Case report

Role of oral physician in the management of pemphigus vulgaris- A geriatric case report

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ABSTRACT

Pemphigus is a blister forming disease of both mucosa and skin of autoimmune origin. Most commonly seen in old age individuals and oral lesions precede the skin lesions in most cases. This is a report of a unique case of pemphigus vulgaris in an elderly male patient where oral lesions started initially followed by skin lesions. It was provisionally diagnosed by our oral medicine specialists on the very first visit and confirmed after a biopsy. The patient was treated successfully with systemic steroids and azathioprine with reduction in dose after 3 weeks. The healing was satisfactory and the patient is under follow-up.

Keywords: Pemphigus; oral lesions; immunofluorescence.

INTRODUCTION

Pemphigus vulgaris (PV) is said to be an autoimmune chronic blister forming disease of skin and mucosa in which the immune system of the body produces autoantibodies against the specific protein molecules of the desmosomal adhesion complex leading to the intraepidermal or intraepithelial blister formation (1). The word 'pemphigus' has originated from a Greek word called pemphix meaning bubble or a blister and the word 'vulgaris' has originated from a Latin word called vulgaris meaning common (2). The overall prevalence rate of this disease is 0.1 to 0.5% per one million population in a year with a much greater prevalence in the ethnic groups like Ashkenazi Jews, Japanese, and Mediterranean ancestry (3). The pathology involves formation of circulating immunoglobulin G autoantibodies against the desmosomal adhesion molecules namely cadherins and desmoglein 1 and 3 present in the epithelium. Desmoglein 1(dsg-1) is associated with the epithelial cells of skin, while desmoglein 3 (dsg-3) is associated with the cells of oral mucosa (4). There are clinically five different types of pemphigus namely P. vulgaris, P. foliaceus, Paraneoplastic pemphigus, IgA pemphigus and drug-induced pemphigus. The oral lesions are common only with P. vulgaris and Paraneoplastic Pemphigus (5). Lesions might occur on any intraoral location, but the buccal mucosa is found to be the commonly affected one (6). Desquamative gingivitis is a very common feature of this disease while involving the mucosa (7). The common age predilection is 4th and 6th decades of life, with a male to female ratio of 1:2 (8). The significance of this case report is that the oral lesions presented first followed by skin lesions and early medical intervention was carried out. Direct immunofluorescence test was done for this case which showed typical features.

Case history

A 74 year old male patient visited the department of oral medicine and radiology, Adhiparasakthi Dental College and Hospital with the chief complaint of painful ulcerations and burning sensation of the mouth and lips for the past three months. Patient gave a history of difficulty in chewing spicy and solid food. The medical and personal history of the patient were non-contributory. On extraoral examination, depigmented areas with multiple ulcerations with black crusts were seen over the scalp, shoulder and chest with positive Nikolsky's sign (Fig. 1). On intraoral examination, there were multiple large irregular ulcerations seen over the buccal mucosa, labial mucosa, lower vestibule, palate and ventral surface of the tongue (Fig. 2). The ulcers showed irregular borders with erythematous bases and were covered by yellowish slough. On palpation, there were bleeding spots and lesions that were severely tender. Based on the age of the patient and clinical appearance of the lesions, a provisional diagnosis of pemphigus vulgaris was considered. Blood investigations like complete blood count, urine analysis, liver function test, blood sugar analysis, Erythrocyte sedimentation rate were done and found to be normal.

Fig. 1: Skin lesions seen over the axilla, scalp and shoulder

Fig. 2: Oral lesions seen over the labial mucosa, tongue and buccal mucosa

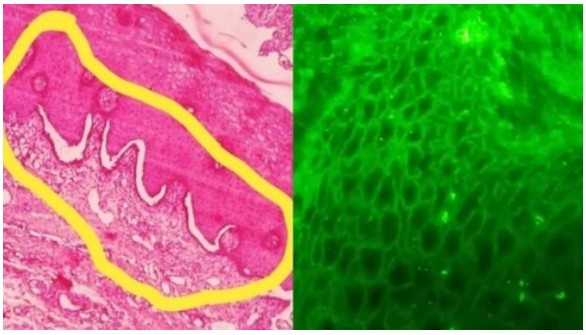

Fig. 3: Histopathological picture showing suprabasilar split and immunofluorescence showing fish net appearance

Fig. 4: Healing of oral lesions appreciated three weeks after initiating the therapy

Incisional biopsy was done from left posterior buccal mucosa region and histopathological examination revealed intraepithelial/suprabasilar split, tombstone appearance of basal layer cells and clumps of acantholytic/tzanck cells within the split. Direct immunofluorescence revealed deposition of IgG around the epithelial cell surface showing the typical fishnet appearance (Fig. 3). The microscopic findings were consistent with pemphigus vulgaris. The patient was treated with the following systemic drugs: Prednisolone 40 mg once daily in the morning for 10 days followed by 30 mg once daily in the morning for next two weeks, Azathioprine 50 mg once daily in the afternoon for 24 days and Pantoprazole 40 mg once daily in the morning before food for 24 days. Topical Clobetasol propionate solution was prescribed for skin lesions. There was a considerable amount of healing after 3 weeks and the patient was kept under follow-up (Fig. 4).

DISCUSSION

Pemphigus is a life-threatening condition which affects both the mucous membrane and skin, clinically presented as vesicle formation and histologically by intraepithelial clefting and acantholysis (9). The classical clinical appearance of pemphigus is thin-walled bullae arising on skin or mucosa or both. The bullae tend to be very fragile, break easily and leave behind denuded surfaces of variable sizes which tend to extend peripherally due to breaking of the epithelium from the underneath dermis. In majority cases of pemphigus vulgaris, the oral mucosa is involved prior to the skin (10). In the present case also, patients had come for the complaint of oral ulcerations with severe pain and burning sensation where the mucosal clefts extended from the periphery. In the oral cavity, the lesions most commonly arise in areas that are exposed to constant masticatory occlusal trauma such as buccal mucosa, followed by palate and gingiva. It can also affect other mucosal surfaces like larynx, pharynx, oesophagus, and conjunctiva. Pemphigus lesions show a positive 'Nikolsky's sign' which is the extension of the lesion along the periphery when pressure is applied tangentially on the lesion or perilesional area using index or thumb finger (3). Nikolsky's sign was positive in our case. Incisional biopsy is best carried out on intact vesicles or bullae which are less than 24 hours old. The biopsy tissue must be taken from the advancing edge of the lesion in order to the area of characteristic suprabasilar acantholysis observed by the oral pathologist. There was no intact bulla identified in our case, at the time of diagnosis and hence the advancing edge of the ulcer was taken for biopsy. Supra basal split seen in pemphigus vulgaris helps to differentiate it histopathologically from other subepithelial blister forming diseases like mucous membrane pemphigoid, bullous lichen planus and chronic ulcerative stomatitis which was consistent with our case as well. Indirect immunofluorescence is helpful in further differentiating pemphigus from pemphigoid and other blister forming diseases and is helpful in following the progress of the disease. Confirmatory diagnosis is made by the characteristic deposition of IgG and C3 antibodies at the cell surface of perilesional tissue (11). The present case showed IgG deposition and

classical fishnet appearance in direct immunofluorescence confirming the diagnosis. The current management of pemphigus vulgaris includes topical and systemic steroids in combination with immunosuppressants such as azathioprine or mycophenolate which was followed in our case (12) But the use of high doses of systemic steroids has been associated with multiple complications such as osteoporosis and hyperglycemia (13). Treatment can also be augmented with IV immunoglobulin or plasmapheresis, aimed at reducing circulating autoantibodies. Treatment with rituximab, an anti-CD20 antibody, is shown to be effective in many cases (12). Pulse therapy for pemphigus such as IV dexamethasone and cyclophosphamide pulse, has been successful in the recalcitrant cases (3).

CONCLUSION

Dentists play a very important role in diagnosing pemphigus lesions since many of the patients first acquire oral lesions followed by skin lesions. Early identification can significantly improve the effectiveness of treatment, reduce complications of long-term steroid therapy, improve quality of living, and reduce morbidity associated with the disease.

CONFLICT OF INTEREST

The authors declare no conflicts of interest.

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