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# Title

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# **CLINICAL VIGNETTE**

# Xeroderma of the Eyelids

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A 24-year-old female with celiac disease, hypothyroidism, and eczema presents with a new rash around the eyelids. Her symptoms began several weeks ago and gradually increased in severity. She tried various over the counter products including aloe and hydrocortisone cream with limited success. She reports that the rash worsened when she applied any cream around her eyes and when she works at construction sites. The rash is also present on her hands and neck. Current medications include oral contraceptive, Loratidine, levothyroxine, and a rare alprazolam. She does not take supplements and reports no medication allergies.

She is a college graduate and works for a construction company. She strictly adheres to a gluten-free diet and has been able to keep her Celiac disease symptoms under control. She does not drink alcohol nor smoke.

Physical Examination: Blood pressure is 98/62, pulse 64, temperature 98.2 F, height 5 feet 3 <sup>1</sup>/<sub>2</sub> inches, 99.0 pounds. BMI is 17.3.

The patient is comfortable and her exam was normal except for skin. Skin was remarkable for a thin scaly pink plaque-like rash on the right side of the neck, dorsal surfaces of the hands, and around the periphery of the eyelids. Oral mucosa and conjunctiva were unremarkable. The remainder of her skin exam including arms, legs, back, chest, stomach, and scalp was within normal limits

Her labs revealed a normal CBC and chemistry panel. Lipids were normal. Thyroid function tests were normal except for a significantly elevated thyroid peroxidase antibody level. Urinalysis was normal.

#### General Discussion and Epidemiology

Xeroderma of the eyelids (or eyelid dermatitis) is a group of inflammatory skin conditions causing an eczematous-like rash around the eyes.<sup>1</sup> 90% of cases of eyelid dermatitis are female.<sup>2</sup> Risk factors including a history of rosacea, psoriasis, atopic dermatitis, and seborrheic dermatitis.<sup>2</sup> Many conditions can mimic this disorder and need to be considered in the differential.<sup>3</sup> These include dermatomyositis, SLE, Sjogren's, psoriasis, infections, and medication reactions.<sup>3,4</sup> Approximately 3-12% of the population will be affected at some point during their lifetime.<sup>1,4</sup> Although there is no clear racial predisposition, risk factors include living in cooler climates and in densely populated areas.<sup>5</sup> The incidence of the disorder is increasing in

Africa, eastern Asia, western Europe, and parts of northern Europe.<sup>5</sup> Most cases occur in children or teens with 80% of patients being diagnosed before the age 10 and only 2% being diagnosed as adults.<sup>4</sup> Most cases resolve by adulthood.<sup>4</sup> Other risk factors include a family history of atopic or autoimmune conditions.<sup>6</sup> Although the majority of patients with periorbital dermatitis have food sensitivities, food avoidance strategies early in life do not appear to affect the risk of the disorder.<sup>7</sup> The risk of the disorder does seem to be reduced by early life exposure to pets and day care.<sup>8</sup>

# Etiology and Pathophysiology

Causes of eyelid dermatitis include contact dermatitis (allergic and irritant), atopic dermatitis, seborrheic dermatitis, and rosacea.<sup>9</sup> Allergic contact dermatitis is the most common cause of eyelid dermatitis.<sup>9</sup> Other conditions that can mimic eyelid dermatitis include dermatomyositis, urticaria, infections, neoplasms, discoid lupus erythematosus, and systemic lupus erythromatosus.<sup>10</sup> Eyelash curlers, make-up applicators, nail polish, facial or hand moisturizers, artificial nails, household cleaners, carpentry products, paint, shampoos, eve drops, mascara, lipstick, hair dyes, and topical antibiotics can be associated with eyelid dermatitis.<sup>11</sup> Dry weather and a windy climate can increase superficial skin permeability to allergens and irritants leading to edema and lichenification of the skin.<sup>11</sup> Allergic contact dermatitis is believed to be mediated by a type IV hypersensitivity reaction, while irritant dermatitis is believed to be mediated by a direct toxic effect.<sup>11,12</sup> Seborrheic dermatitis is related to colonization by various yeast strains in association with a disrupted immune response.<sup>11,12</sup>

#### **Clinical Features**

Eyelid dermatitis can affect one or both eyes and cause a pruritic, painful eczematous scaly rash around the eyes.<sup>1</sup> In some cases, the rash becomes crusty and oozing vesicles are noted.<sup>13</sup> The conjunctivae is usually spared.<sup>11</sup> As time progresses, the skin becomes thickened and secondary infections can develop.<sup>3,11</sup> Isolated involvement of the upper eye lids is more suggestive of atopic conditions.<sup>11</sup> Some atopic patients often have involvement of both the upper and lower eyelids.<sup>11,12</sup> Ocular complications can occasionally occur such as corneal abrasions and conjunctival infections.<sup>12</sup> Signs and symptoms can be intermittent or can be chronic and long-standing.<sup>12</sup> The shorter the duration, the more likely it is to be allergic contact dermatitis.<sup>12</sup> Atopic patients often have eczematous skin changes in the antecubital regions and sparing of the nasal

region.<sup>12</sup> Irritant contact dermatitis is also common and can be associated with vitamin A creams and alpha hydroxyl acid creams.<sup>12</sup> Upper lid involvement tends to be associated with airborne allergens, while lower lid involvement is more commonly associated with an exposure to a topical preparation.<sup>11</sup> The presence of vesicles can be a sign of herpetic viral infection while honey-colored crusting may indicate bacterial impetigo.<sup>11</sup>

### **Diagnosis and Testing**

Diagnosis is typically made through a careful history and by noting improvement with avoidance of potential causative agents.<sup>13</sup> Although patch testing is often used to isolate specific allergens, many allergens are not included in standard allergy testing.<sup>11</sup> Moreover, it can be challenging to eliminate each of the potential allergens or irritants.<sup>11</sup> If vesicles are present, a viral culture or Tzanck smear should be obtained.<sup>10</sup> Bacterial cultures can be checked if suspected.<sup>11</sup> Biopsies are rarely performed, but occasionally can be helpful.<sup>11</sup> KOH examination should be checked if appropriate.<sup>13</sup> Blood tests (such as total serum IgE, RAST testing, ANA, CPK, and adolase) and skin prick testing are often abnormal, but rarely helpful in terms of diagnosis and treatment.<sup>13</sup>

#### Treatment

Treatment for eyelid dermatitis depends on the suspected etiology.<sup>14</sup> Patients with suspected contact dermatitis usually undergo patch testing and suspected allergens completely eliminated.<sup>11</sup> Patient education is vital in ensuring that the patient avoids any potential exposures.<sup>14</sup> Acute episodes of contact or atopic dermatitis are usually treated with low to midpotency topical steroids for 5-10 days or a short course of oral steroids.<sup>11</sup> Topical steroids should be used judiciously because of the risk of skin atrophy, telangiectasias, and striae.<sup>11,14</sup> Once symptoms have improved, proper moisturization is important to ensure maintenance of the skin barrier.<sup>14</sup> All potential causative agents should be removed if possible.<sup>14</sup> An oral antihistamine at bedtime can sometimes be very helpful during the acute period.<sup>3</sup> Seborrheic dermatitis is usually treated with ketoconazole cream while treatment shampoos (with selenium, zinc, ketoconazole, or salicyclic acid) are used to treat the scalp.<sup>3,14</sup> Steroids are usually avoided in these patients.<sup>2</sup> Eyelid dermatitis related to rosacea can be treated with oral tetracycline, doxycycline, or minocycline for 4-6 weeks.<sup>3</sup> Eyelid hygiene is also very important including warm compresses and antibiotic eye drops.<sup>14</sup>

# Prognosis

Eyelid dermatitis typically has a good prognosis assuming the correct diagnosis is made and treatment plan followed.<sup>2</sup> Some patients improve and then relapse over time.<sup>2</sup> Some patients suffer relapses related to an allergic contact dermatitis from topical antibiotics and inactive ingredients in other preparations.<sup>3</sup> Preparations with as few extra ingredients as possible should be used.<sup>11</sup> Follow-up patch testing may be considered in difficult cases or if patients relapse.<sup>11</sup>

# Clinical Course and Follow-Up

The patient was managed with low-potency topical corticosteroids. Over the course of 6-8 weeks, she gradually improved. She also had patch testing and tested positive for dimethylaminopropylamine. Going forward, she carefully avoided products that contain dimethylaminopropylamine.

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