Persistent Eyelid Swelling in a Patient with Rosacea

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Persistent facial swelling may occur as a rare complication of rosacea. This finding has been referred to as lymphedematous rosacea, Morbihan's disease or persistent solid facial edema. A literature review for cases of lymphedematous rosacea revealed that the chronic inflammatory process that accompanies the disease contributes to an increase in the permeability of blood vessels. With time, the lymphatic drainage system becomes permanently impaired, leading to fluid accumulation in the affected skin. Herein, we report the case of a 58-year-old female with history of rosacea who developed bilateral periorbital swelling with associated erythema of the conjunctiva (ocular rosacea) over a 4 month period that only responded to oral corticosteroids. Biopsy revealed changes of lymphedematous rosacea.

Key words: Lymphedematous rosacea, Morbihan's disease, Rosacea

Rosacea is a common cutaneous disease that mostly affects facial convexities of middle-aged individuals. The primary clinical features are transient or persistent erythema, papules, pustules, and telangiectasias (1). The standard classification system established by the National Rosacea Society in 2002 accepted four subtypes of rosacea, namely erythematotelangiectatic, papulopustular, phymatous and ocular (2).

Persistent facial swelling may occur as a rare complication of rosacea which can present in any stage of the disease (3-6). It has been referred to as lymphedematous rosacea, Morbihan's disease or persistent solid facial edema (7). A similar complication has been observed in patients with acne vulgaris (8).

Ocular involvement in rosacea is a common finding, but lymphedematous rosacea of the eyelids, or blepharophyma is a scantily reported ocular complication of the disease (9-11).

Herein, we present a case of lymphedematous rosacea affecting the eyelids as a rare complication of the disease.

Case Report

A 58 year-old woman presented for evaluation of a 4-month history of persistent bilateral swelling of the eyelids. The patient complained of a sensation of dryness of both eyes and an occasional facial burning sensation that started years ago.

On physical examination, she had a bilateral non-pitting edema involving the eyelids, accompanied by a diffuse facial erythema and some telangiectasias. A prominent erythema of the conjunctiva was noted (Figure 1). Laboratory studies revealed a normal blood cell count, erythrocyte sedimentation rate, liver function tests, serum albumin levels and thyroid function tests.

The patient was treated initially by her ophthalmologist with oral doxycycline 100mg daily for ocular rosacea, later increased to twice daily. Despite these therapies she developed bilateral eyelid edema without pruritus, reason why her ophthalmologist discontinued all medications and gave her a course of prednisone 60mg daily with a short 2 week tapering. She had good response with clearing of the edema, however after prednisone was discontinued the bilateral periorbital edema returned and became persistent. It was at this point that she was referred to us for a skin biopsy.

Histopathologic examination of a punch biopsy specimen from the eyelid showed dermal edema in association with dilated vessels and a predominant perivascular lymphocytic infiltrate (Figure 2). An increased number of fibroblasts throughout the dermis were also observed (Figure 2). Mucin stain was negative.

Discussion

Rosacea is a common disease of the skin, which typically affects the central region of the face (cheeks, chin, nose and central forehead) (2). Patients with rosacea usually present a combination of cutaneous signs such as flushing,
erythema, telangiectasias, edema, papules, pustules and phymatous changes (2). The disease is characterized by remissions and exacerbations.

The course of the disease may be complicated by a persistent and progressive swelling with no tendency of spontaneous involution (5, 9-11). This rare complication of rosacea is known as lymphedematous rosacea.

The first description of lymphedematous rosacea was given in 1957 by a French dermatologist Robert Degos. Later the term ‘morbus morbihan’ was introduced to describe a group of patients from Morbihan, France who presented this rare complication of rosacea (6).

Clinically, lymphedematous rosacea presents as a swelling predominantly affecting the central face (forehead, glabella, nose and cheeks) (7). Initially, the lymphedema associated with rosacea is pitting (reversible stage), but with persistence, fibrosis occurs, leading to an irreversible non-pitting induration of the skin (3). It may occur in association with a relatively mild rosacea and in the absence of any other signs of the disease (9-10).

Ocular involvement may occur in 3-58% of patients with rosacea, depending on the series (9-10). Blepharitis and conjunctivitis are the most frequent manifestations of ocular involvement. Lymphedematous rosacea involving the eyelids, as in our case, is a rare reported complication of rosacea.

The pathogenesis of rosacea remains unclear. However, several factors may contribute to recurrent episodes of vasodilatation that lead to an increased permeability of the facial blood vessels and an increase in the lymph load to the affected skin (5-6).

The chronic inflammatory process that accompanies the disease leads to the destruction of the perivascular collagen and elastic fibers, contributing to an increase in the permeability of blood vessels (4-5, 12). With time, the lymphatic drainage system becomes permanently impaired, leading to fluid accumulation in the affected skin. Chronic lymphedema of the facial skin progresses to the end-stage phymatous changes of rosacea (fibrosis and sebaceous hyperplasia) (3).

Histochemical and immunohistochemical studies have shown that up to one third of the dilated vessels in lymphedematous skin are lymphatics (3). Recently, Gomaa, et al. (4) found a significant increased expression of the D2-40 marker for lymphatic endothelium in skin affected by rosacea, in comparison with non-lesional skin. Their findings supports that a process of lymphangiogenesis is involved early in the pathogenesis of rosacea.

The therapeutic options for the disease usually provide unsatisfactory results. Although treatment should address the ongoing inflammatory process, patients with lymphedematous rosacea may fail to respond to...
oral corticosteroids (10). High doses of antibiotics, thalidomide and clofazimine have apparently failed to control the disease. Promising results have been achieved in some patients with a combination therapy of low-dose isotretinoin (0.1-0.2 mg/kg/day) and ketotifen (1-2 mg/day) (7). It is unknown whether an early, aggressive treatment could prevent the onset of the irreversible changes associated with lymphedema. However, surgical management is useful in improving skin lesions in patients refractory to conventional treatment (13-14).

In conclusion, lymphedematous skin changes are a rare complication of rosacea. It may accompany or present in the absence of other signs of rosacea. An impaired cutaneous lymphatic system may play a crucial role in the pathogenesis. Treatment of lymphedematous rosacea is difficult, and no therapeutic approach has been entirely effective.

References


Resumen

El entumecimiento facial persistente puede ocurrir como una complicación poco común de la rosacea. Esta es conocida como rosacea linfedematosa, enfermedad de Morbihan o edema facial sólido persistente. Una revisión de la literatura de los casos de rosacea linfedematosa reveló que el proceso inflamatorio crónico que acompaña la enfermedad contribuye al aumento en la permeabilidad de los vasos sanguíneos. Con el tiempo, el sistema linfático se deteriora, llevando a una acumulación de fluido en la piel afectada. A continuación, reportamos el caso de una paciente de 58 años con historial de rosacea que desarrolló entumecimiento bilateral de los párpados asociado a eritema de la conjuntiva (rosacea ocular) por un período de 4 meses, que respondió solamente a corticoesteroides orales. La biopsia reveló cambios de rosacea linfedematosa.