Chronic Eyelid Lymphedema and Acne Rosacea

Report of Two Cases

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Objective: The authors describe the clinical findings and surgical treatment of two patients affected by chronic eyelid lymphedema associated with facial acne rosacea.

Design: Two interventional case reports.

Methods and Intervention: The clinical diagnosis of acne rosacea was based on the physical examination and confirmed by the histopathologic findings obtained from biopsy of the involved tissue. Surgical treatment was required to address the disfiguring chronic eyelid lymphedema and to correct the resultant mechanical lower eyelid ectropion in both patients.

Results: Surgical debulking of the affected soft tissue resulted in very satisfactory cosmetic and functional improvement in both patients.

Conclusions: To our knowledge, this is the first series of cases of chronic eyelid lymphedema secondary to acne rosacea reported in the ophthalmic literature. Six similar cases have been described previously in the dermatologic literature; all of which had been treated medically without satisfactory results. Surgical debulking of the involved eyelids should be considered in patients affected by persistent symptomatic rosacea lymphedema. *Ophthalmology 2000;107:2220–2223 © 2000 by the American Academy of Ophthalmology.*

Ocular involvement in acne rosacea is very common, affecting up to 58% of patients.[1] This involvement most often consists of blepharitis, meibomianitis, conjunctivitis, and keratitis. Rosaceous lymphedema, or *blepharophyma*, involving the eyelids is a rare complication that may occur in any stage of the disease and represents a diagnostic and therapeutic challenge for the ophthalmologist. Although previously reported in the dermatologic literature, chronic eyelid lymphedema as a consequence of acne rosacea has not been featured in the ophthalmic literature. We report two patients with disfiguring chronic lymphedema of the eyelids secondary to acne rosacea.

Originally received: November 10, 1999.
Accepted: July 19, 2000. Manuscript no. 99748.

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Case Reports

Patient 1

A 53-year-old white female had, at presentation, a 9-month history of chronic progressive lymphedema involving the left upper and both lower eyelids. She also reported recurrent intermittent swelling of the upper cheeks. Five years earlier, the patient had undergone a right radical neck dissection for metastatic squamous cell carcinoma presenting in a cervical node. This was followed by radiation therapy. The source of the primary tumor was not located, and the patient had subsequently showed no other evidence of disease. Her past medical history was negative for thyroid disease, dermatomyositis, systemic lupus erythematosus, angioedema, or sarcoidosis. Before presentation, the patient had been treated for acne rosacea with oral macrolide antibiotics by her dermatologist for several months with no improvement.

Examination revealed erythematous, nonpitting edema affecting both lower eyelids and the left upper eyelid (Fig 1). The skin overlying both lower eyelids showed a peau d'orange appearance. Visual acuity and pupillary reaction were within normal limits. Laboratory studies, including a complete blood count, differential white blood cell count, eosinophil level, erythrocyte sedimentation rate, electrocardiogram, liver function tests, thyroid stimulating hormone levels, serum creatinine levels, and chest radiograph, were normal. Orbital computed tomography imaging revealed preseptal soft tissue swelling, affecting most markedly the left lower eyelid. A punch biopsy from the left lower eyelid revealed dermal edema with dense lymphocytic infiltrate, consistent with rosacea (Fig 2). No histopathologic evidence of lupus erythematosus, dermatomyositis, or lymphoma was present.

ISSN 0161-6420/00/$–see front matter
PII S0161-6420(00)00429-2

Figure 1. Chronic lymphedema affecting both lower eyelids and left upper eyelid. The figure shows marked erythema of the involved eyelids and scattered teleangectasias of upper cheeks.

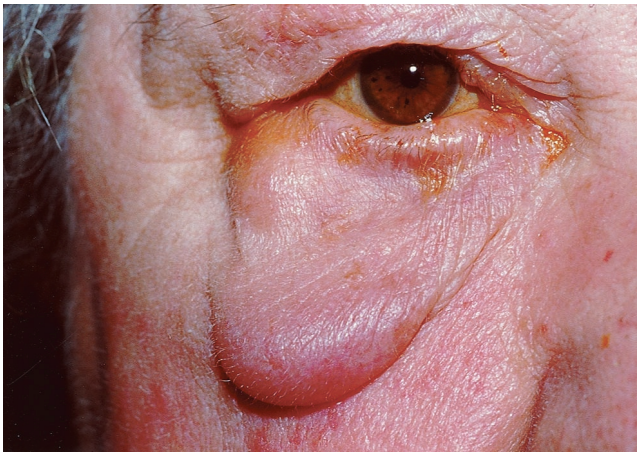

Figure 3. Chronic lymphedema affecting the right lower eyelid. The figure shows marked erythema and teleangectasias of the skin overlying eyelids and upper cheeks.

Patient 2

A 68-year-old white female had, at presentation, a 1-year history of chronic right lower lid swelling and erythema occurring after an otherwise uncomplicated cataract surgery. The patient reported visual field impairment secondary to the mass effect and constant tearing from the affected side as a result of her mechanical ectropion. Her past medical history revealed hypertension and hypercholesterolemia, but was otherwise noncontributory. She had been diagnosed with facial acne rosacea several years earlier and treated with various antihistamines and macrolide antibiotics without relief of her symptoms. Her visual acuity was 20/20 in both eyes with normally reactive pupils. Examination of her right lower eyelid showed chronic lymphedema and mechanical ectropion with punctal eversion; there also was marked erythema of the skin overlying the area (Fig 3). Upper cheeks, nose, and forehead revealed multiple teleangectasias and erythema. Laboratory studies, including a complete blood count, differential white blood cell count, eosinophil level, erythrocyte sedimentation rate, electrocardiogram, liver function tests, thyroid stimulating hormone levels, serum creatinine levels, and chest radiograph, were normal. Orbital computed tomography scanning showed subcutaneous soft tissue swelling of the affected area. No underlying pathologic condition was noted in the sinus or in the orbit. A punch biopsy from the right lower eyelid revealed dermal edema, vascular hyperplasia, and stellate fibroblast infiltration, findings consistent with rosacea.

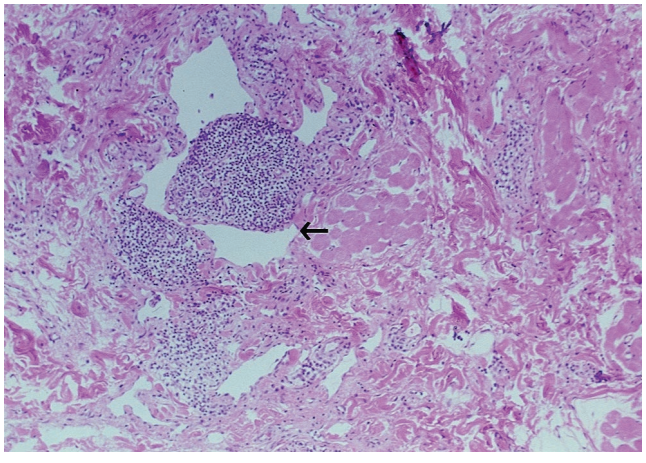

Figure 2. Histologic examination of the excision specimen from patient 1 reveals dilated lymphatic channels and lymphocytic infiltrate (arrow; stain, hematoxylin–eosin; original magnification ×100).

Surgical Procedure

Both patients underwent debulking of the lower eyelid mass, with horizontal eyelid tightening. A subciliary incision was made across the full length of the lower eyelid, and the dissection was then carried through the orbicularis muscle into the postorbicularis fascial plane, down to the level of the inferior orbital rim. In both patients, the bulk of the edema appeared to be concentrated within the preseptal and pretarsal orbicularis muscle, although the orbital septum was also grossly thickened and infiltrated by the process. The orbicularis muscle and orbital septum were excised. The skin was then redraped, excised, and closed. A horizontal tightening was performed via a standard lateral tarsal strip procedure to resuspend the ectropic lower lid. Second and third week postoperative results are shown, respectively, in Figures 4 (patient 1) and 5 (patient 2).

Histopathologic analysis of the excised specimens from each patient confirmed the findings of the punch biopsies. Both speci-

Figure 4. Patient 1 after debulking and horizontal tightening (3 weeks after surgery).

Figure 5. Patient 2 after debulking and horizontal tightening (2 weeks after surgery).

mens were reviewed by a dermatopathologist and believed to represent acne rosacea.

Discussion

Chronic facial edema may complicate the course of many diseases (Table 1).[2] Acne rosacea is a chronic disease involving the skin of the upper third of the face of middle-aged, fair-skinned patients, with women being more commonly affected than men.[3] Blacks are rarely affected.

This disease has three phases. The initial phase consists of erythema and teleangectasia; patients may subsequently develop an "acneiform" component (papules, pustules), and some cases evolve to the third stage, characterized by inflammatory nodules, tissue hyperplasia, or rhinophyma. Rosaceous lymphedema of the eyelids is a rare and disfiguring variant and may present at any stage of the disease. Chronic facial edema may also be found in association with acne vulgaris,[4] suggesting a similar mechanism.

The exact pathogenesis of the lymphedema remains obscure. It has been postulated that chronic inflammation may lead to destruction of elastin around the vessels, resulting in transudation of fluids.[5,6] The chronic inflammation may also cause fibrosis and permanent obstruction of lymphatic vessels of the deep dermis, leading to accumulation of fluids, congestion, and lymphedema of the underlying tissues.

Medical treatment for acne rosacea includes topical metronidazole and oral macrolide antibiotics. Severe cases may require systemic corticosteroids or isotretinoin. However, these treatments are ineffective for rosacea-induced chronic lymphedema.

At presentation, both our patients had clinical evidence of the vascular stage of rosacea involving the forehead, upper cheeks, and nose, with no papules or pustules. Biopsy in both cases was consistent with rosaceous lymphedema. Although chronic lymphedema of the eyelids as a consequence of acne rosacea has not been previously reported in the ophthalmologic literature, there have been six previous cases described in the dermatologic literature; the lymphedema persisted unchanged in all cases treated medically.[7–10]

Table 1. Differential Diagnosis of Chronic Facial Edema

Inflammatory
Acne rosacea
Acne vulgaris
Lupus erythematosus, systemic
Sarcoidosis
Allergic dermatitis
Angioedema
Dermatomyositis
Panniculitis
Granulomatous cheilitis
Infectious
Erysipelas
Mononucleosis
Herpes zoster
Tuberculosis
Mucopolysaccahridoses
Sturge-Weber syndrome
Congenital
Facial hemiatrophy
Infantile cortical hyperostosis
Apert's syndrome
McCune-Albright syndrome
Malignant
Angiosarcoma
Lymphoma
Mycosis fungoides
Leukemia cutis
Lymphosarcoma
Melkersson-Rosenthal syndrome
Myeloma
Kaposi's sarcoma
Miscellaneous
Trauma
Leprosy
Trichinosis
Hypothyroidism
Superior vena cava syndrome
Nephrotic syndrome
Fat pads
Rosai-Dorfman disease

Adapted from Harvey DT, Fenske NA, Glass LF. Rosaceous lymphedema: a rare variant of a common disorder. Cutis 1998;61:321–4.

Surgical debulking of the affected soft tissue in our two patients resulted in very satisfactory cosmetic and functional improvement and should be considered in patients with persistent symptomatic lymphedema.

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