How to Cite:
Bathija, V. S., Pandey, P., Jain, A., Chhabra, S. K., Deshpande, P., & Bhati, C. A. (2022). Ear nose throat manifestation of autoimmune blistering disorder. *International Journal of Health Sciences*, *6*(S6), 9766–9772. https://doi.org/10.53730/ijhs.v6nS6.12606

Ear nose throat manifestation of autoimmune blistering disorder

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Abstract---Introduction: Autoimmune blistering diseases (AIBD) are rare blistering disorders of mucous membrane and skin caused by autoantibodies against specific cells. Ear, nose and throat (ENT) examination is must and should be a routine procedure to evaluate these patients for mucosal involvement as mucosal involvement significantly increases morbidity and mortality. These patients should be asked for oropharyngeal, ear, nose and throat symptoms and should be evaluated for the same by direct and endoscopic examination. Materials and Methods: 60 patients were taken in total, who had mucosal involvement. Ear, nose and throat (ENT) examination was done. Every case of mucosal involvement was promptly enquired about oropharyngeal, ear, nose and throat symptoms. All patients were clinically seen for ENT and

International Journal of Health Sciences ISSN 2550-6978 E-ISSN 2550-696X © 2022.
Manuscript submitted: 9 May 2022, Manuscript revised: 18 July 2022, Accepted for publication: 27 August 2022

dermatological manifestations by direct and endoscopic examination, cutaneous examination even in absence of symptoms. Result: Out of 60 patients, 42 were pemphigus vulgaris, 3 were pemphigus foliaceus, 8 were paraneoplastic pemphigus, 7 were mucous membrane pemphigoid. Conclusion: It is very important to evaluate every patient of mucosal lesion by both ENT and dermatologist for the better management of the patient to reduce mortality and morbidity.

Keywords---autoimmune blistering diseases, mucosal involvement, ear nose throat.

Introduction

Autoimmune blistering diseases (AIBD) are a bunch of rare blistering disorders of the mucous membrane and skin because of autoantibodies directed against specific cells in our own body causing morbidity and mortality. AIBD be divided into 2 subgroups: (1) subepidermal blistering diseases (SEBD) (2)intraepidermal blistering diseases (IEBD) (1). Bullous pemphigoid is chief AIBD of the subepidermal blistering group. It's the foremost common AIBD in developed countries and seen in people of age group >70 years. It affects mucosal membranes ,it is not as common as PV [2]. On the other hand, mucous membrane pemphigoid (MMP) mainly affects the mucosal surfaces of oropharynx, larynx, eyes , nose, oesophagus and genitals and leads to tissue destruction and functional limitations due to chronic mucosal inflammation and scarring [3].

IEBD includes pemphigus foliaceus (pf), pemphigus vulgaris (pv) and paraneoplastic pemphigus (pnp) . Pv is that the major type and it's most typically seen between the ages group of 40 and 50 years. It always starts as erosions within the oral mucosa, and continues with blistering and erosive skin lesions. Other involved areas include the nasal mucosa , pharynx , larynx and oesophagus. (4). Pnp is a paraneoplastic blistering disease characterized by cheilitis and ulcerative stomatitis, persisting painful erosions of mucosa and cutaneous polymorphic involvement .it always presents with severe mucosal involvement including the pharyngeal, laryngeal, ocular and genital mucosal surfaces. Its prognosis is poor thanks to accompanying malignancy. On the opposite hand, pf doesn't affect the mucosal surfaces [5].

Materials and Method

60 patients were taken in total, who had mucosal involvement. Every case of mucosal invlovement was romptly enquired about oropharyngeal, ear, nose and throat symptoms. All patients were clinically evaluated for ent and dermatological manifestations by direct and endoscopic examination, cutaneous examination even in absence of symptoms.[6] In these cases the ent involvement was studied and found to be much more than expected, especially in pv, [5-11]. Our study additionally suggested that the severity of ent involvement in pv was significantly associated with symptoms and endoscopic finding in cases of severe disease (pemphigus disease area index (pdai)> 16). However apart from severe scarring form of aibd even mild mucosal involvement can cause discomfort, bleeding and

erosions on mucosa. Therefore every ent surgeons should do a proper examination and procedural approach in care of these patients (7)

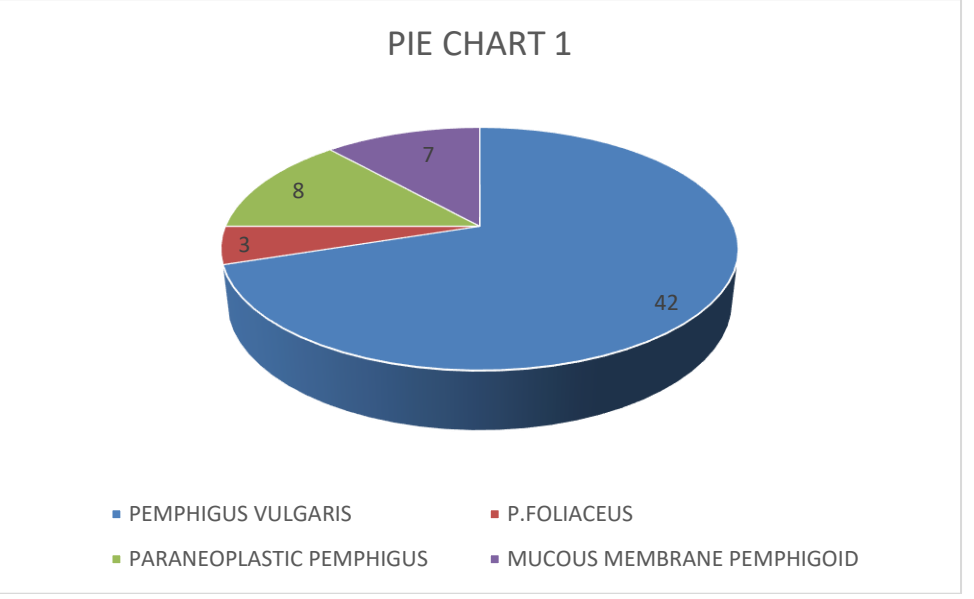
- Inform the patients and his /her families about the disease, its nature, its evolution available treatments, side effects of treatment and complications.
- Inform the patients about follow up to monitor disease activity, treatment tolerance and any side effects.
- Inform the patients about triggers of disease such as drugs, physical trauma and operations
- Although there is insufficient evidence in terms of restrictions about diet, patients can be informed about avoiding certain foods (such as garlic, onion,) which are thought to be triggers of aid (especially for pemphigus) (15)
- Patients should be vaccinated against seasonal influenza, pneumococci most appropriately before any therapy however, if patients are on immunosuppressive drugs like high dose corticosteroids, azathioprine, rituximab), live attenuated vaccines are contraindicated. (9)
- Steps taken by practitioners for ENT examination are:
- Ask about recent drug-intake as it is well-known that drugs like d-penicillamine, angiotensin-converting enzyme inhibitors, beta-blockers, etc.) Can trigger aibd (15,16).
- Check and document the patient's general condition and comorbidities.
- Examine all mucous membranes to determine the degree of mucosal damage and functional impairment like dysphagia, dysphonia, hoarseness (10)
- Remember not cause any mechanical trauma by direct pressure and/or friction on the skin and mucosal surfaces, as trauma can create new blisters and erosions, especially in active disease due to the nikolsky phenomenon.
- Use flexible nasogastric tubes and endoscopes and the ones with lubrication to decrease the trauma risk.
- Drain new blisters by a sterile needle and overlying mucosa should be left intact to provide a biologic cover of the erosion.
- A dermatologist with a good knowledge of AIBD should also be involved in the process of diagnosis and the treatment plan for these patients. (11)
- Document all therapies (topical and systemic)
- Explain risks of the operation to the patients and his/her care takers in detail and in simple language
- Patients should continue their usual skin care habits, such as emollients and antiseptic washes, and topical therapies during, before and after surgery.
- Lubricate each and every equipment like laryngoscopes, endotracheal and nasogastric tubes (12)
- Electrocardiogram and electrocautery electrodes should be done without adhesive tapes especially in patients with active disease
- Patients with erosions have an increased risk of fluid and heat loss during the surgery therefore operation theatre should have all the facilities needed for the same

- There is an increased risk of secondary infection and pain after surgery in patients with aid due to direct (erosions) and indirect effects (usage of immunosuppressive medications, anaemia, malnutrition) of the disease. For which proper antibiotic cover should be given (13)

Result

Table 1
Frequency of various oral/oropharyngeal mucosal lesions in the study subjects

Lesions	Frequency (n=113)	Percentage (%)
Pemphigus vulgaris	42	71
Pemphigus foliaceus	3	5
Paraneoplastic pemphigus	08	13.4
Mucous membrane pemphigoid	07	10.6



Investigations and Diagnosis

Biopsy, clinical features, light microscopy, positive direct immunofluorescence microscopy (dif) findings and positive specific enzyme-linked immunosorbent assay (ELISA) autoantibodies are necessary for the diagnosis of aibd [2-3]. Specimens should be taken from early bullae arising (from the edge of the lesion) on erythematous or normal mucosa or skin and placed in 10% formalin (8)

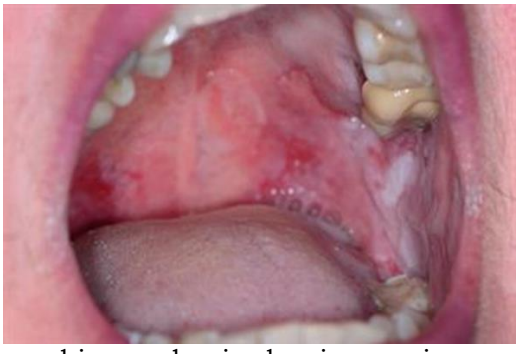


Figure 1. Patient of pemphigus vulgaris showing erosions and ulceration over left side of buccal cavity

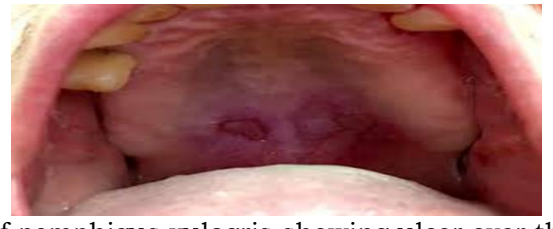


Figure 2. Patient of pemphigus vulagris showing ulcer over the posterior side of hard palate

Figure 3. Patient of mucous membrane pemphigoid showing a large ulcer over the upper gums

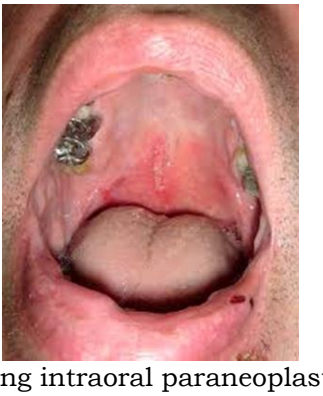


Figure 4. Showing intraoral paraneoplastic pemphigus

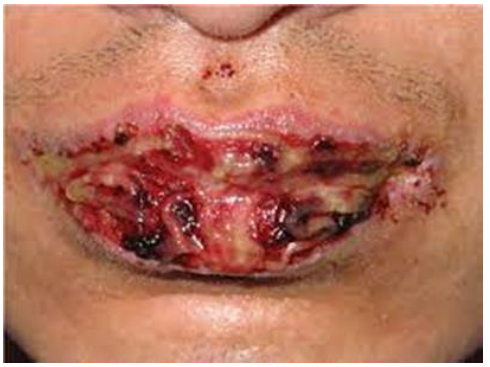


Figure 5. Patient of paraneoplastic pemphigus showing lips involvement

Discussion

As seen from our study we concluded that in patients coming with complaints of mucosal involvement should be thoroughly examined as it is missed. Hence examining these patients should be a combined approach of both the ENT specialist and a dermatologist , for better management of the patient

Conclusion

Every ENT specialist should be aware of AIBD so they can give proper care to the patient in association with a dermatologist. Early diagnosis which results in improved management and reduces morbidity and mortality.

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