Persistent Palatal Ulcerations. A Potential Manifestation of Juvenile Systemic Lupus Erythematosus

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Oral ulcerations in children and adolescents is a common occurrence and affects about 20-30% of this population. This case report describes a unique and serious autoimmune condition that presented with distinct oral findings that significantly supported the differential diagnosis of Juvenile Systemic Lupus Erythematosus in a 15 year-old female. Pediatric and general dentists should familiarize themselves with the condition to facilitate diagnosis with collaborative efforts with the medical team

Keywords: *Juvenile Systemic Lupus Erythematosus, oral ulcers, autoimmune disease, malar rash*

INTRODUCTION

Juvenile-onset systemic lupus erythematosus (JSLE) is common autoimmune condition affecting children with clinical variability and severity.[1] Twenty percent of systemic lupus erythematosus (SLE) will be diagnosed before puberty, with a median age of 11–12 years.[2-5] The prevalence of JSLE is high in Asian and African communities, with a female predominance 4.7:1–6.2:1).[6,7] JSLE patients usually manifest with mucocutaneous lesions, kidney ,central nervous system (CNS) diseases and hematological dysfunctions.[4] The "butterfly"- rash across the cheeks and bridge of the nose, oral ulcers are the most prevalent presentation of mucocutaneous lesions in JSLE patients.[8] Up to 30 % of JSLE patients is will present with oral ulcerations which is significantly higher than in adult SLE [4,5] with no gender preference in children.[9] The common oral ulcers in JSLE patients are palatal erythematous ulcers and aphthous-like ulcers.[10,11]

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The purpose of this manuscript is to demonstrate the oral manifestations of JSLE in an adolescent female and to underscore the importance of exploring the oral involvement in order to enhance and facilitate the diagnostic process.

Case report

A 15-year-9 month-old Hispanic female was referred to the Pediatric Dentistry Graduate Clinic for a consultation regarding a lesion/ulcer in the hard palate persisting for six months. Patient was admitted to the Pediatric Rheumatology Clinic at UF Health Hospital at the University of Florida for a second opinion regarding a potential diagnosis of SLE. Patient reported having headaches and fatigue for the past three years, had developed polyarthralgias on her hands, knees, ankles and morning stiffness. She had been experiencing fever (37.5˚C -38˚C) and night sweats since the past month. Her initial lab work revealed strong positive serology for SLE including positive antinuclear antibody (ANA) (high titer), high anti ds-DNA levels, positive Sm, Ro, La and RNP. The direct anti-globulin test was positive +++. Her last visit to the dentist was two years ago. Patient's chief complaint included pain on the roof of the mouth.

Extra-oral examination revealed hypopigmented malar rash that spared the nasolabial fold. Lymphadenopathy presented in anterior cervical, posterior cervical, and submandibular nodes that were non-fixed, non-tender, and a size of 2.0 x 2.0 cm diameter was noted. Enlargement of the right parotid gland was noted and the inferior lobe was palpable. Intra-oral findings included moderate pooling of saliva on the floor of the mouth, multiple raised erythematous lesions on the ventral aspects of the tongue and a 1.5 cm exophytic erythematous lesion on hard and soft palate. Numerous ulcerations of the mandibular buccal gingiva of tooth #35, ventral surface of the tongue, lower lip and the roof of mouth (palatal) mucosa was noted. (Figure 1). Tissue sample from the buccal gingival tissue of tooth

#16 and the palate area were excised, embedded in formalin and Michel's solution respectively and submitted to biopsy and routine hematoxylin and eosin (H&E) stained examination with direct immunofluorescence (DIF) studies. Routine histology revealed superficial parakeratinized stratified squamous epithelium overlying a slightly nodular mass of dense fibrous connective tissue. The epithelium was detached in one portion of the specimen and the surface keratin layer was slightly frayed and shaggy. The epithelium was hyperplastic with areas that remain attached exhibiting a mildly eosinophilic coagulum in the superficial lamina propria. Tissue was also submitted to direct immunofluorescence antibody staining, which was positive at the point of the basement membrane for IgM, IgA, IgG and C3 and negative for fibrinogen. The clinical parameters and the oral presentation when combined with the histological findings were compatible with the serological and biopsy supported diagnosis of JSLE.

DISCUSSION

Our patient presented with the common oral manifestations of JSLE. Most significantly, she experienced painful tender palatal lesions (which is one of the hallmarks in JSLE spectrum and is included in the diagnostic criteria of this condition), chelitis of the lower lips, presence of erythematous nodules on the ventral aspect of the tongue, and ulcerations of the gingiva. However, since the clinical differential diagnosis included also other ulcerative conditions an incisional biopsy had to be performed. Immunofluorescence study was positive for IgG, IgM, IgA and C3 at the basement membrane and was consistent with SLE. Combined with the positive autoimmune serology, the oral biopsy confirmed the diagnosis of JSLE.

Mucocutaneous lesions are frequently presented in JSLE patients at diagnosis, and affect approximately 70–75% of JSLE populations worldwide. [7,10,11] According to the American College of Rheumatology (ACR) revised criteria for the classification of SLE, four mucocutaneous features are postulated including oral ulcers. The description of oral ulcers in these criteria is "painless oral or nasopharyngeal ulceration observed by a physician", yet these may be unclear, especially in children.[12] In the new Systemic Lupus International Collaborating Clinics group classification criteria, oral ulcers remain included. They are defined more specifically as oral ulcers at palate, buccal mucosa or tongue without other causes (e.g., vasculitis or infection).[13] Several types of oral ulcers may be presented in JSLE patients; some of them (e.g., palatal erythematous ulcers and aphthous ulcers) associated with the disease activity. Other forms of oral ulcerations may be not related disease severity.[14] Erythematous ulcers of the palate are presumed to be specific for SLE lesions they may present as a painless, single/ multiple lesion(s) on the masticatory or keratinized mucosa, especially the hard palate (Figure 1). It is an acute sign occurring when disease is active, and sometimes is the first clue of JSLE without skin lesions.[15] The early lesion may be hemorrhagic in nature prior to development of an actual open ulcer.[10] When ulcers coalesce into a large erythematous

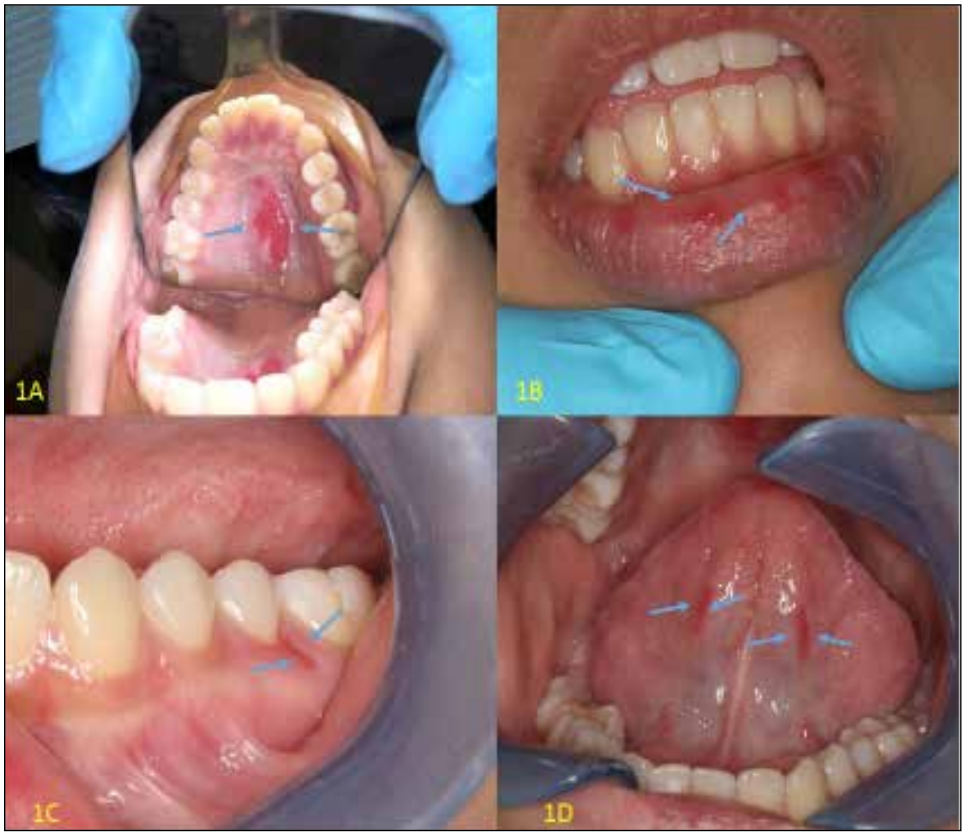

Figure 1: Multiple ulcers and erythematous papules (marked with blue arrows) involving the roof of the mouth (1A), the lower lip 1B, mandibular buccal gingiva area of tooth #35 (1C) and the ventral surface of the tongue (1D).

center on the hard palate extending to the soft palate this may be suggestive of a diagnosis of onset oral JSLE

Other manifestations of JSLE include chelitis of the lips presenting as small or diffuse, red edematous, or crusty painful ulcers.[17] Chelitis often involves the vermilion zone of the lower lip (typical lupus cheilitis).[18] As the lesion is found in the sun-exposed area, lupus chelitis is usually associated with photosensitivity in JSLE patients.[19] Extensive erosive lupus chelitis that involves both upper and lower lips has also been reported in JSLE patients and appears in active disease.[20]

Other oral presentation in JSLE include lingual infarction (localized necrosis), higher incidence of periodontitis and TMJ dysfunction[22]. In spite of the high incidence of JSLE among SLE patients and the high prevalence of oral lesions in this condition, there are scanty reports in the dental literature on the oral manifestation of JSLE. In view of the clinical and diagnostic importance of oral and especially palatal lesions, increased awareness to this condition among dentists who treat children and adolescents is warranted.

CONCLUSION

Palatal, persistent, and erythematous ulcers in a pediatric patient with a history of severe arthritis and positive bloodwork for autoimmunity should alert the oral health care provider about the potential diagnosis of JSLE. Close collaboration between the dental and medical disciplines, may provide a prompt diagnosis and a beneficial medical outcomes for the patient.

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