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RESEARCH ARTICLE

ORAL LESIONS – THE CLUE TO DIAGNOSIS OF THE MOST COMMON BLISTERING DISEASE

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ABSTRACT

Pemphigus Vulgaris is a chronic autoimmune intradermal blistering mucocutaneous disease that initially manifests in the form of intraoral lesions, followed by other mucous membranes and the skin. The aetiology of the disease still remains unclear but the presence of auto antibodies is consistent with an autoimmune disease. Systemic corticosteroids remain the treatment of choice but topical corticosteroids are also used for long term non-healing lesions of the oral cavity. In this article, we have discussed a case of a 60-year-old woman showing oral ulceration without skin lesions.

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INTRODUCTION

Pemphigus is a potentially life-threatening autoimmune mucocutaneous disease, commonly characterised by blistering; pemphigus vulgaris (PV) is the main variant and the one which most commonly involves the oral cavity (Alessio Gambino *et al*., 2014). The term being derived from the Greek word *Pemphix* (bubble or blister) (Crispian, 2002). Pemphigus vulgaris is rare disease, with a reported incidence of 0.1-0.5 cases per 100,000 individuals globally per year. It has slight female predominance and primarily manifests in adults during the fifth or sixth decade of life (Dagistan, 2008). Pemphigus can be classified into five major groups: Pemphigus vulgaris (PV), pemphigus foliaceus, paraneoplastic pemphigus (PNP), drug-induced pemphigus and immunoglobulin A (IgA) pemphigus. Oral lesions have been associated with only PV and PNP (Kumar *et al*., 2016). The etiology of the disease still remains unclear but the presence of autoantibodies is consistent with an autoimmune disease. The autoimmune bullous dermatoses are divided into 2 main groups: diseases of the dermoepidermal junction, that are due to abnormalities at the interface between the dermis and the epidermis (example of which is pemphigoid) and intraepithelial dermatoses, which include the various forms of pemphigus.

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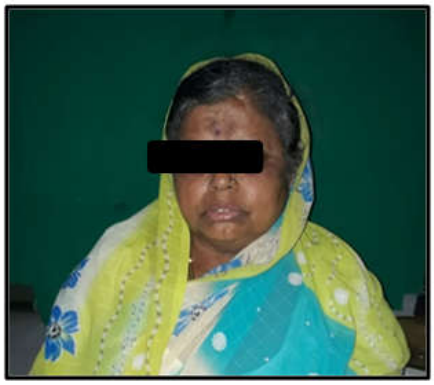
Pemphigus results from circulating immunoglobulin G (IgG) antibodies directed against desmosomes; these antibodies interfere with keratinocyte adhesion due to which acantholysis occurs and results in formation of bullae (Nadia Ben Lagha, 2005). The prognostic factors are age, time between onset of symptoms and initiation of treatment, extent of the lesions and the dose of corticosteroids required to initially control the disease (Nadia Ben Lagha, 2005). In this article, we have discussed the case of female patient with multiple chronic oral ulcers and its management.

CASE REPORT

A 60-year-old woman reported to the department of oral medicine and radiology with the chief complaint of painful ulcers in the mouth and bleeding gums since 10 months. She had similar lesions on scalp, hands and legs 10 years ago. For the treatment of lesions on scalp, hands and legs patient reported to the dermatologist, who gave the diagnosis as Pemphigus Vulgaris. All lesions healed after taking systemic corticosteroids (tablet prednisolone 100mg per day, later on dose was tapered to 40 mg per day). It took 4 years for the lesions to get healed. Patient again started taking tablet prednisolone 40mg for the oral lesions but it did not show any improvement in the lesions. So she reported to Oral Medicine and Radiology department. Personal and family history were uneventful. Patient is suffering from Type 2 diabetes mellitus for which she is taking tablet Glycomet 500 S.R. (Metformin hydrochloride sustained release tablet). History revealed that the lesions started as blisters or vesicles and quickly busted, leading to erosions.



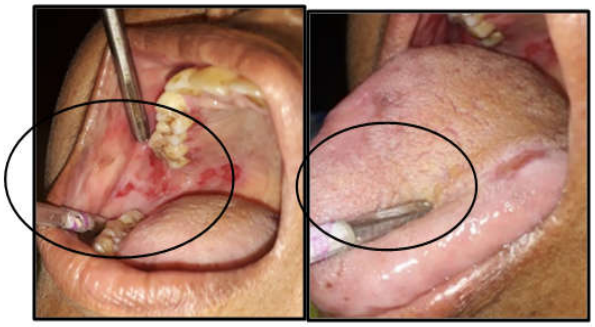
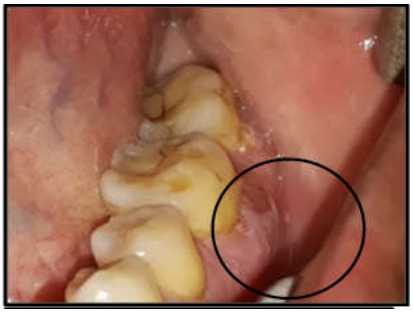
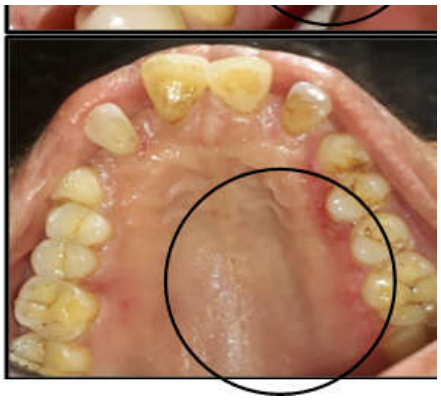
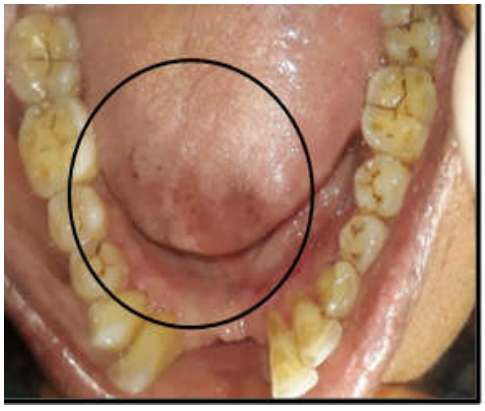


Fig 1. Clinical presentation of lesions







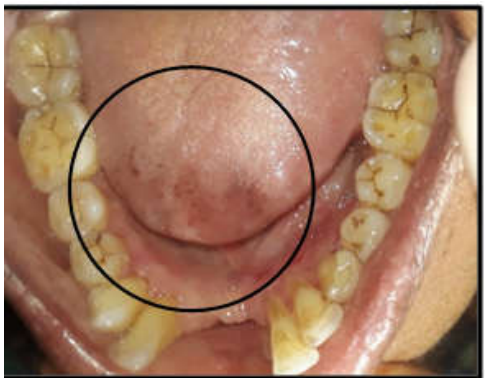


Fig 2. H and E stain-showing suprabasilar split

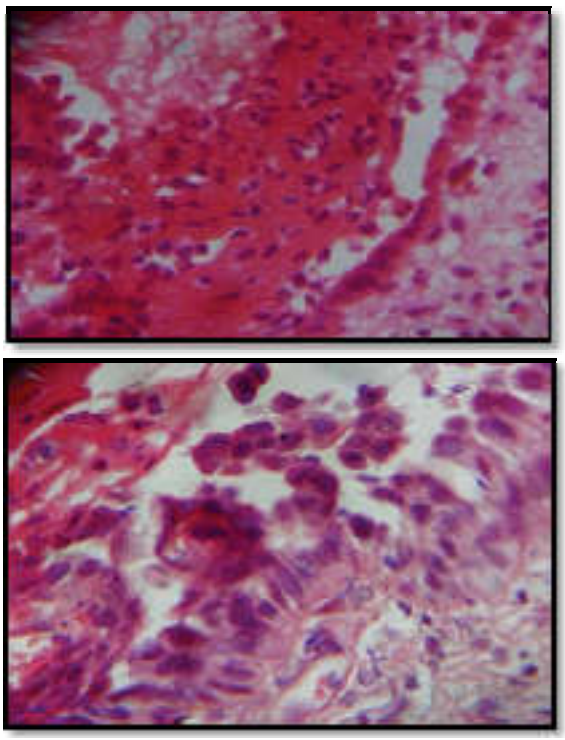


Fig. 3. H and E stain – showing Acantholytic Tzanck cells within the suprabasilar split

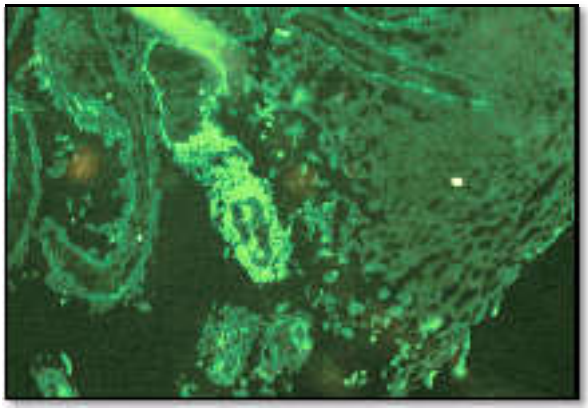


Fig 4 – Direct immunofluorescence stain showing deposition of C3 in the areas of autoantibodies in the epithelium

The lesions are painful and patient was unable to eat or drink because of the lesions. Presently, multiple red erythematous lesions were seen on hard palate, right buccal mucosa, tongue and gingival (Fig 1). The lesions were approximately 0.5 to 1cm each in diameter, roughly oval in shape with ragged borders and ulcerated surface.

On palpation the ulcers were tender, depth is shallow and showed bleeding on manipulation; bleeding on probing was positive with gingiva and inflammation was present with marginal gingiva but there was no other part of the body seen affected except the oral cavity. Light pressure was applied to intact skin but that did not show any formation of bullae (negative Nikolsky's sign). On the basis of history and clinical examination, provisional diagnosis given was pemphigus vulgaris. Differential diagnosis given was Erosive Lichen Planus, Benign Mucous Membrane Pemphigoid or Cicatricial pemphigoid and erythema multiforme. Investigations advised to the patient were complete blood count, random blood sugar level and biopsy and also Direct immunofluorescence test; and orthopantomogram were advised. Complete blood count report revealed that the patient was anaemic (Hb- 9gm%) and her random blood sugar level was slightly higher (129.1mg%). Orthopantamogram showed moderate horizontal bone loss, missing 31,32,41,42, grossly carious 27,28 and 46. Along with periapical abscess with 27,28 and 46.

Perilesional biopsy, the tissue was taken from right buccal mucosa showed ulcerated squamous epithelium and suprabasilar split (Fig 2). Also, it was positive for acantholytic cells (Tzanck cells) i.e round shape cells with prominent nuclei and the connective tissue showed, moderate chronic inflammatory cell infiltrate (Fig 3), these histopathological features are suggestive of pemphigus vulgaris. Direct immunofluorescence showed intercellular deposits of antibodies against IgG and C3 (complement), which confirmed the diagnosis of pemphigus vulgaris. After the diagnosis was confirmed, we prescribed topical application of kenacort ointment (0.1% triamcinolone); to be applied thrice daily and not eat or drink at least an hour after the application. Patient showed marked reduction in the size of lesions at the 15th day of follow up. Patient was given iron supplements to correct the haemoglobin levels. Scaling and root planning was done and endodontic treatment was advised with 27,28 and 46.Also replacement of missing teeth was advised to the patient.

DISCUSSION

In 70 to 90% of the cases, the first signs of disease appear on the oral mucosa. Although the lesions can be located anywhere within the oral cavity, but most often found in areas that are subjected to frictional trauma, such as the cheek mucosa, tongue, palate, and lower lip (Dagistan, 2008). Where as in the present case the skin was affected initially and later on oral manifestations were encounter. The disease may affect other mucous membranes of the body such as the conjunctiva, nasal mucosa, pharynx, larynx, esophagus and genital mucosa, as well as the skin where intact blisters are seen commonly (Tan, 2006). Autoimmune blistering skin diseases show circulating auto antibodies which are directed against the epidermal keratinocyte cell surface or the epidermal basement membrane zone (BMZ), that in turn induces separation either between epidermal keratinocytes or at the dermoepidermal junction (Masayuki, 2010; Takashi Hashimoto, 2003). The most common disease showing antibodies against antikeratinocyte cellsurface is pemphigus and bullous pemphigoidis the representative disease among diseases that develop sub epidermal blister (Takashi Hashimoto, 2003). The initial lesions are often insidious and localized. The mouth is affected by persistent, painful ulcers and a burning sensation, which affects the appetite (Vaillant, 1999). The skin becomes affected several weeks or months after the mucosal lesions appear, with the appearance of flaccid blisters filled with clear fluid. These fragile blisters are easily broken, which leaves behind erosions surrounded by epidermal rings. Putting pressure on healthy skin causes either a bulla or an erosion; this effect is known as Nikolsky's sign (Vaillant, 1999; Bickle, 2002; Cotell, 2000). This sign, although highly suggestive of pemphigus, is not specific and may be absent. Histologically, pemphigus vulgaris is characterized by intraepidermal cleft, with a basal cellular layer forming the base of the blister. Direct immunofluorescence reveals a homogeneous deposit of IgG and C3 in the intercellular substance (Thivolet, 2001).

The diagnosis is usually based on the oral manifestations, while confirmation is provided by the histological findings, which show the presence of intraepithelial blisters, acantholysis, and Tzanck cells (Femiano, 2002). Direct immunofluorescence stain of the fresh lesion specimens shows IgG or IgM and complement fragments in the intercellular space (Femiano *et al*., 2002). In our case, a biopsy of the intraoral lesions was obtained. The specimen sections were stained with Hematoxylin-Eosin, and the principal histological characteristics were evaluated. Direct immunofluorescence studies were also carried out. Local and systemic corticosteroid therapy is usually employed in management of Pemphigus Vulgaris (Crispian, 2002; Gregoriou *et al*., 2015). The main aim of treatment is to induce disease remission. This should be followed by a period of maintenance treatment (Darling, 2006). Drug doses should be tapered slowly and patients should remain under regular follow-up while they remain on treatment (Erik, 1999). Ultimately, treatment may be withdrawn and there should be prolonged clinical remission.

CONCLUSION

Pemphigus vulgaris usually shows the oral signs first, it is an opportunity for a dentist to diagnose it correctly. Early diagnosed cases of pemphigus can be managed easily. And if there appear the extra-oral symptoms, prompt referral to the specialist should be done.

Conflicts of interest: The authors report no conflicts of interest related to this study.

Glossary of Abbreviations

PV – Pemphigus Vulgaris
PNP – Paraneoplastic pemphigus
IgG – Immunoglobulin G
C3 – Complement 3

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