Chronic Mucocutaneous Candidiasis

by Hatem Kobtan, M.D.

A 48 y white female was referred with the clinical diagnosis of progressive bilateral cicatricial pemphigoid .

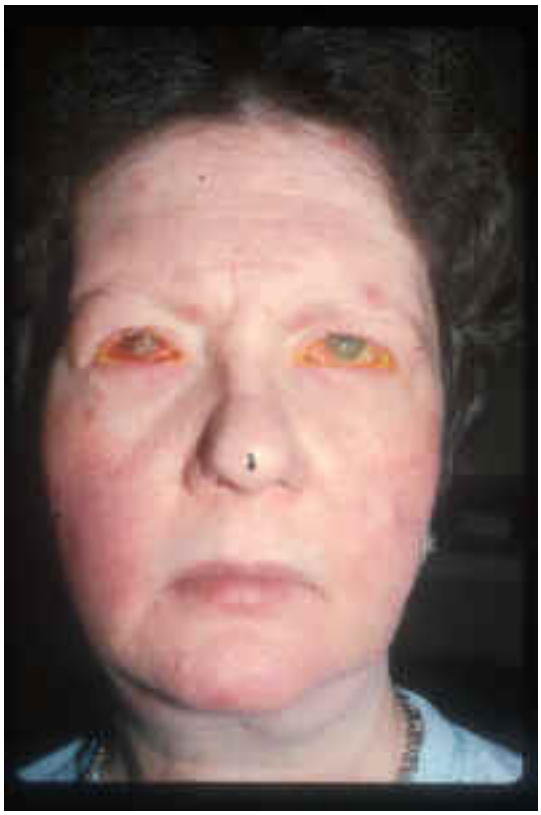


FIG 1

Past Ocular History : chronic irritation and redness, dating over the preceding decade.

Past Medical History :

* oral mucosal involvement with blister formation,

* oral discomfort with eating, particularly food containing acid (tomato).

* Chronic vaginitis unresponsive to topical medication.

* Progressive abnormal finger nails originally believed to be fungal infection refractory to antifungal treatment.

Family History : None

Ocular Medications : Artificial tears.

Topical steroids 8 times/ day .

Examination : General

Lips , buccal mucosa ,nail lesions.

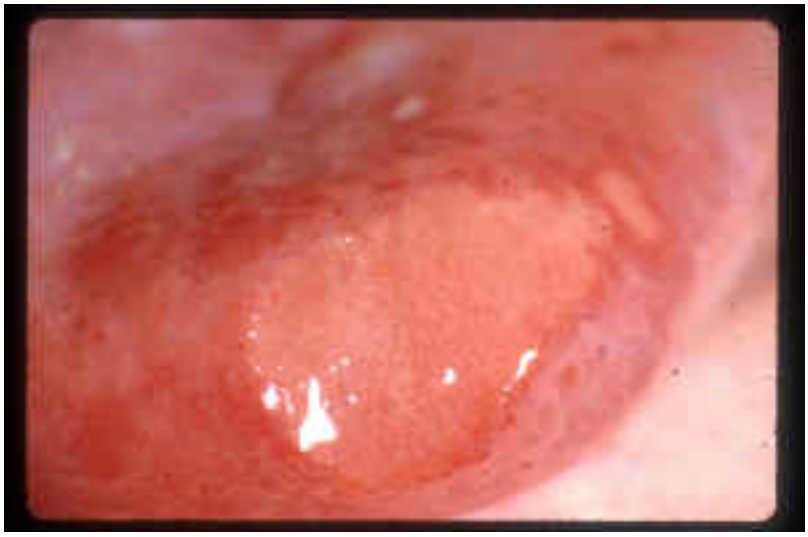
FIG 2 

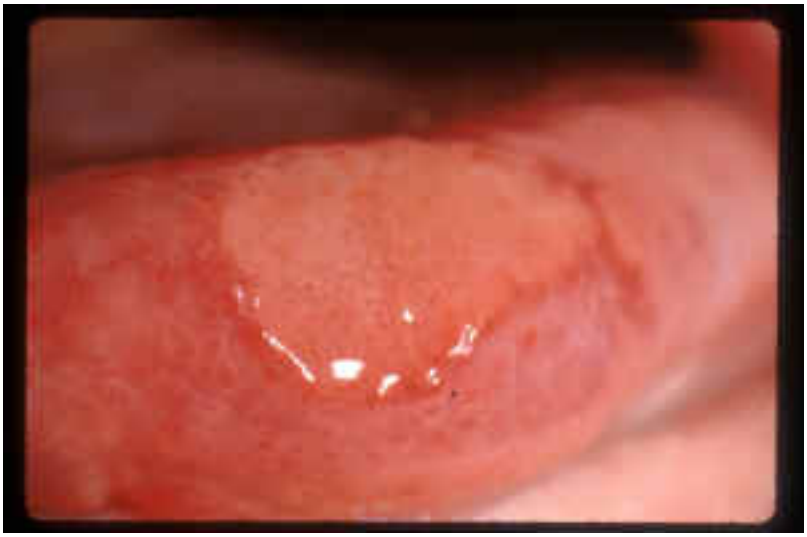
FIG 3 

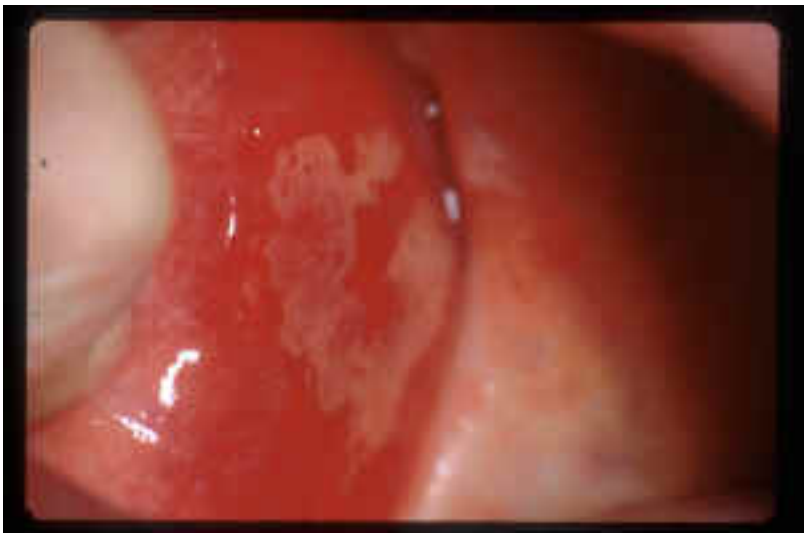
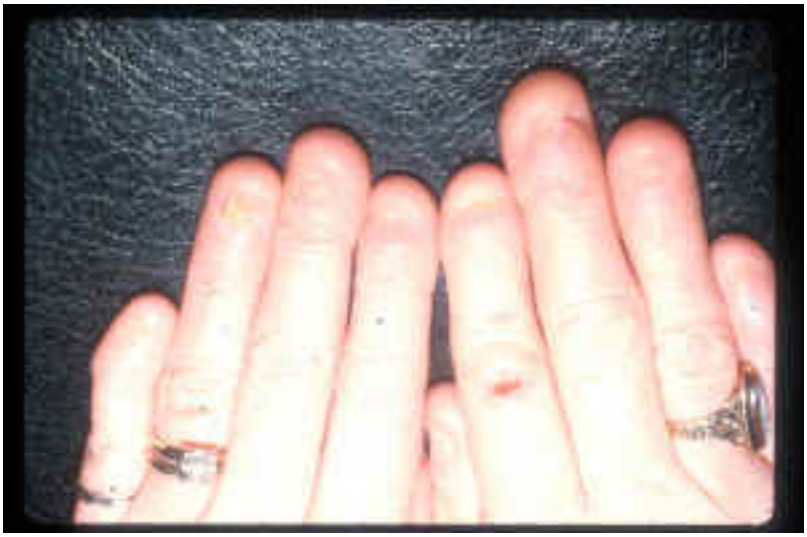
FIG 4 

FIG 5 

Ocular

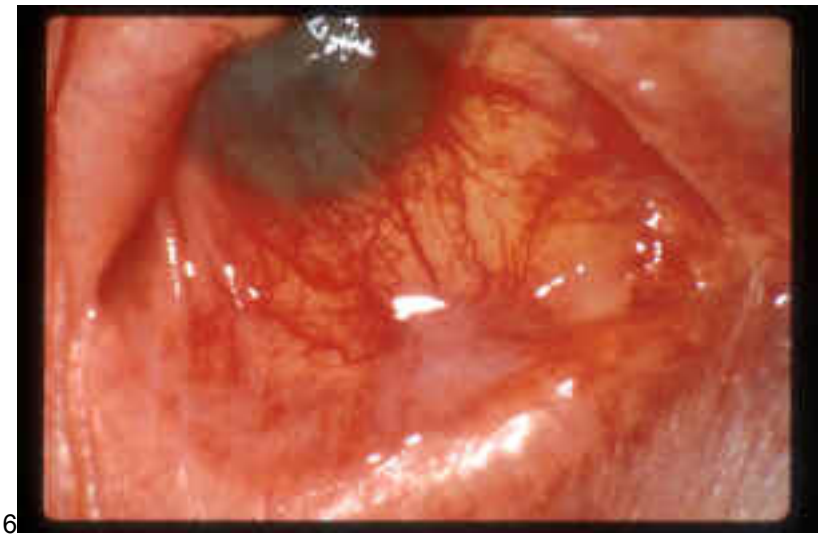
* V/A : 20/400 OD CF5 feet OS

* Pupils: Normal OU

* Motility: Normal OU

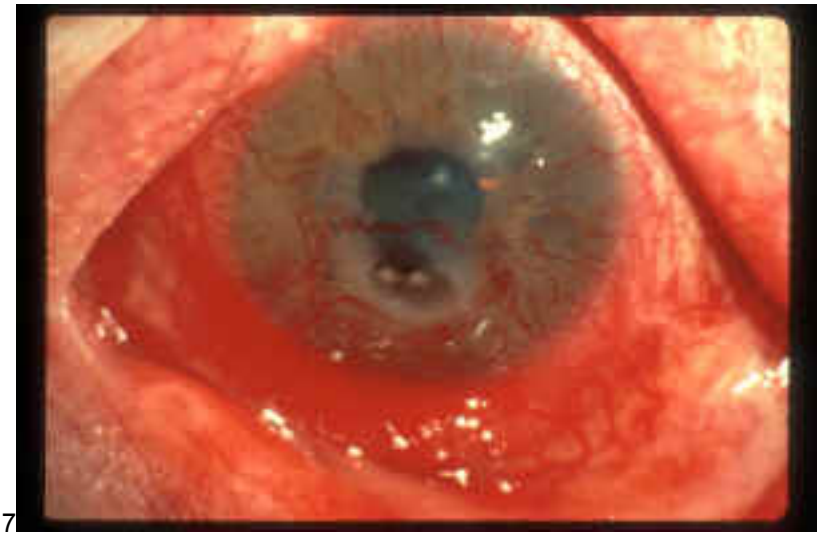
* External exam: Marked injected conj. OU

* Slit lamp: Fornix foreshortening, symblepharon OU.

FIG 6 

keratopathy, scarring & neovascularization OU.

Descematocele OS.

FIG 7 

Investigations :

Scrapping of the corneal infiltrate, Samples of

oral and vaginal lesions underwent microscopy

and culture confirming our suspicion of CMCC.

Treatment :

Medical

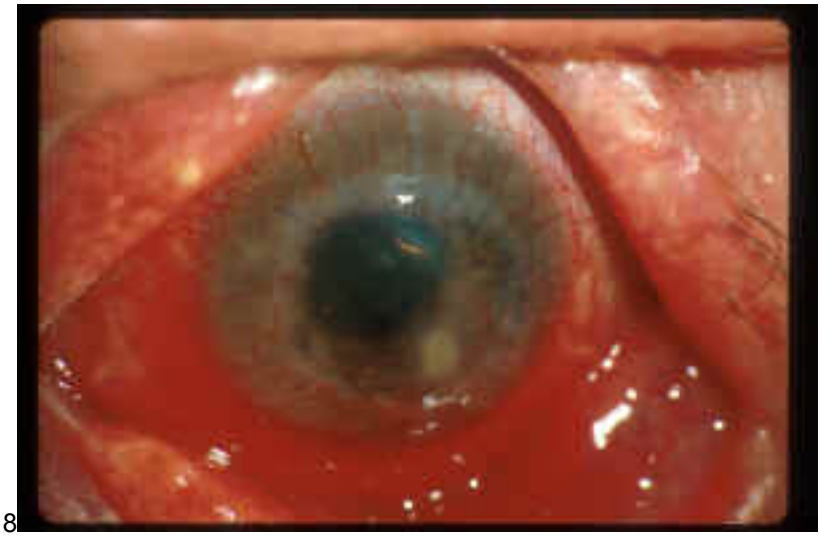
Oral: Ketoconazole 400 mg twice /day.

Topical: Miconazole 1% & 5 flucytosine 1% hourly.

Surgical

Lamellar keratoplasty OS after surgical debridment

Of the edges of the descematocele.

FIG 8 

Follow up : V/A dramatically improved to 20/80 0D & 20/40 OS

Topical antifungals were stopped 3 weeks after surgery.

Systemic antifungal were continued for life. Oral and vaginal mucosal lesions resolved.

Chronic Mucocutaneous Candidiasis (CMCC)

A heterogenous group of clinical syndromes characterized by chronic, treatment-resistant superficial Candida infection of the skin, nails & oropharynx. There is virtually no propensity for disseminated visceral candidiasis.

Immunology:

There are specific abnormalities in cell mediated immunity in many cases

of CMCC, whereas humoral immunity is largely intact.

The following is a listing of some of the commonly described immunologic

abnormalities:

1. Complete anergy to common skin test antigens or selective unresponsiveness to C. albicans antigen.
2. Selective IgA deficiency.
3. Plasma inhibitor to lymphocyte transformation by C.albicans.

4. Serum inhibitor of PMN leukocyte chemotaxis & killing of C.albicans.
5. Combined abnormality of monocyte mobility, phagocytosis and killing.
6. Abnormal complement function.
7. Abnormal macrophage function.
8. Defective suppressor T cell function.
9. Impaired generation of helper T cells.
10. Defective handling by monocytes.

In some cases the Immunologic defect is reversed after successful antifungal therapy. Patients with AIDS usually develop CMCC instead of systemic candidiasis.

Classification of CMCC (Clinical Syndromes)

1. Chronic diffuse candidiasis

2. Chronic localized candidiasis

3. Chronic candidiasis with endocrinopathy.

4. Chronic candidiasis without endocrinopathy.

5. Chronic oral candidiasis.

6. Chronic candidiasis with thymoma.

Diagnosis:

1. Clinical evaluation.

2. Direct microscopic examination of specimens for yeast

(observing the typical budding yeast with hyphae).

3. Isolation of yeast in culture is needed for definitive proof of infection,

(Sabouraud agar with added antibiotic is usually recommended for isolation, whitish mucoid colonies grow within 2-5 days, negative cultures are discarded after 4 weeks)

Differential

Diagnosis:

The lesions of CMCC should be differentiated from Tinea (favus),

Bacterial pyoderma, Acrodermatitis enteropathica.

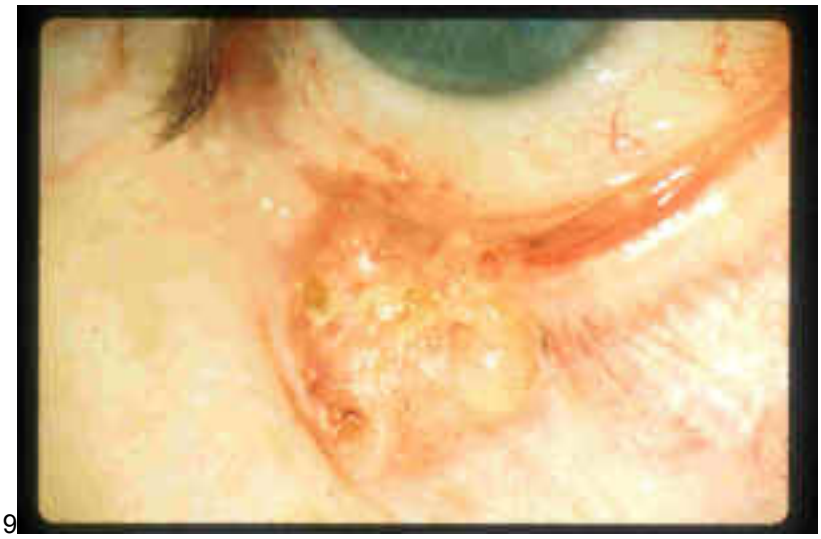
Ocular Manifestations:

1. Conjunctival infection: Patchy raised white areas on the palpebral conjunctiva, follicular conjunctivitis, ulceration and scarring.

2. Cornea infection: Bilateral keratitis occurs in 25-50 % of patients with CMCC with a chronic central epithelial defect, undermined infiltrated edges, anterior stromal scarring ,superficial vascularization is usually present. Keratitis can persist for many years before becoming quiescent.

3. Eye lids: Candida blepharitis resembles Staph. blepharitis (ulceration with loss of lashes) presence of small granuloma near

lash follicles serve as a useful clue to candida infection.

FIG 9 

4. Alopecia of the eye brow skin ,eye lashes can occur in CMCC.

5. Canaliculitis, chronic dacryocystitis or acute purulent dacryocystitis.

Mucocutaneous Manifestations:

1. CMCC usually begins with typical oral lesions which then extend to involve lips, larynx rarely esophagus.

2. Concomitant vaginal candidiasis is typical.

3. Onychial and paronychial lesions arise.

4. Skin of the hand, fingers, nails & scalp becomes desquamated crusted, hyperkeratotic & even granulomatous.

Conditions associated with CMC include

1. Endocrinopathies (Hypoparathyroidism, Hypoadrenalism, Hypothyroidism)

2. Circulating autoimmune antibodies

3. Diabetus mellitus

4. Chronic active hepatitis

5. Pernicious anemia

6. Malabsorption

7. Deafness

8. Ichthyosis

9. Iron deficiency

10. Recurrent pyogenic ,viral and fungal infections

11. Vitiligo

Treatment of Ocular involvement :

A. Eye lids: Stop antibiotic or steroid used on the eye lid,

Nystatin dermatologic cream 100,000 unit/gm 2-3 times /day.

Alternatively,

Amphotericin B (0.3%) sol applied to the lid 3-6 times /day.

5 ´Flucytosine applied 6 times / day.

B. Conjunctiva: Topical Amphotericin B(0.15%) 4 times /day.

C.Cornea : Medical treatment :

1.Topical Amphotericin B (0.15%)/ every 1ô2 hour

during the day and every 1 hour during sleep.

2.Topical Miconazole (10 mg /ml) every 1ô2 hour &

3.IV Miconazole (1.2- 3.6 gm) daily especially

if perforation seems likely.

Surgical treatment :

1. Debridment of necrotic tissue; Superficial keratectomy helps

by removing majority of organisms.

2. Penetrating keratoplasty if impending perforation is present

or deep cornea is involved. At the time of PK make several iridotomies.

3. Glucocorticoids are not used postoperatively.

4. Antifungals are used before and after PK. with subconjunctival

Miconazole(5mg) at time of surgery.

Complications of PK:

1. Secondary glaucoma, peripheral anterior synechiae.

2. Recurrent infection in the graft and graft failure.

Chronic Mucocutaneous Candidiasis

Khayyam Durrani, M.D.

1) Concommitant vaginal candidiasis occurs rarely in patients with MCC T/F

2) In mucocutaneous candidiasis (MCC), most immunologic abnormalities occur in the cellular arm of the immune system T/F

3) Conjunctival involvement in MCC is manifested as a follicular conjunctivitis T/F

4) Patients with MCC tend to have higher rates of disseminated candidal infection than the normal population T/F

5) The underlying immunologic defect persists in all patients with MCC after successful antifungal therapy T/F

6) The differential diagnosis of MCC includes the following except

A) acrodermatitis enteropathica

B) bacterial pyoderma

C) vitiligo

D) Tinea favus

E) All of the above

7) MCC initially manifests signs and symptoms involving the

a)eyes

b)esophagus

c)skin and nails

d)oral mucosal involvement

e)none of the above

8) MCC has been associated with all of the following except

a)Deafness

b)Diabetes mellitus

c)Iritis

d)Hypothyroidism

e)Vitiligo

9) Corneal involvemnt in MCC can be treated with all of the following except

a) IV Miconazole

b) Topical Amphotericin B coupled with topical steroids

c) Penetrating keratoplasty with subconjunctival Miconazole at the time of surgery

d) Superficial keratectomy

e) Topical Miconazole

10) Corneal involvement in MCC may include

a) pannus formation

b) central epithelial defect

c) anterior stromal scarring

d) peripheral ulcerative keratitis

e) all of the above

Answers: 1) F 2)F 3) T 4)F 5)F 6)C 7)D 8)C 9)B 10)D