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Original Research Article

Profile of Ocular Manifestations in Pemphigus Pateints

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Abstract

Purpose: To study the ocular manifestations in pemphigus and bullous pemphigoid (BP) patients.

Methods: Cross-sectional Observational Study was conducted on patients diagnosed with all types of pemphigus attending Outpatient Department at Basaveshwara Hospital, Chitradurga from December 2021 to September 2022.

Exclusion criteria being Patients on chronic antiglaucoma medications, Pre-existing dry eye or xerophthalmic patients, Patients on long-term topical steroids usage in eye, Patients who underwent previous ocular surgeries, Patients with previous corneal scar.

Results: A total of 50 patients of pemphigus were examined. Out of 50, 12 patients had ocular involvement. Duration of disease of all patients was more than 6 months. Redness and burning sensation was the most common symptom. The most common ocular signs, being conjunctival hyperemia, and moderate dry eye. Ocular manifestations are more common with bullous pemphigoid patients than other types.

Conclusions: High occurrence of dry eye and conjunctival scarring can lead to visual morbidity. Thus ocular examination of pemphigus patients prevents long-term sequelae and complications.

Keywords: Pemphigus, Autoimmune Disease, Dry Eye, Cicatrisation, Hyperemia.

INTRODUCTION

Pemphigus are group of autoimmune blistering illnesses which causes acantholysis and blistering of the skin and mucous membranes which are brought on by autoantibodies against desmosomal proteins that serve as intercellular adhesion molecules.[1,2] The desmosomal antigens desmoglein 1 and 3 are among the autoantibodies that contribute to the etiopathogenisis of this illness.[3] The glycoprotein desmoglein commonly express systemic findings in skin, mucosa, larynx, oesophagus, cervix, anus and vulva, the ocular findings of the disease like non-cicatrizing conjunctivitis, erosions in conjunctiva and lid margin.[4]

Autoantibodies against antigens expressed in the conjunctival epithelium's basal cells are released in ocular PV (OPV), diminishing in the suprabasal layers. These desmosomal cadherin's, desmoglein 1 and 3, are the pemphigus antigens. It is thought that the build-up of autoantibodies against intercellular adhesion molecules causes early conjunctival inflammation. These

autoantibodies also triggers complement to activate, which induces inflammation and disrupting the basement membrane Zone (BMZ).[5,6]

Intraepithelial activity is observed in ocular pemphigus vulgaris, but autoimmune activity is evident in linear immunoglobulin. Sub epithelial disease sites include those for mucous membrane pemphigoid and epidermolysis bullosa acquisita. For a blister to form, inflammatory cells and complement must be present.[7] Acantholysis and blister development are caused by autoantibodies in pemphigus vulgaris. Early detection of ocular symptoms in these disorders is essential due to the increased risk of blindness with a delay in diagnosis and care.[8] The study was conducted since an accurate identification of these autoimmune bullous illnesses is necessary for effective treatment.

MATERIALS AND METHODS

This cross sectional observational study conducted in department of ophthalmology at Basaveshwara Medical College and Hospital, Chitradurga, Karnataka from December 2021 to September 2022 included after obtaining informed consent form and Institutional Ethics Committee (IEC) clearance.

A total of 50 patients of pemphigus were examined. Out of 50, 12 patients had ocular involvement Patients underwent ophthalmic examination including Visual acuity both for distant and near, best corrected visual acuity assessed by snellens chart. Torch light and slit lamp bio microscopy, Conjunctival fluorescein staining and corneal staining, Tear film break up test, Schirmer test.

Inclusion Criteria: Patients whoever diagnosed with Pemphigus aged between 30 to 70 years. Exclusion Criteria: patients on long-term anti-glaucoma drugs, individuals with xerophthalmia or prior dry eye patients with chronic topical steroid use in the eyes, Patients who had previously undergone pterygium surgery for their eyes, Patients who underwent lid surgery, corneal scarring, or who are unwilling to participate in the trial are excluded..

Statistical Analysis: All the statistical analysis was done by using Microsoft Excel Spread Sheets.

RESULTS

This cross sectional observational study recruited 50 patients diagnosed with pemphigus, out of which 12 patients had ocular manifestations. All patients had duration of disease more than 6 months. Unilateral involvement was more common compared to bilateral. Females were more affected compared to males. Among 12 patients 8 patients were diagnosed with bullous pemphogoid, and 4 patients with pemphigus vulgaris. Rednes, dryness and photophobia were most common symptom. All the patients had Conjunctival hyperaemia, six patients had papillae, eight patients had Blepharitis, two patients had Symblepharon, one patient had corneal opacities and Schirmer Test out of 12 patients six patients had mild and six had moderate dryness. seven patients had abnormal (5,8 and 10 seconds). TBUT-tear film break up time - five patients shown normal (10, 15, 20 and 25 seconds) and all the symptoms and signs are depicted in the table 1, 2, 3, 4.

Table 1: Age and gender distribution of the study subjects

Patient Details	C1	C2	C3	C4	C5	C6	C7	C8	C9	C10	C11	C12
Age	48	53	56	49	45	53	54	43	45	47	50	52
Gender	F	F	F	F	F	F	M	F	M	M	F	F

C- Case; M- Males; F-Females

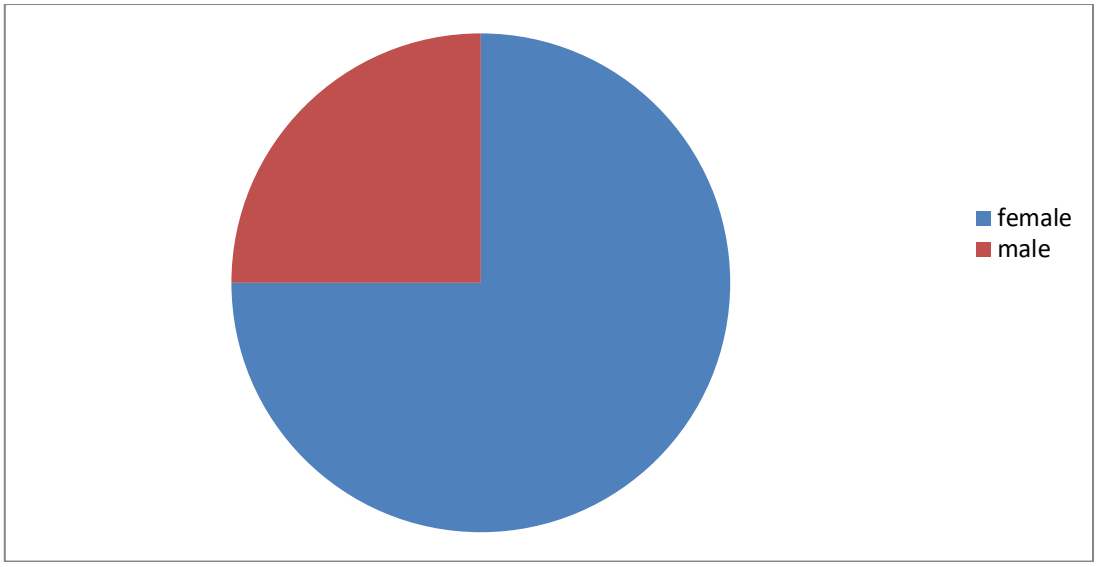

Figure 1: Gender Distribution

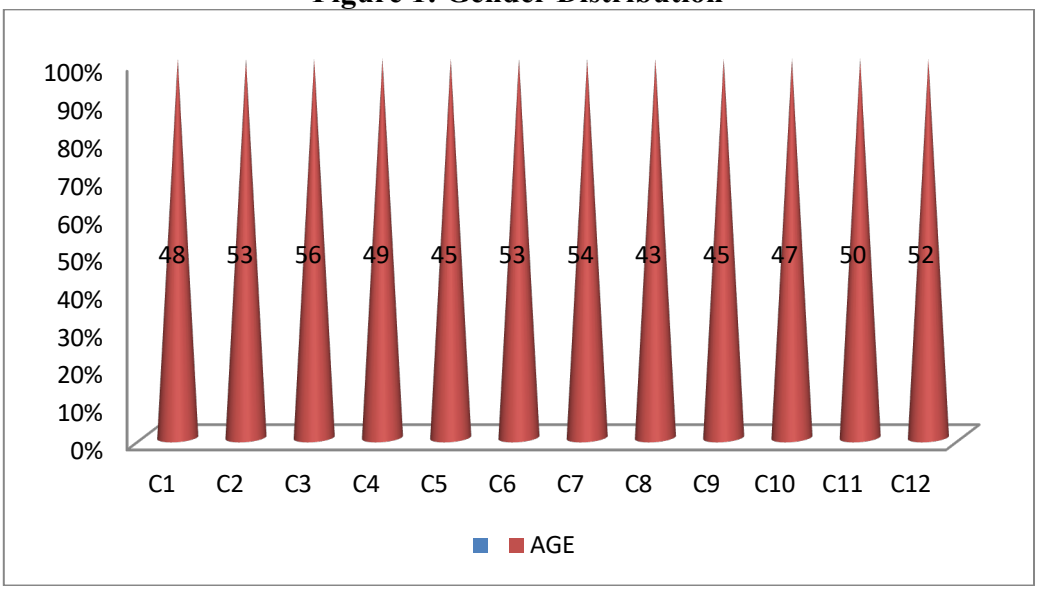

Figure 2: Age distribution of the study subjects

Table 2: History and symptoms of the study subjects

Symptoms and History	C1	C2	C3	C4	C5	C6	C7	C8	C9	C10	C11	C12
Redness	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes
Dryness	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	No	No	No
Itching/stinging	N0	No	Yes	Yes	Yes	No	No	No	Yes	Yes		Yes
Blurring of vision	No	No	No	No	No	No	No	No	No	No	Yes	No
U/L OR B/L	U/L-R	U/L-R	U/L-R	U/L-L	B/L-	B/L-	U/L-L	U/L-R	U/L-R	B/L	U/L - L	B/L
Type of pemphigus	BP	BP	PV	BP	PV	BP	BP	PV	BP	BP	BP	PV
Duration of pemphigus	1YR	6m	7m	1yr	9m	6m	6m	7m	1yr	10m	9 M	1YR
Pain	NO	YES	NO	NO	NO	YES	YES	YES	NO	NO	NO	NO
Photophobia	YES	YES	YES	YES	NO	YES	NO	NO	YES	YES	NO	NO

U/L-Unilateral, B/L- Bilateral. BP-Bullous Pemphigoid, PV – PemphigusVulgaris,m-Months,Yr-Year

Table 3: Signs distribution of the study subjects

Signs	C1	C2	C3	C4	C5	C6	C7	C8	C9	C10	C11	C12
Conjuctival hyperemia	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes
Papillae	No	No	Yes	Yes	No	No	No	Yes	Yes	No	Yes	Yes
Blepharitis	Yes	No	No	Yes	Yes	No	Yes	Yes	Yes	Yes	No	Yes
Symblepharon	Yes	No	No	No	No	No	No	No	Yes	No	No	No
Schirmer Test	Mod	Mil	Mil	Mod	Mod	Mil	Mil	Mil	Mod	Mod	Mil	Mod
Corneal Opacities	No	No	No	No	No	No	No	No	Yes peripheral at 6 clock macula grade of 2x2mm in C9	No	No	No

MIL-Mild,MOD-Moderate

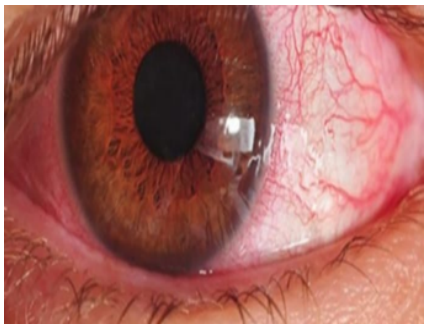

Figure 3: Hyperemia

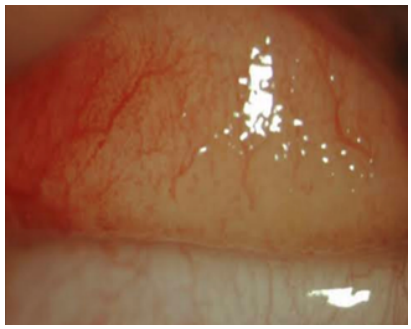

Figure 4: Papillae

Table 4: TBUT distribution of the study subjects

TBUT	C1	C2	C3	C4	C5	C6	C7	C8	C9	C10	C11	C12
	AN 10 sec	N 20 sec	N 15 sec	AN 8 sec	AN 20 sec	AN 10 sec	N 20 sec	N 15 sec	AN 5 sec	AN 10 sec	N 25 sec	AN 5 sec

AN-abnormal,N-normal, Sec- Seconds

DISCUSSION

The muco-cutaneous tissue of the body is affected by Pemphigus vulgaris (PV), a group of autoimmune diseases in which the epithelium separates from the basal layer at a point slightly above the basal layer. This causes huge stretches of exposed tissue to the elements. Drug-induced PV, mucous membrane PV, mucocutaneous PV, pemphigus vegetans of Neuman, and pemphigus vegetans of Hallopeau are some of the different types of pemphigus vulgaris. It's crucial to distinguish between PV and Bullous pemphigoid.[9,10] In PV, the desmosomes that hold the epithelial cells together are affected, whereas the hemi-desmosome, which connects the epithelium to the dermis, is affected in Bullous pemphigoid. Although the prevalence and clinical features of ocular involvement in PV have been documented, however they

are not well understood.[11]Autoimmune-induced conjunctival inflammation is the first stage of ocular involvement. Autoantibodies and complement are deposited in the basement membrane zone or intracellular substance of the conjunctival epithelium, activating inflammatory cells that then release cytokines that remodel the extracellular matrix and induce progressive fibrosis of the cornea, conjunctiva, or eyelid. [12]Corneal epithelial defects may progress into secondary bacterial infections, ulceration and scarring. Conjunctival shrinkage, corneal neovascularization, and opacification of the ocular surface, which can result in reduced vision or total blindness, are the hallmarks of end-stage ocular involvement.[13,14]Clinical symptoms of autoimmune blistering eye illnesses are brought on by the conjunctiva's increasing cicatrization. Dry eyes are caused by destruction of the conjunctival goblet cells, obstruction of the meibomian gland orifice, and blockage of the lacrimal gland ductules. The ocular surface is more susceptible to harm from the outside when the tear film is inadequate or unstable.[15] According to a study by GhazalaButt et al, 32 (32%) of the patients had ocular involvement, with conjunctival redness being the most prevalent symptom, occurring in 18 patients (18%). In 17 cases (17%), eyelid skin involvement was also quite common. 15% of individuals had conjunctival discharge. Only 4 (4% of patients) showed signs of visual haze. About 21 patients (21%) reported experiencing pain, which was extremely common.[16] In a research that involved 147 patients, Daoud et al. revealed that 7% of cases have ocular involvement. Bilateral conjunctivitis and ulcers along the lid edge were found, according to the authors. The average time between the commencement of PV and the involvement of the eyes was 20 months, and the average duration of the eye disease was 12 months. According to the study, patients fully recovered from conjunctival illness with no long-term consequences.[17]According to a study by Jeremy C.K. Tanet et.al colleagues, blepharitis (68.1%), conjunctival hyperemia (22.7%), and limbal widening (18.2%) were the most prevalent ocular symptoms in both groups.[18] In their cohort of 15 patients with PV, Chirinos-Saldana et al study revealed subconjunctival scarring, symblepharon, trichiasis and entropion, corneal opacities, and ankyloblepharon, showing the existence of long-term cicatricial alterations brought on by ocular disease..[19] According to Akhyani et al., conjunctivitis and erosion of the palpebral conjunctiva were the most prevalent findings in their research of 103 PV patients with ocular involvement, which occurred in 16.5% of the patients.[20] Our study also demonstrated conjuctival hyperemia and dryness as most common manifestations which is comparable to previous studies.

Limitations of present study include small sample size and lack of follow up.

CONCLUSION

According to the findings of the current study, ocular involvement is frequently present in pemphigus patients, therefore ocular examinations and subsequent patient care can decrease visual morbidity and subsequent complications including blindness in pemphigus patients who have ocular involvement.

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