

Systemic Immunological Diseases With Oral & Mucocutaneous Manifestations

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ABSTRACT

Background

The oral and mucocutaneous manifestations of systemic immunological diseases vary greatly in its presentation ranging from ulcerations to blisters accompanied with or without burning sensation or pain. The main objective of this study was to identify to prevalence of oral and mucocutaneous manifestations of systemic immunologic diseases in Chennai, South India.

Method

Archived records of 6300 cases from an institution in Chennai, South India was done. A total of 105 cases with oral and mucocutaneous manifestations were narrowed down. The case history, clinical features and histopathological features of these cases were examined. The descriptive statistics was done using SPSS.

Results

Females had a greater presentation of oral lesions when compared to males and age group of 60 years and above exhibited more lesions compared to the others. The most common lesions included lichen planus, pemphigoid and pemphigus.

Conclusion

Sometimes these diseases present oral manifestations as the first sign and symptom which can be identified by dentists. Early diagnosis is necessary for proper management and prevention of any serious sequelae

Keywords: Oral manifestations, Lichen planus, Pemphigoid.

INTRODUCTION

The oral cavity can be regarded as a mirror of the general health because few systemic diseases present with local symptoms and/or lesions in the oral mucosa (1). Signs and symptoms of immunologic diseases, hematologic conditions, endocrinopathies, systemic infections, and nutritional disorders can be identified by careful intraoral examination (2). The manifestations of these diseases may vary in frequency and location (3) and they can manifest as ulcerations, discolorations or loss of elasticity associated to the systemic disease in both the soft and hard tissues in the oral cavity (4). Even though from histological and embryological perspective, the oral mucosa is similar to that the skin, the pattern of disease presentation varies (1,5).

Several immune mediated inflammatory mucocutaneous diseases affect the oral cavity that present as blisters, sloughing, erosions, ulcerations and pain (6). The common mucocutaneous disorders that affect the oral cavity include oral lichen planus, mucous membrane pemphigoid and pemphigus vulgaris (7). These conditions can have a serious sequelae if left untreated therefore early and proper diagnosis is essential for patient management(6)

A dental surgeon is the first person to come across these lesions that have similar clinical characteristics, therefore their identification based solely on oral lesions is a challenging issue for dentists, leading to a delay in the establishment of a correct diagnosis and suitable management of the patient. The main objective of this study was to identify to prevalence of oral and mucocutaneous manifestations of systemic immunologic diseases in Chennai, South India.

Material and methods

A total of 6300 case records from the archives from archives dated 1997 – 2018 was assessed. Out of which 105 cases with systemic immunological diseases exhibiting oral& mucocutaneous manifestations were assessed. Clinical information was retrieved from records and histopathologic features were reviewed. Demographics, chief complaint, history of presenting illness, past medical history, extra oral and intra oral examination were assessed. Intraoral examination details involved assessment of hard tissue, soft tissue and radiographic examination. The site, size, shape, colour, consistency and extension of the lesions were assessed. The results were collected and analysed with descriptive statistics using Statistical Package for Social Sciences (SPSS version 21).

Results

Demographics

The gender distribution was 27% male and 51% female (Figure 1). The age group of the records examined ranged from 20- 60 years. 8% of the patients were in the age group of 20-30 years, 18% in the 31-40 age group, 30% in 41-50 age range and the remaining majority of 38% above 50 years of age (Figure 2).

Site of the lesion

Most of the lesions diagnosed where present in the buccal mucosa and gingiva followed by tongue.

Prevalence of lesion

The majority of the lesion examined present were lichen planus (86%), pemphigoid (15%) followed by pemphigus (4%).

DISCUSSION

Out of 6300 specimens, these immunologically mediated diseases accounted for 105 (1.66%) cases, of which 86 (1.36%) were lichen planus, 4 (0.06%) were pemphigus and 15 (0.23%) were pemphigoid.

Lichen planus is an immune mediated disease with unknown aetiology. It is a non-scrapable white lesion which can also present as smooth red ulcers. Burning sensation may be present occasionally. The characteristic feature is the presence of Wickhams striae. The histopathological features include hyperkeratosis, acanthosis, saw tooth rete ridges, civatte bodies and sub epithelial infiltration of inflammatory cells (Figure 4).(8) Pemphigoid is the most common autoimmune bullous disorder first characterized by Lever in 1953. It most commonly affects the elderly with autoantibodies directed against hemidesmosome proteins (BP180 and BP230) leading to destruction of the basement membrane zone. It presents intraorally as multiple ulcers or erosions with burning sensation. The histopathological features include the characteristic subepithelial clefting, intact epithelium and dense inflammatory cell infiltrate (Figure 5) (9,10).

Pemphigus is an IgG mediated autoimmune disease with three major subtypes including pemphigus vulgaris, pemphigus foliaceus and paraneoplastic pemphigus. The word pemphix means blister or bubble (11). The IgG autoantibodies are characteristically directed against desmogleins 1 and 3 (12). They can present as ill-defined ulcers, erosions with burning sensation and characteristic Nikolsky's sign. The histopathological features include suprabasilar spilt, acantholysis, presence of Tzanck cells and relative scarcity of inflammatory cell infiltrate (Figure 6)(13).

It is noteworthy that the lesions in these 3 diseases have similar clinical characteristics, therefore their identification based solely on oral lesions is a challenging issue for dentists. A correct & early diagnosis is of paramount importance for a proper therapeutic decision

and appropriate approach to the patient's treatment.

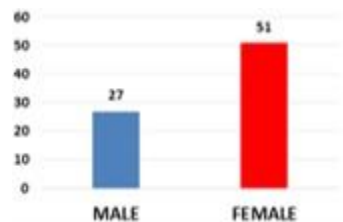
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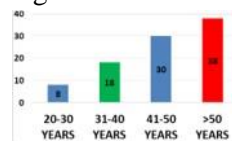
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Figure 1

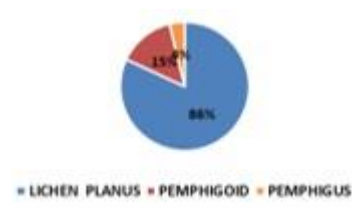
Gender distribution

**Figure 2**

Age distribution

**Figure 3**

Prevalence of the lesion

**Figure 4**

Histopathology of lichen planus

**Figure 5**

Histopathology of pemphigoid

**Figure 6**

Histopathology of pemphigus

