

Pemphigus vulgaris presenting as bleeding gums: A case report

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Abstract

Pemphigus vulgaris (PV) is a chronic autoimmune disease which may be potentially life threatening. PV manifests as blisters and erosions of the skin and the mucous membrane. It can manifest initially as oral lesions. It is essential to diagnose this condition early in order to avoid the serious complications. We describe a case of 58-year-old male presented with bleeding gums and burning sensation for 6 months. Patient was previously misdiagnosed as gingivitis. Though it is rare for pemphigus vulgaris to present with gingival involvement as primary site of manifestation, our case emphasizes the importance of thorough history and clinical examination for the early diagnosis and management of such condition.

Keywords: Pemphigus Vulgaris; Oral Lesions; Gingivitis.

INTRODUCTION

Pemphigus Vulgaris (PV) is an autoimmune disorder of the skin and mucous membrane associated with intraepithelial blistering (1,2). It is a chronic disease that affects both the genders. It is commonly seen in middle-aged individuals around the 5th and 6th decade of life (2).

The word pemphigus originated from the Greek word "pemphix" which means 'to blister or bubble'. The intraepithelial blistering is caused due to acantholysis within the epithelium (2). Pemphigus vulgaris, Pemphigus vegetans, Pemphigus erythematosus and Pemphigus foliaceus are four major form of pemphigus. Only the vulgaris and vegetans types are associated with oral mucosa with pemphigus vulgaris being the commonest type (3). Pemphigus Vulgaris accounts for over 80% of cases of pemphigus (3,4).

In about 70-90% of the cases, the first sign of the disease is seen in the oral cavity. The buccal mucosa is the commonest site followed by the palate, lingual and labial mucosa (4). Gingiva is rarely affected and when involved manifests as desquamative gingivitis (DG) (4). In most patients with PV, the oral lesions are followed by involvement of the skin (1,4). However, in few cases, the oral cavity may be the only site of involvement for nearly a year (4).

Thus early diagnosis of PV is essential so that treatment can

be initiated at the earliest and progression of the disease and development of skin lesions can be prevented (4). This report describes a patient complaining of bleeding gums and burning sensation in the oral cavity for nearly 6 months who was diagnosed to have PV.

CASE REPORT

A 58-year-old male patient reported to the Outpatient Department of our dental hospital with complaint of bleeding gums and burning sensation in the oral cavity since 6 months. Bleeding gums was present since 2-3 months and aggravated on brushing. Burning sensation was continuous in nature and increased during salt intake. Patient complained of difficulty in consumption of spicy food.

Patient visited a local doctor for the same purpose, where he was diagnosed of having gingivitis and was prescribed certain medications. No history of relief with the medications. History of throat pain few months ago for which the patient took medication after which he noticed vesicle or blister formations in the oral cavity. Patient reported that the blisters used to form in certain areas then ruptured and was then followed by formation of blisters in new areas intraorally. Patient also gave a history of rashes and allergic reactions on the skin of upper limb, lower limb, back and irritation and redness in the eyes. There was no history of prolonged hospitalization, systemic illness or use of long term

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medication. The patient had history of betel quid chewing and smoking for past 10 years.

On extra oral examination bilateral submandibular lymph nodes were enlarged, palpable and tender. On intra-oral examination multiple erosions were noticed involving the gingiva, tip of the tongue, floor of the mouth, soft palate, labial and buccal mucosa (Figure 1A). Tissue tags were present at the periphery of erosions present over left labial and buccal mucosa (Figure 1B). On inspection of the left labial mucosa an ulcer measuring about 0.5cm to 1 cm in size was noticed. The ulcer was ovoid in shape with irregular borders which was covered with whitish pseudo membrane (Figure 2A and 2B). On palpation there was bleeding and the lesion was tender to touch. Round to oval erosion was noticed on the tip of the tongue measuring about 0.5cm in size. On palpation there was bleeding and was associated with pain. Multiple areas of ulceration and erosion measuring less than 1cm were noticed on the lateral border of the tongue, floor of the mouth, buccal mucosa and lower vestibule which were associated with bleeding and tenderness on palpation. Nikolsky's sign was positive in our case.

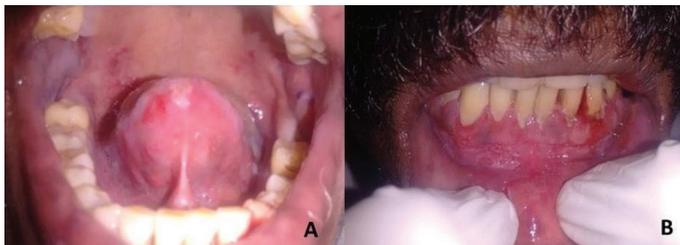


Figure 1 A. Intraoral picture showing erosions involving tip of the tongue and soft palate

Figure 1 B. shows erosion on the marginal and attached gingival and lower labial mucosa. Tissue tags were present at the periphery of erosions present over left lower labial mucosa



Figure 2A. Intraoral picture showing multiple areas of ulceration and erosion noticed on the floor of the mouth

Figure 2 B. Intraoral picture showing erosion on the right upper labial mucosa

On inspection of the gingiva erythematous and oedematous areas noticed including both the marginal as well as the attached gingiva indicative of desquamation (Figure 3). Generalized attrition of the teeth and gingival inflammation with bleeding on probing was present. History of repeated episodes of multiple blisters in the oral cavity, features of multiple oral ulcers, gingival desquamation and positive Nikolsky's sign lead to the diagnosis of Pemphigus vulgaris. The differential diagnosis of mucous membrane pemphigoid was considered.

Exfoliative cytology of the left buccal mucosa was done. Medical opinion from the department of dermatology and ophthalmology was sought. The patient was advised for biopsy, indirect and direct immunofluorescence. Histopathological examination of the smear revealed few round epithelial cells with enlarged and hyperchromatic nuclei suggestive of Tzanck cells. Numerous acute inflammatory cells were also seen. Based on the histopathological features, a diagnosis of pemphigus vulgaris was made (Figure 4).



Figure 3. Intraoral picture showing erythematous areas including both the marginal as well as the attached gingiva of the lower anterior teeth indicative of desquamation. Also bleeding areas are noticed in the lower left premolar region

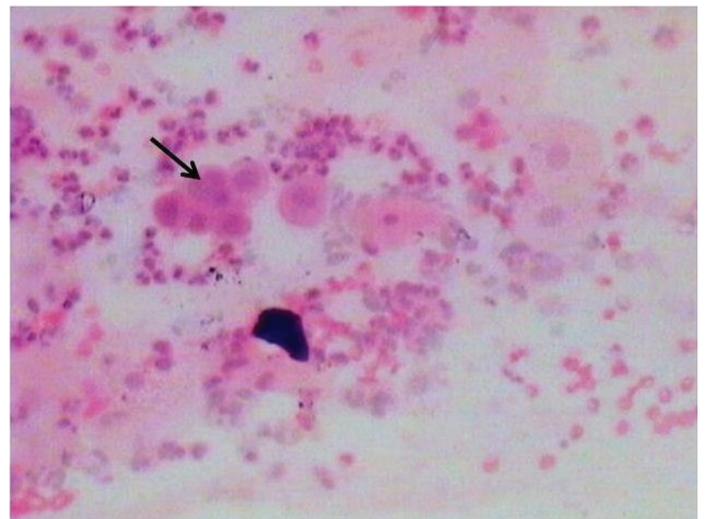


Figure 4. Histopathological picture of the smear showing round epithelial cells with enlarged and hyperchromatic nuclei suggestive of Tzanck cells with numerous inflammatory cells. (Arrow indicative of Tzanck cells)

The treatment plan comprised of oral prednisolone 40 mg/day for 2 weeks along with chlorhexidine mouthwash. Triamcinolone acetonide 0.1% ointment was also prescribed for the patient. Patient reported about the reduced pain, burning sensation and healing of the lesions when contacted through telephone and he didn't report personally as his symptoms subsided.

DISCUSSION

Pemphigus is a disease characterised by blistering of the oral mucosa and skin caused due to acantholysis (4). This process of acantholysis i.e the loss of keratinocyte to keratinocyte adhesion is caused by the circulating antibodies against the intracellular adhesion molecules (4).

Initially the disease begins as small asymptomatic blisters which are made up of thin wall, which ruptures easily resulting in painful and haemorrhagic erosions. In most cases, the oral mucosa is the first involved site. Intraorally the lesions are mostly seen in areas with constant trauma such as buccal mucosa, tongue, hard palate. The gingiva is the rarest affected site within the oral cavity. A literature search in pubmed revealed only three case reports in the past 20 years with gingival involvement as the first site of manifestation of pemphigus (1,5,6).

In our case the patient presented with bleeding gums and burning sensation. Only oral lesions along with gingival involvement were present in our patient at the time of reporting. Ulcerations can also be seen involving the conjunctiva, larynx, nasal mucosa, pharynx and oesophagus (7). Desquamative gingivitis (DG) is a clinical manifestation of the gingiva associated with desquamation loss of the gingival epithelium followed by redness and ulceration with or without blister formation (1). Nisengard and Levine proposed the following criteria for the diagnosis of DG: (1) Gingival erythema not resulting from plaque, (2) desquamation of the gingiva, (3) Other intraoral and sometimes extraoral lesions, and (4) Complaint of sore mouth, especially from spicy foods (8).

Pemphigus is generally associated with lesions that are formed in different layers of epithelium. These lesions are formed due to antibodies that are directed against target cell surface antigen. Out of all the cases of Pemphigus reported 80% of the cases are of Pemphigus vulgaris (9). The main antigen responsible for the manifestation of PV is desmoglein (Dsg) 3 (1). In PV circulating IgG autoantibodies are present against Dsg3. These bind to the desmoglein on the epithelial membrane of the cells leading to acantholysis. Acantholysis is usually seen occurring in the lower layers of stratum spinosum and as a result causes suprabasilar bulla. The bullae formed spreads involving larger areas of mucosa and skin (4,10,11).

The blisters may be few centimetres in size and contain clear fluid. Under pressure they spread through the surrounding epidermis thereby increasing in size. This is termed as indirect Nikolsky's sign. Healing process is slow without formation of scar. In the oral mucosa the bullae formed ruptures immediately because of the thin wall. After rupturing they get detached from the epithelium (11,12). In our case intact blisters were not present. Patient showed positive Nikolsky's sign. Acantholytic cells also called as Tzank cells contain round hyperchromatic nuclei

with clear perinuclear halo. The presence of these cells can be seen in cytological smear (1,13). Tzank cells were observed in present case.

A proper history taking can help the clinician in distinguishing these lesions from herpes virus infections and erythema multiforme. Pemphigus Vulgaris can be differentiated from other closely related diseases by biopsy and immunofluorescence. Biopsy should be done on intact bullae that are less than 24 hours old. The supra basilar split which is characteristic of Pemphigus vulgaris helps in differentiating it from bullous lichen planus, mucous membrane pemphigoid, and chronic ulcerative stomatitis. Indirect immunofluorescence can differentiate PV from pemphigoid. Diagnosis is confirmed by IgG and other C3 antibodies which bind to skin or mucosa causing intraepithelial blistering (14).

Treatment

An important phase in the management of PV is the early diagnosis. This enables symptomatic relief, easier control of disease using lower doses of medication and limitation of disease progression. Local and systemic corticosteroid therapy is the mainstay of PV management. Treatment consists of use of paste, ointments or mouthwash either used alone or in combination with systemic agents. Treatment usually involves 2 phases: a loading phase which brings the disease under control and a maintenance phase. Intralesional administration of corticosteroids is used for the management of persistent lesions. In case of extensive lesion systemic corticosteroids are administered. Prednisone 0.5–2 mg/kg is the recommended dose for systemic administration. If steroids are needed for longer periods of time, agents such as Azathioprine or Cyclophosphamide are added to reduce the complications of long term corticosteroid therapy (15,16). In our case patient was treated with prednisolone 40mg/day for two weeks and recalled. This report describes rare initial presentation of PV as bleeding gums and burning sensation. Thorough knowledge regarding PV is essential to all dental professionals to avoid the misdiagnosis and delayed treatment in such cases.

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