1902). The description of the false positive test for syphilis in Systemic Lupus Erythematosus (SLE) by Reinhart and Hauck from Germany (1909); the description of the endocarditis lesions in SLE by Libman and Sacks in New York (1923); the description of the glomerular changes by Baehr (1935), and the use of the

term diffuse connective tissue disease' by Klemperer, Pollack and Baehr (1941). The beginning of the modern era in SLE was the discovery of the 'LE' cell by Hargraves, Richmond and Morton at the Mayo Clinic in 1948.[1]

SLE is a chronic inflammatory condition. Its cause is still unknown and genetic, immunologic, hormonal and environmental factors have been implicated in its pathogenesis. The disease is more prevalent amongst women of childbearing age, although it can affect both sexes equally at any age.[2] Siblings of SLE patients are approximately 30 times more likely to develop SLE compared with individuals without an affected sibling[1].A higher SLE incidence is reported in Asian (especially Chinese), African, and Hispanic populations.[3]

GWAS in lupus have confirmed the importance of genes associated with immune response and inflammation (HLA-DR, PTPN22, STAT4, IRF5, BLK, OX40L, FCGR2A, BANK1, SPP1, IRAK1, TNFAIP3, C2, C4, CIq, PXK), DNA repairs (TREX1), adherence of inflammatory cells to the endothelium (ITGAM), and tissue response to injury (KLK1, KLK3). These findings highlight the importance of Toll-like receptor (TLR) and type 1 interferon (IFN) signaling pathways. Some of the genetic loci may explain not only the susceptibility to disease but also its severity. For instance, STAT4, a genetic risk factor for rheumatoid arthritis and SLE, is associated with severe SLE. One of the key components of these pathways is TNFAIP3, which has been implicated in at least six autoimmune disorders, including SLE.[1]

Environmental triggers of SLE include ultraviolet light, demethylating drugs, and infectious or endogenous viruses or viral-like elements. Sunlight is the most obvious environmental factor that may exacerbate SLE. Epstein–Barr virus (EBV) has been identified as a possible factor in the development of SLE. EBV may reside in and interact with B cells and promotes interferon-α (IFNα) production by plasmacytoid dendritic cells (pDCs), suggesting that elevated IFNα in lupus may be—at least in part—due to aberrantly controlled chronic viral infection.[1,3]

Classically, LE has been subdivided into a systemic and a cutaneous form. While systemic lupus erythematosus (SLE) is a multiorgan disease with variable prognosis, cutaneous lupus erythematosus (CLE) is a more benign condition – limited to skin and/or mucosal surfaces.[2] The disease appears to be more common in urban than rural areas and 65% of patients with SLE have disease onset between the ages of 16 and 55 years. Men with lupus tend to have less photosensitivity, more serositis, an older age at diagnosis, and a higher 1 year mortality compared to women. SLE tends to be milder in the elderly with lower incidence of malar rash, photosensitivity, purpura, alopecia, Raynaud's phenomenon, renal and central nervous system involvement, but greater prevalence of serositis, pulmonary involvement, sicca symptoms, and musculoskeletal manifestations.[1] The prevalence of mucosal involvement in LE patients is debatable. The oral lesions are present in 9–45% of patients with SLE and in 3–20% in those with CLE.[2]

The classic presentation is the oral discoid lesion, characterized by a well-demarcated zone of erythema, atrophy, or ulceration surrounded by white, radiating striae. These lesions appear similar to those found in patients with erosive lichen planus. Variations include honeycomb plaques (silvery white, scarred plaques), raised keratotic plaques (verrucous lupus

erythematosus, and non-specific erythema. Purpura, petechiae, or irregularly shaped ulcers also are possible. Discoid lesions like those typically found elsewhere on sun-exposed skin may be found on the lip vermilion. Cheilitis may be evident as well.[4] Oral lesions in patients with SLE typically resolve with systemic immunosuppressive therapy. For patients with limited skin or oral mucosal disease, topical corticosteroids or systemic anti-malarial drugs are appropriate.[5]

2. Case Report

A 45 year old female patient reported to Darshan Dental & Oral cancer center with complaints of reddish lesion on the tongue. She was also having generalized joint pain, but not having any skin lesion. Medical reports showed high cholesterol and she was not under any medication.

Clinical examination revealed well demarked bluish grey lesion on left dorsal part of the tongue. Its looks like hematogenic or degenerating changes, but patient was not having any history of trauma. (Figure 1 and Figure 2)

Histopathological features shows parakeratinized stratified squamous epithelium of variable thickness with atrophy in some places and basal cell degeneration (Figure 3 and Figure 4). A homogenous eosinophilic, acellular band was evident at the epithelial-connective tissue interface. The underlying fibrous connective tissue stroma shows patchy mono-nuclear inflammatory cell infiltrate. PAS staining revealed homogenous positivity in the basement membrane zone.Correlating the clinical findings and serum ANA values of 345 reported, histopathological features are suggestive of Lupus Erythematosus. She was advised dsDNA, SSA, SSB, centromere, Scl70 and ENA estimation for further treatment planning.

She was given Deflazacort 6mg twice a day along with topical Triamcinolone application on the tongue. The patient is under regular follow up for continuous monitoring of systemic complications.

3. Discussion

Immune responses against endogenous nuclear antigens are characteristic of SLE. Autoantigens released by apoptotic cells are presented by dendritic cells to T cells leading to their activation. Activated T cells in turn help B cells to produce antibodies to these self-constituents by secreting cytokines such as interleukin 10 (IL10) and IL23 and by cell surface molecules such as CD40L and CTLA-4. In addition to this antigen-driven T cell-dependent production of autoantibodies, recent data support T cell-independent mechanisms of B cell stimulation via combined B cell antigen receptor (BCR) and TLR signaling. The pathogenesis of SLE involves a multitude of cells and molecules that participate in apoptosis, innate and adaptive immune responses. Increased amounts of apoptosis-related endogenous nucleic acids stimulate the production of IFNα and promote autoimmunity by breaking self-tolerance through activation of antigen-presenting cells. Once initiated, immune reactants such as immune complexes amplify and sustain the inflammatory response.[1]

In a study of American College of Rheumatology, 46 patients with confirmed diagnosis of LE and presenting oral lesions were biopsied and analyzed microscopically by two pathologists. Sections of all biopsied lesions were stained with routine hematoxylin–eosin and periodic acid–Schiff (PAS). PAS stain was used to disclose the presence of colloid bodies in the epithelium and basement membrane thickening. Direct

immunofluorescence examination (DIF) was also performed in all specimens. The main histological aspects in all the oral biopsied lesions corresponded to a lichenoid mucositis with a deep and perivascular inflammatory infiltrate associated. The covering epithelium presented areas of acanthosis alternated with areas atrophy. In some specimens a pseudo-epitheliomatous proliferation was seen. Variable degree of spongiosis was observed in most cases. On three specimens focal areas of mild to moderate epithelial atypia was detected. Foci with hydropic degeneration of the epithelial basal layer were evident in all biopsies. Widespread or focal basal cell apoptosis, sometimes with the presence of colloid bodies, was frequently observed in the sections examined. These aspects are shown in. Thickening of epithelial and vascular basement membranes was clearly demonstrated on PAS-stained sections. DIF showed linear deposits of IgG and/or C3 in the basement membrane zone of all cases. Immunoglobulin (IgM) fluorescence on cytoid bodies was also observed in all samples. [6]

Our patient was diagnosed with SLE, on the basis of clinical findings, histopathology and ANA test. Right diagnosis at the right time could be life saving for the patient. Our patient had gone around multiple hospitals over the past few months for the tongue lesion. We performed the biopsy, appropriate serology and could reach the proper diagnosis.

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Figures with Legends:

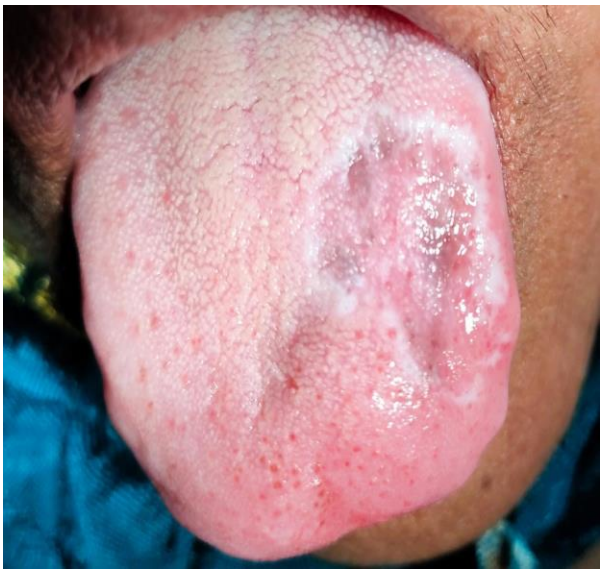

Figure 1 is the clinical picture showing the reddish-white lesion on the left dorsum of the tongue

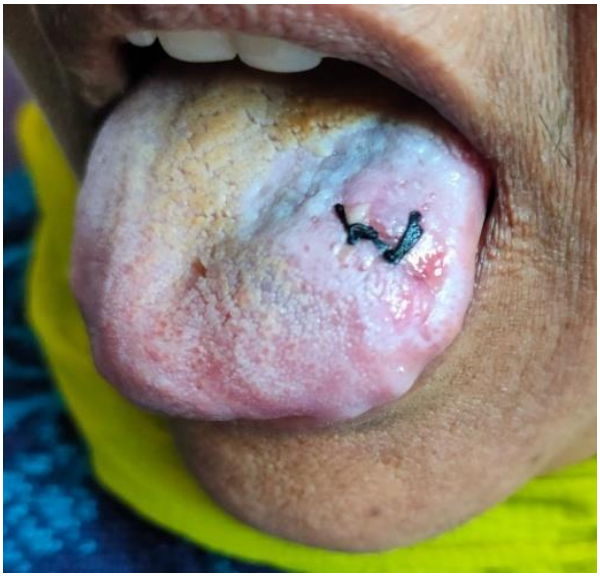

Figure 2 is the clinical picture showing the sutures after biopsy

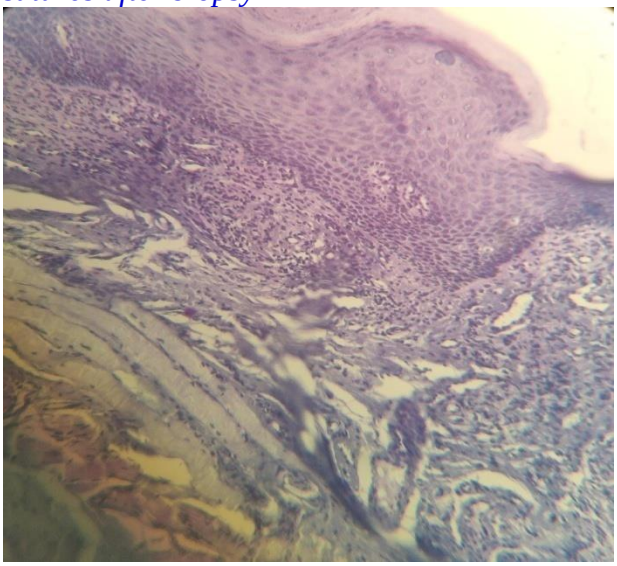

Figure 3 is the photomicrograph showing hyperkeratosis, basal cell degeneration and sub-epithelial lymphocytic infiltrate. (PAS, 10x)

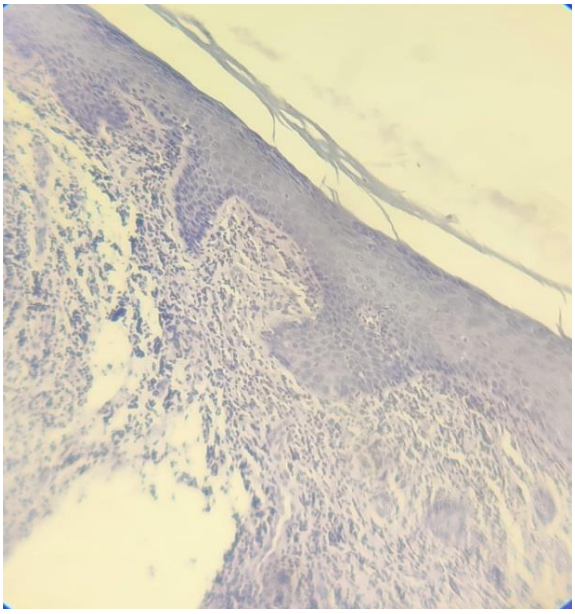

Figure 4 is the photomicrograph showing basal cell degeneration, sub-epithelial lymphocytic infiltrate and diffuse PAS positivity in the basement membrane zone. (PAS, 10x)