Oral Findings of Systemic Lupus Erythematosus- A Grand Round Case

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Received: 2 Feb., 2016
Accepted: 28 Mar., 2016
Published: 5 May, 2016

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Preferred citation for this article:

Abstract A case of an SLE patient with an erythematous plaque at hard palate surrounded by white striae and a chronic cutaneous lupus erythematosous (CCLE) history is being reported. Other authors’ findings are being critically discussed and we focus on the following:

i. Need of oral lesions histopathologic examination in SLE patients as they contribute to diagnosis establishment.
ii. Oral ulcers do not certainly constitute neither the unique nor the main oral lesion in SLE since apart from them are reported red and white plaques, red macules, lechinoid reactions as well as periodontal manifestations (marginal or/and desquamative gingivitis).
iii. Biopsy is crucial for excluding oral squamous cell carcinoma, especially in oral ulcers.
iv. In the case of SLE xerostomia minor salivary glands biopsy contributes to secondary Sjögren syndrome diagnosis.
v. Contribution of oral manifestations in early SLE diagnosis, leads to effective treatment and prevention of serious systemic malfunctions, potentially lethal.

Keywords Systemic lupus erythematosus; Oral findings; SLE diagnostic criteria

1 Introduction
Systemic Lupus Erythematosus (SLE) is an autoimmune connective tissue disease commonly affecting patients in the fourth decade of life, especially women with a ratio 7 to 10:1.

Clinical features include general symptoms and signs, such as fever, fatigue and weight loss, as well as involvement of a variety of internal organs and systems.

In 1982 the American College of Rheumatology classified all those manifestations in eleven groups, establishing the SLE criteria of classification. Disease diagnosis is established when a patient presents with at least four of the previous criteria. Among these, oral (or/and nasopharyngeal) ulcers stand in the fourth place (Klasser et al., 2007; Tang et al., 1982).

2 Case Report
A 74 –years old woman was referred to our clinic because a six days fever accompanied with neutropenia and normochromic anaemia.

Her medical history included Chronic Cutaneous Lupus Erythematosus without any specific treatment since 20 years, hypertension as well as a transient ischemic attack ten years before without any disability.

Patients’ vital signs were normal and her initial clinical examination was normal too.

Laboratory examination: WBC=2200 (neutrophiles 28%, lymphocytes 48%, monocytes 20%), Ht=27,4,
PLT=267.000, MCV=77, MCH=26, ESR=76, CRP was normal despite high ESR, urine analysis was normal, Glucose =161, urea=30, Na=129, K=4,5.

Serum blood and urine cultures were negative. Hepatic enzymes were within normal limits. Serologic examination for HBV, HCV and HIV was negative. Complement fragments C3, C4 were found extremely low.

3 Clinical and histopathological oral findings
Oral examination revealed an erythematous plaque of about 5mmx 3mm at right hard palate mucosa, near the soft palate border. Lesion was surrounded by a painless white border (Figure 1).

There was also a red painless macule at the soft palate, close to the previous lesion, about 2 mm x 3 mm (Figure 1).

Figure 1: An erythematus plaque at right hard palate mucosa surrounded by white border. The dorsal tongue surface is furred

Remain oral cavity appeared normal. There was moderate alveolar bone resorption due to the fact of her being edentulous for the last twenty years. Oral mucosa appeared dry and her tongue was furred (Figure 1), due to fever or/and possible secondary Sjögren syndrome (SS). The patient did not complain for burning sensation.

In accordance to her medical situation and oral examination, oral lesions were diagnosed to be SLE manifestations. Differential diagnosis included oral lichen planus. Clinical diagnosis was established by incisional biopsy, performed under local anesthesia in lesional mucosa, extending to clinically healthy mucosa. Biopsy for the establishment of probable secondary SS was performed in labial minor salivary glands.

Histological examination reveals atrophic areas of covering epithelium. Moderate inflammation exists in chorium, with lymphocytes, plasma cells and a few neutrophils. Hydropic degeneration of basal epithelium layer exists focally, along with presence of lymphocytes and a few colloid bodies. All the above findings are compatible with SLE (Figure 2, 3). Minor salivary gland histological examination reveals no pathological signs.
4 Clinical course

The patient was initially treated with broad spectrum antibiotics and was administered two RBC units. On the fourth day she complained for acute chest pain (with non-specific ECG changes) so she was administered sublingual nitrates. She was still febrile and on the eighth day she presented focal seizure with absence and consciousness loss. Urgent brain CT revealed leucoencephalopathy and cerebral atrophy. In consequence of her episode she was treated with phenytoine. Starting from the following day, the patient's general status was gradually worsened (fatigue, anorexia, dyspnea, hypoxemia, confusion, and tachycardia). Chest x-ray revealed pulmonary infectious infiltrations of the right lung. She developed acute renal and liver failure accompanied by INR prolongation. Those findings were compatible with septic multiorganic failure. Since then she was administered full-dose corticosteroids as well as oxygen, hydration and inotropes. Urgent chest and abdomen CT scans revealed pneumonia and pleural fluid collections and on the 14th day the patient died.

Her medical history, clinical course and laboratory findings confirmed septic shock due to SLE pneumonia as a result of undertreated CCLE.
5 Discussion

Oral lesions constitute an appreciable clinical manifestation in SLE patients. In particular, ulcers of oral mucosa possess the fourth place among the eleven diagnostic criteria, as revised by the American Rheumatology College. Incidence is reported to be between 7% and 41% (Brennan et al., 2005; Sverzut et al., 2008).

Oral ulcers do not, certainly, constitute neither the unique nor the main oral lesion in SLE, since apart from them, are reported: red or/and white plaques (Lourenço et al., 2006; López-Labady et al., 2007), red macules surrounded by white limits with radiating striae (López-Labady et al., 2007), lichenoid reactions (Lourenço et al., 2006; López-Labady et al., 2007), atrophic plaques mainly on the vermillion border of lower lip (Rhodus and Johnson, 1990; Lourenço et al., 2006).

Lourenço et al. (2006) studied 188 patients with SEL; eighty one (81) of them SLE patients and the rest Cutaneous Lupus Erythematosus (C.L.E) patients. Twenty six (26) patients had oral lesions, thirteen (13) of them being S.E.L and thirteen (13) CLE patients. The percentage of SLE patients who presented oral lesions is 16%. Writers did not make clear about the type of oral lesions concerning SLE patients, while CLE patients as well as S.E.L patients belong to the same study group. However, out of 26 patients there is reported only one case with ulcerated plaques of lower lip and buccal mucosa and two cases with vesiculoulcered lesions of tongue, lips and buccal mucosa. Findings are not examined histopathologically.

López-Labady et al. (2007) studied 90 CLE patients; out of them, 36 were SLE patients. Six of them had oral lesions (16, 6%) and three ulcers surrounded by white radiating striae, while all of them had red macules or/and plaques as well as lichenoid lesions of buccal mucosa. Oral findings are examined histopathologically.

Finally, Sverzut et al (2008) refer a case of a young female patient with oral ulcers. She was diagnosed as a SLE patient. Unfortunately, oral lesions were not biopsied.

Histopathologic findings do not, certainly, prove SLE existence, but are only indicative. However, they establish the diagnosis since, as already mentioned they are included among the diagnostic criteria of SLE. Consequently the oral physician can contribute in an early diagnosis of the disease as well as in its documentation.

Marginal gingivitis, desquamative gingivitis and the presence of gingival erosions are among SLE oral lesions (Jayakumar et al., 2006; Albilia et al., 2007). Jayakumar et al. (2006) report a case of a young woman with desquamative gingivitis, followed by the existence of erythematous lesions surrounded by white border radiating striae of the buccal mucosa opposite from the upper third molar. Writers proceed to gingival biopsy; histopathological findings were compatible with SLE. However, the buccal mucosa lesion was not investigated histopathologically, which ought to have been done, since oral lesions are used, among other systematic findings, in order to establish SLE diagnosis.

Oral lesion biopsy, apart from its contribution to establish SLE diagnosis, also contributes in differential diagnosis from other serious diseases (particularly in the existence of ulcers) like oral squamous cell carcinoma which, as known, in an early stage might present in the form of an ulcer or in the form of a white or red plaque.

Opinions concerning the presence of oral ulcers as an indicator of clinical activation of the disease are contradictory (Rhodus and Johnson, 1990; Brennan et al., 2005).

Xerostomia constitutes a common finding among SLE patients. Patients with xerostomia present increased sensitivity in tooth decay and in periodontal disease. They also present difficulty in chewing, in ingestion and in speech, intolerance in artificial dentures as well as dysgeusia. Oral mucosa appears red and shiny (particularly tongue mucosa) and often there are erosions present, leading to unpleasant burning or pain sensation, which, when located on tongue, is characterized as glossodynia. Oral candidiasis constitutes a usual complication of xerostomia, while its frequency is increased on immunosupressed patients, as it often happens in SLE patients who are under pharmaceutical immunosupression (eg long-term steroidal therapy) (Grimaldo-Carjevschi et al., 2011).
Salivary glands malfunction is due to their chronic autoimmune inflammation, as happens in Sjögren syndrome. As known, Sjögren syndrome is likely to present secondarily and along with autoimmune disease of the conjunctive tissue, usually rheumatoid arthritis (Rhodus and Johnson, 1990; Brennan et al., 2005; Fernandez et al., 2010; Baer et al., 2010).

Rhodus and Johnson (1990) while studying 16 SLE patients (women) focused their interest in salivary glands function and found that they all suffered from xerostomia, while, using sialometric methods, they proved decreased salivary glands function.

Oral findings of the previous authors were considered as a result of xerostomia. Among them, in first two places stand caries (100%) and periodontitis (93%) while at the third place (87, 5%) are angular cheilitis as well as Cheilitis ochilosis and glossodynia. Angular cheilitis can be attributed to oral candidiasis.

However, it ought to be mentioned that in all previous patients, biopsy of minor salivary glands in order to establish secondary Sjögren syndrome, was not performed.

In conclusion, it becomes perceptible that SLE patients have oral pathological interest (Nico et al., 2011; Heath et al., 2015). The localisation of oral lesions and their evaluation along with the other clinical pathological and laboratory findings contribute in the early diagnosis of the disease, and consequently in a more effective treatment along with the prevention of serious malfunctions (potentially lethal) of various internal organs of those patients.

References
http://dx.doi.org/10.1002/art.1780251101
http://dx.doi.org/10.1016/j.dcen.2004.07.006
http://dx.doi.org/10.1111/j.1600-0560.2009.01368.x
http://dx.doi.org/10.4103/0970-9290.29883
http://dx.doi.org/10.1111/j.1600-0714.2007.00569.x
http://dx.doi.org/10.1111/j.1600-0560.2006.00518.x
http://dx.doi.org/10.2340/00015555-1103
http://dx.doi.org/10.1002/art.1780251101

5