

FREQUENCY AND COMPLICATIONS OF OCULAR PEMPHIGIOD IN N.W.F.P.

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ABSTRACT

Objective: To study the frequency of "Ocular Pemphigoid" in the population of NWFP and to study the mode of presentation and presence of complications

Material and Methods: It is a retrospective case study conducted in the department of Ophthalmology, Hayatabad Medical Complex, during the period from January 1998 to December 2001.

Results: During this period six cases were identified with the proven diagnosis of ocular cicatricial pemphigoid (OCP). The incidence of OCP in the admitted patients was 1:2427. Fifty percent of them were male & 50% female. The mean age of presentation was 56.6 years. Sixty seven percent of the patients had systemic mucous membrane involvement. Only 17% had skin involvement. All of them were previously treated as trachoma.

Conclusion: This paper emphasizes the presence of ocular pemphigoid and should be kept in mind when dealing with the patients of trichiasis, entropion and dry eyes.

Key words: Ocular pemphigoid, Trichiasis, Dry Eyes, essential shrinkage of conjunctiva.

INTRODUCTION

Ocular pemphigoid is also known as benign mucous membrane pemphigoid and essential shrinkage of conjunctiva.¹ It is a rare, bilateral (one eye may be involved first), chronic blistering conjunctival disease of unknown etiology.² It affects the eyes

particularly but usually also involves other mucous membranes and less commonly the skin. In this condition the essential and destructive process is not the formation of blisters, but the invasion of the sub mucous tissue by new-formed connective tissue, which subsequently contract and cause complications.³ As the disease is virtually blinding and can also cause systemic

complications and that appropriate therapy can retard the progression of the disease, the correct diagnosis is of immense importance. The aim of the study was to ascertain the incidence of ocular pemphigoid and also to see the rate and severity of the complications in our population.

MATERIAL AND METHODS

It is a retrospective study conducted in the department of ophthalmology Hayatabad Medical Complex during the period January 1998 to December 2001. Records of all the patients were analyzed with a proven diagnosis of ocular pemphigoid. The following parameters were recorded: Age, sex, address, and mode of presentation. Visual acuity and condition of the lids, adnexa, fornices and anterior segment examination with slit lamp biomicroscopy were also recorded. Systemic examination including examination of mucous membrane of mouth, nose, throat and skin examination was also recorded. History was also obtained regarding the previous diagnosis and treatment modalities.

RESULTS

During the period January 1998 to December 2001, a total of 14560 patients were admitted to the department of Ophthalmology Hayatabad Medical Complex Peshawar. Six cases were identified as having proven diagnosis of ocular pemphigoid (Incidence 1: 2427). All of the patients have involvement of both eyes. Out of 6 cases, 3 (50%) were males and 3 (50%) were females. The mean age of presentation was 56.6 years (Range 45-65 years). The details of the patients are given in Table No. 1. All of the cases were previously treated as trachoma. All of them had received local and systemic antibiotics for trachoma. One patient presented with advanced stage of ocular pemphigoid with

total keratinization and epidermalization of the cornea and conjunctiva. One patient (Case No. 1) presented with dysphagia and otolaryngologist diagnosed it to be cicatricial pemphigoid. All of our patients presented with obliterated inferior fornices suggesting being a late stage of the disease. Except for one patient (Case No. 2) all had already developed trichiasis and entropion and three of them had undergone repeated surgeries, which failed. Three (50%) of the cases had received systemic steroids in belief it to be Sjogren's syndrome although none of them gave history of arthritis or other collagen disorder.

DISCUSSION

The diagnosis of ocular cicatricial pemphigoid (OCP) is extremely important, given the natural history of the disease, the effectiveness but the potential toxicity of therapy and the potential confusion from other causes of chronic cicatrizing conjunctivitis. The reported incidence of the disease in the literature ranges from 1: 8000 to 1: 20,000.^{2,4} In our study the incidence was 1: 2427 which seems to be much higher but as our unit is a referral center it may be only due to the reason that cases have been concentrated. Most reports describe a slight but clear female predilection⁵ but in our study we could not found any sex predilection. Average age of our patients is 56.6 years, which is in accordance to other reports⁶. All of our patients presented with complications (except one case no. 2), which indicates late presentation due to unawareness regarding the condition. Four of our cases (67%) had systemic mucous membranes involvement, which is also confirmed by other studies.³ Only one of our patients (17%) had skin involvement, which is a much less incidence as compared to other studies, which indicates involvement of skin in 50% of cases.³ Only one of these patients had a prior diagnosis of cicatricial pemphigoid and

Patient	Age	Sex	VA		Ocular Involvement						Systemic		
					Fornices	Lids	Cornea		Conjunctiva		Mucous Membrane	Skin	
			R	L			R	L	Re	RL			
1	65 yrs	M	6/60	PL	Both Upper and lower obliterated in (both eyes)	Trichiasis Entropion (both eyes)	Opacity	Perforated Ulcer	Xerotic	Keratinized	+	-	-
2	45 yrs	F	6/12	6/9	Lower obliterated in (both eyes)	Both eyes OK	Clear	Clear	Inflamed	Inflamed	+	-	-
3	65 yrs	F	6/60	6/60	Lower obliterated in (both eyes)	Trichiasis Entropion (both eyes)	Opacity	Opacity	Xerotic	Xerotic	-	-	-
4	50 yrs	M	6/36	FC	Lower obliterated in (both eyes)	Trichiasis Entropion (both eyes)	Hazy	Opacity	Xerotic	Xerotic	-	-	-
5	65 yrs	F	FC	FC	Lower obliterated in (both eyes)	Trichiasis Entropion (both eyes)	Opacity	Opacity	Xerotic + Keratinized	Xerotic + Keratinized	+	-	-
6	65 yrs	M	PL	PL	Total Symblepharon	Trichiasis Entropion (both eyes)	Opaque	Opaque	Epidermalization	Epidermalization	+	+	+

TABLE - 1

that also for his esophageal lesion. All of the patients with conjunctival disease had been treated for trachoma.

Immunologic basis as the cause of ocular pemphigoid is suggested by findings that include: i) Immunoglobulins and C_3 complement bound to the basement membrane of involved conjunctiva.⁷ ii) Circulating antibodies directed to conjunctival epithelium, basement membrane of skin and buccal mucosa.⁸ iii) There is an increased incidence of HLA DQw7 antigens.⁹ iv) Approximately half the patients have elevated serum immunoglobulins A levels.¹⁰ Histologically the benign mucous membrane pemphigoid is characterized by sub-epithelial bullae, which ruptures and are replaced not by epithelium but the fibro vascular tissue containing lymphocytes and plasma cells. The vascular and inflammatory component lessens with chronicity, resulting in contraction of the fibrous tissue with subsequent shrinkage and scarring.³

The condition is chronic and progressive. Bullae are rarely seen clinically on the conjunctiva. Bullae may be seen in the mucous membrane of the mouth, nose and pharynx. Skin lesions may be seen on the face, scalp or other parts of the body. The ocular condition continues as a non-responsive chronic conjunctivitis with aropy discharge. As the disease advances, there is gradual obliteration of fornices due to sub-epithelial scarring. Typically the inferior fornix is compromised first as opposed to trachoma where the upper fornix is involved first. With scarring of the conjunctiva there is occlusion of the lacrimal gland openings, entropion and trichiasis. Keratinization & epidermalization of the conjunctiva leads to keratoconjunctivitis sicca. Corneal ulceration is a frequent secondary complication. Symblepharon results which progresses to ankyloblepharon and finally loss of eye.^{11,12,13}

The management is usually difficult and disappointing. Both topical and systemic steroids have been tried with limited success.¹⁴ Chemotherapy for cases that are extremely active may be tried with prednisolone, cyclophosphamide, dapsons and azathioprine with good results in experienced hands.¹⁵ Treatment of these patients is supportive in nature. For dry eyes various types of artificial tears and lubricants should be used. Trichiasis, entropion and epithelial keratinization from drying and exposure may be managed with bandage soft contact lenses and artificial tears.¹⁶ It seems that ocular surgery for correction of conjunctival scarring, entropion and symblepharon ultimately results in a more rapid progression of conjunctival scarring if the inflammation is not controlled with immunosuppressants. Mucous membrane grafting may benefit in cases of cicatricial entropion.¹⁷ Corneal ulcers are treated with appropriate antibiotics. When there is appreciable visual loss due to corneal scarring, penetrating corneal graft is usually unsuccessful because of the drying, exposure, trichiasis, entropion and susceptibility to infections. Keratoprosthesis may be considered in some patients and may have some limited success.¹⁸ Although trachoma may simulate cicatricial pemphigoid but trachoma affects the upper conjunctival fornix and upper lid first and only in very late cases the lower fornices are affected and even then they are not fully obliterated. The complication of trachoma occurs late in life but the history of inflammation can be traced to early childhood while cicatricial pemphigoid typically starts in 4th decades of life.¹⁹ Repeated surgery on three of our patients would have caused more rapid progression as no prior control of inflammation had been achieved. Topical steroids are of little help and systemic therapy has to be instituted to retard the progression of the disease. The rate of complications indicates the severity of the disease and poor treatment modalities.

CONCLUSION

It is important that Ocular Pemphigoid may be considered in the differential diagnosis of cicatrizing conjunctivitis and repeated inflammations of other mucous membrane, as early and prompt diagnosis may help in the retardation of progression of the disease and its complications.

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