LICHEN PLANUS

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CLINICAL CASE

Referring Diagnosis

"Cicatricial Pemphigoid involving the conjunctiva and the oral mucosa."

History of Present Illness

42-year-old female with a 3 months history of bilateral redness and discomfort of her eyes. The patient denied any acute changes in her vision and had no previous episodes of a similar foreign body sensation. She was referred for treatment of Ocular Cicatricial Pemphigoid.

Past Ocular History

Non-contributory.

Past Medical History

Oral Lichen Planus of 5 years duration.

Ocular Medications

Artificial Tears OU qid. (No relieve of symptoms)

Family History

Non-contributory

Review of Systems

Non-contributory

Examination:

Vision: 20/20 OU Intraocular Pressure: 18 OU

Slit Lamp Exam:

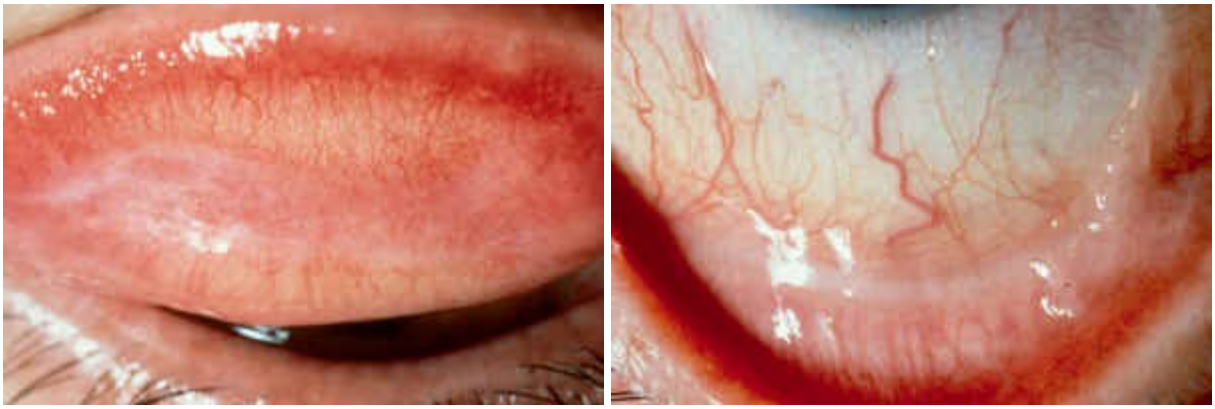

Figure 1: 2+ injection OU (OD>OS) with subepithelial fibrosis and fornix foreshortening.

Laboratory Work-up

Negative

Differential Diagnosis of Cicatricial Conjunctivitis

Infectious	Allergic	Autoimmune	Miscellaneous

Trachoma Adenovirus C.Diphteria	Atopic Keratoconj. Stevens-Johnson	OCP Sarcoidosis Lupus Scleroderma Lichen Planus	Ocular Rosacea Chemical Burns Trauma Medicamentosa Radiation Neoplasia

Conjunctival Biopsy

1) No evidence of immunoreactant deposition in the epithelium Basal Membrane Zone (BMZ).

2) Immunofluorescence staining for Collagen IV and Fibrinogen: Abnormally broad and fragmented BMZ with reduplication

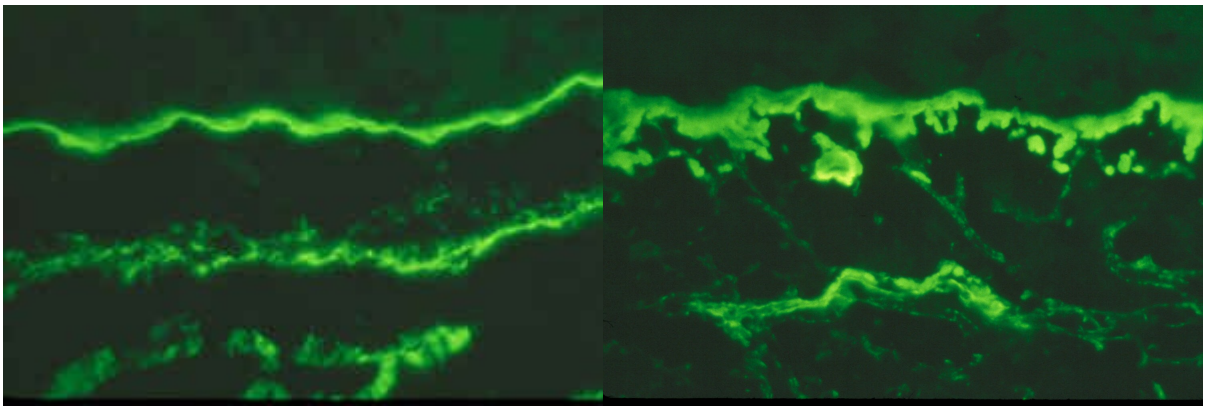

Figure 2: Immunofluorescence staining Collagen IV: Normal (left) and Patient(right)

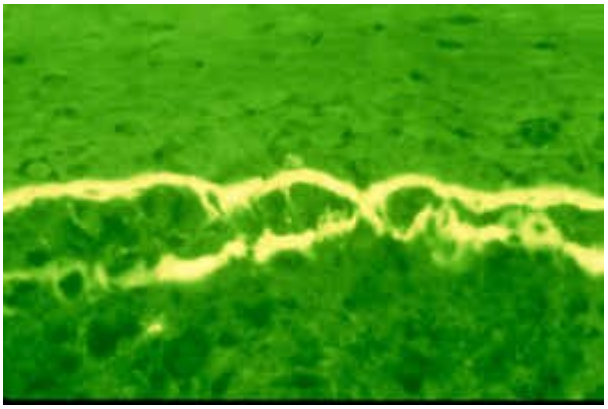

Figure 3: Immunofluorescence staining Fibrinogen: Patient

Diagnosis: CONJUNCTIVAL LICHEN PLANUS

Plan

Topical Cyclosporine-A 2% qid OU

1 Month Follow Visit

Patient complained of burning sensation associated with the use of cyclosporine. Clinical assessment revealed improvement of the ocular surface with decreased fornix inflammation. Cyclosporine was decreased to bid and this was well tolerated.

1 Year Visit

The patient symptoms have completely resolved and cyclosporine treatment was tolerated.

Examination:

Vision: 20/20 OU

Slit Lamp Exam

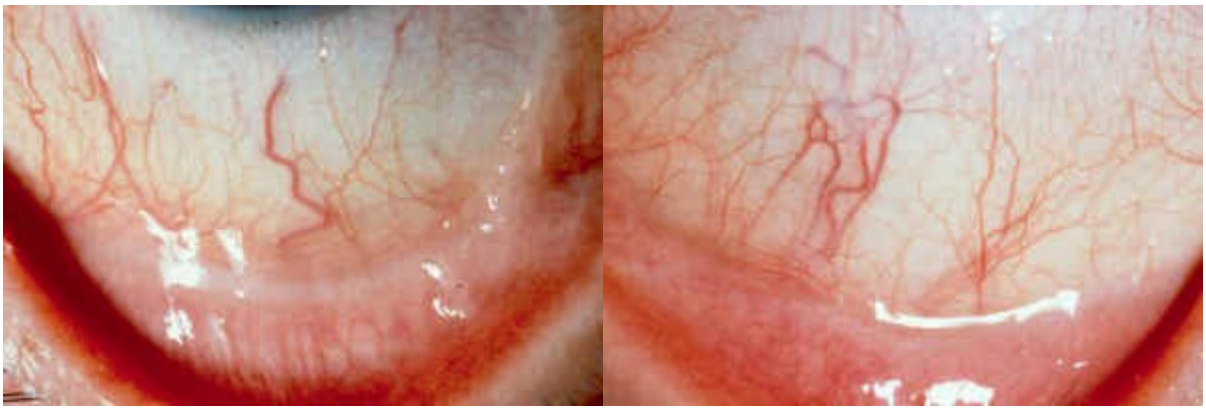

Figure 4: No progression of subepithelial fibrosis: Initial Exam and 1 Year exam.

LICHEN PLANUS

Lichen planus is a common skin disorder that can also involve mucous membranes, scalp, nails and the genitalia. Although its cause is unknown, an autoimmune mechanism is thought to play a major pathophysiological role . There has been no gender or racial predilection associated with lichen planus. The incidence of this disease in dermatology clinics is 1.4%, as compared to 5.0% found in oral medicine clinics . The age of the individuals affected is 30 to 60 years.

CLINICAL FEATURES

The clinical presentation of lichen planus can be characterized by its configuration (linear vs annular), morphology (hypertrophic, follicular, vesicular or bullous) and the site of the lesions. This review will concentrate in the mucous membranes and skin manifestations.

Skin

The cutaneous lesions of lichen planus are typically erythematous to violaceous papules, scaly with a flat topped, polygonal form . The papules tend to involve more commonly the flexural areas of the wrist and forearms. Other skin sites involved include; legs, thighs, lower back, trunk, neck and scalp.

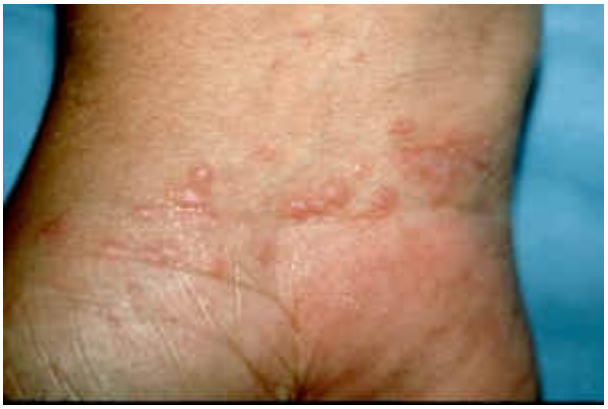

Figure 5: Lichen planus skin lesions

The skin lesions in lichen planus tend to be intensely pruritic. However, evidence of skin trauma, secondary infection or bloody crust is infrequent. In 20% of the patients, the lesions can be asymptomatic .

Mucous Membranes: Oral Mucosa

The oral lesions have a radiating velvet, white or gray appearance, most commonly in a reticulated pattern. This represents the "lace-like" network of linear subepithelial fibrosis found in these lesions. The buccal and glossal mucosas are most commonly affected, however, lesions can also be found in the palate and gingiva (erosive gingivitis).

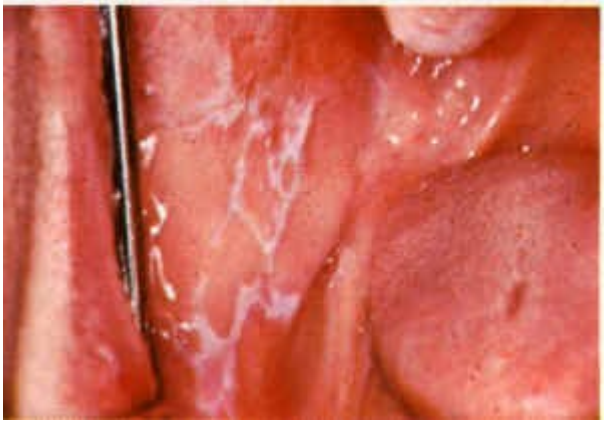

Figure 6: Lichen planus oral mucosal lesion

Pain and burning sensation have been associated with the erosive form of oral lichen planus. Nevertheless, the lesions can also be asymptomatic. In 15% to 30% of the patients with lichen planus, oral lesions can be the sole manifestation of the disease .

Mucous Membranes: Conjunctiva

Involvement of the conjunctiva is rare, however, it has been described since the early 20th century . Most recently, in a study of 584 patients with lichen planus who were evaluated in an oral medicine clinic for extra-oral manifestations, only 1 patient was found to have conjunctival manifestations . Although rare, conjunctival inflammation can be severe and lead to subepithelial fibrosis and scarring .

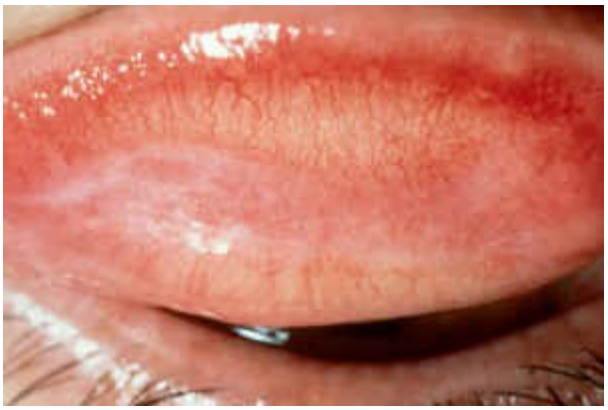

Figure 7: Conjunctival Lichen Planus

The presence of cicatrizing conjunctivitis in a patient with known history of lichen planus,**does not** establish the diagnosis of conjunctival lichen planus. Lupus, bullous pemphigoid and other autoimmune disorders have been described to occur in patients with lichen planus . Therefore, a thorough histological analysis, including immunofluorescence staining, most be performed in order to make a definite diagnosis.

IMMUNOPATHOLOGICAL CHARACTERISITICS

Conjunctival lichen planus has been found to have distinct immunopathological characteristics in biopsy specimens . First, no immunoreactants complexes deposition is present in the BMZ of the conjunctiva. This is important in ruling out other immunobullous diseases such as ocular cicatricial pemphigoid. Secondly and most

important, thickening and reduplication of the BMZ is shown by staining for Collagen IV and VII, fibrinogen and laminin .

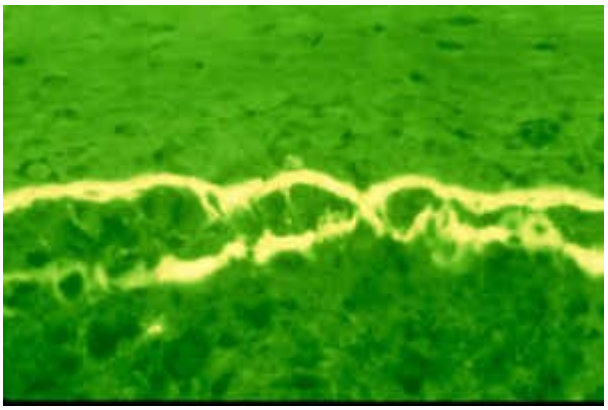

Figure 8: Fibrinogen immunofluorescence staining in conjunctival LP

This is in contrast to oral lesions of lichen planus, where fibrin and fibrinogen are not normally found in the oral mucosa. When present, it is diagnostic of the disease . In the BMZ of the conjunctiva, these proteins are normally present. Thus, the morphological appearance rather than its presence is important in making the diagnosis of lichen planus .

DIFFERENTIAL DIAGNOSIS: IMMUNOBULLOUS DISEASES WITH OCULAR INVOLVEMENT

Pemphigoid vulgaris

Bullous pemphigoid

Cicatricial pemphigoid

Paraneoplastic pemphigoid

Linear IgA bullous dermatosis

Epidermolysis bullosa acquisita

Dermatitis herpetiforme

Lichen planus

CLINICAL COURSE AND

Prognosis

In most patients lichen planus is benign and usually a self-limited disease. The duration is related to the extension and site of involvement. In general, two third of the patients will have spontaneous resolution in a period of 8 to 12 months.

Skin disease alone= 11 months

Skin and mucous membranes= 17 months

Oral mucosa alone= 4 to 20 years

Recurrences and exacerbations can occur in 18% to 20% of the cases.

TREATMENT

The use of many different drugs has been used in the treatment of cutaneous and oral lichen planus. More recently, the use of Cyclosporine-A mouthwash in patients with oral lichen planus was found to be effective in decreasing the severity of symptoms, erythema, erosion and reticulation of oral lesion . Since the use of topical cyclosporine has also been shown to be safe and useful in the treatment of immune-mediated ocular surface diseases, we decided to use it for the treatment of this patient . This treatment successfully relieved the patient from symptoms and halted disease progression. Moreover, elimination of the treatment resulted in recurrence of the inflammation, and improvement with reinstitution of the medication .

Recommendation: Cyclosporine-A 2% gtt qid

TEACHING POINTS

Lichen planus can affect the conjunctiva causing inflammation, subepithelial fibrosis and scarring.

The coexistence of cicatricial conjunctivitis in a patient with history of lichen planus, **does not** establish the diagnosis of conjunctival lichen planus.

Immunopathological analysis of conjunctival tissue is critical in making the diagnosis.

Immunofluorescence staining of the conjunctiva from patients with lichen planus shows: A) No deposition of immunoreactants complexes on the BMZ ; B) Thickening and reduplication of the BMZ, shown with staining against fibrinogen, laminin, collagen IV and VII.

Cyclosporine-A 2% eyedrops can be used for the treatment of conjunctival lichen planus.

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