Blepharokeratoconjunctivitis is an important and underdiagnosed chronic inflammatory disorder observed in children. This disorder describes a spectrum of clinical manifestations, ranging from chronic eyelid inflammation, recurrent chalazia, and conjunctival and corneal phlyctenules to neovascularization and scarring. This condition may compromise visual acuity and lead to amblyopia. At the most extreme end, corneal perforation rarely may occur. Therapy includes eyelid hygiene, topical antibiotic ointment, and steroid therapy. Oral therapy, in the form of tetracycline, has been reported to be successful, but it is only useful in patients older than 8 years because of destructive effects on dental enamel in those younger than 8 years. Oral erythromycin therapy has also been described as effective treatment in several small case series and articles.

While the clinical characteristics and response to therapy are typical, the nomenclature to describe the disease is less so and illustrates the lack of definite etiology. Many terms, including nontuberculous or staphylococcal phlyctenular disease, childhood acne rosacea, and blepharokeratitis have been used. Chronic blepharokeratoconjunctivitis, a term used by Farpour and McClellan, will be used herein, as it describes the full spectrum of clinical manifestations, including a variety of corneal changes; staphylococcal, anterior blepharitis, and meibomian gland dysfunction; and posterior blepharitis. This study evaluated the incidence, history, symptoms, clinical signs, and treatment outcomes of blepharokeratoconjunctivitis in a pediatric population at a tertiary cornea practice.

METHODS

A search of the computerized patient database was performed to identify all consecutive new pediatric patients from January 1, 1997, through December 31, 2002. A child was defined as an individual 12 years or younger. All medical records were examined to determine the reason for consultation and subsequent diagnosis. The medical records of children identified as having blepharokeratoconjunctivitis were further examined. Preceding history and treatment, clinical characteristics, recommendations, and ultimate outcomes were noted.
RESULTS

One hundred ninety-five pediatric medical records were reviewed. Blepharokeratoconjunctivitis was the most common single diagnosis at consultation, accounting for 15% of referrals (29 patients) (Figure 1). This was followed closely by trauma (27 patients [14%]) and central congenital corneal opacities (25 patients [12%]). There was an increase in patients seen with blepharokeratoconjunctivitis during the 6-year period (Figure 2).

Of the 29 cases identified, there were 16 girls (55%) and 13 boys (45%). The mean age at consultation was 6½ years (age range, 2-12 years), while the mean age at onset of symptoms was 4.1 years (range, 0-8 years; n=25). Twenty-six of the patients were white (90%), 1 was Asian (3%), and 2 did not have their race reported (7%). Twenty-one patients (73%) had a history of styes or chalazia noted, and 3 patients had undergone surgical excision of the eyelid lesion. Six patients had a history of atopy, with asthma (2 patients), eczema (1 patient), and significant allergies (3 patients).

At consultation, 11 (38%) of 29 patients were taking high-potency steroids, in the form of topical 1% prednisone or 0.1% dexamethasone, and 4 patients (14%) were taking oral erythromycin. Disease was bilateral in 28 (97%) of 29 patients but significantly asymmetric in 6 (21%) of 29 patients. Amblyopia, attributed to blepharokeratoconjunctivitis, was present in 2 (7%) of 29 patients. Of the 29 patients, eyelid inflammation was present in all patients (100%), superficial punctate keratitis in 16 (55%), and corneal vascularization in 15 (52%). Corneal infiltration was noted in 8 (28%) of 29 patients, with 4 patients (14%) having small, peripheral corneal infiltrates and 4 patients (14%) having classic phlyctenule formation. Corneal scarring was seen in 11 (38%) of 29.

Warm compresses were prescribed to all patients; topical antibiotic ointment was prescribed to 27 (97%) of 29 patients. Oral therapy, in the form of erythromycin (n=21) and doxycycline (n=1), was prescribed to 22 (76%) of 29 patients. The length of oral therapy ranged from 1 to 14 months and was dictated by the clinical course. Two patients had gastrointestinal tract distress, of whom 1 was successfully prescribed a lower dosage and 1 developed mouth ulcers, which were subsequently believed to be unrelated to the therapy.

Steroid therapy was tapered at the initial visit in 11 of 11 patients. Follow-up was available for 15 of 29 patients, with a mean follow-up of 5.4 months (range, 2-25 months). All patients showed clinical improvement. Follow-up was available for 8 of 11 patients in whom steroid therapy was tapered. Therapy with steroids was discontinued for 3 patients or tapered to lower-potency steroids for 5 patients, including fluorometholone, and loteprednol etabonate (0.5% Lotemax; Pharmos Corporation, Alachua, Fla; and 0.2% Alrex; Bausch & Lomb, Tampa, Fla). Recurrences were noted in 6 (40%) of 15 patients; all were successfully managed with the same lower-potency steroid therapy.

COMMENT

Blepharokeratoconjunctivitis is an important inflammatory condition that is commonly seen in children. The
Once again, oral tetracycline was a successful therapy. Keratitis, and findings consistent with acne rosacea, in children with meibomian gland inflammation, bilateral keratoconjunctivitis, one of whom had clinical improvement while taking oral erythromycin. They acknowledged that some believe acne rosacea and phlyctenulosis are disparate presentations of the same process or disease. Ezurum et al described 2 prepubescent children with meibomian gland inflammation, bilateral keratitis, and findings consistent with acne rosacea, including malar erythema, telangectasias, and pustules. Once again, oral tetracycline was a successful therapy.

In a larger series, Culbertson et al described 17 patients younger than 18 years who had phlyctenular disease. Five of the patients were suspected to have acne rosacea, with a definite diagnosis in 2 cases. Five of 10 patients treated positively for Chlamydia species. Most patients received oral erythromycin, with the treatment course ranging from 3 weeks to 9 months. One patient received oral erythromycin (25 mg/kg in 4 divided doses) and preservative-free steroids. Laboratory cultures was given; thus, the descriptive term “blepharokeratoconjunctivitis” for this entity. In our retrospective review, no information about patients’ dermatologic condition or bacterial cultures was given; thus, the descriptive term “blepharokeratoconjunctivitis” is best suited.

This study describes 29 patients with blepharokeratoconjunctivitis: the largest to date, to our knowledge. All of the previous articles refer to this disease as a rare disorder. Similar to previous articles, oral erythromycin therapy was successful and well tolerated. It is unclear if the mechanism of action is a direct effect on lipid synthesis or the influence on the microflora that leads to differences in lipid composition. A study that assessed the ability of erythromycin to alter the microflora concluded that it behaves similarly to tetracycline. The dosage of erythromycin ranged from one quarter strength of that used for an acute infection (50 mg/kg) to 250 mg twice a day.

As this condition is a chronic, recurrent one that affects young patients, the importance of minimizing steroid therapy cannot be overemphasized. The treatment regimen of all patients was successfully tapered from high-potency steroids during initial treatment and recurrences. We recommend the institution of oral erythromycin or doxycycline therapy, age-dependent, in any child who requires steroids to manage his or her disease. While a limitation of this study is the lack of follow-up on half of the cases, this is consistent with the consultation nature of the practice.
Blepharokeratoconjunctivitis is a common reason for cornea referral in children. Oral erythromycin therapy is an effective therapy with a steroid-sparing effect. Recurrences are common and may be successfully managed with low potency steroids.

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REFERENCES


ARCHIVES Web Quiz Winner

Congratulations to the winner of our August quiz, Igor Kozak, MD, University of California San Diego, Shiley Eye Center, La Jolla, Calif. The correct answer to our August challenge was cyst formation due to epithelial downgrowth. For a complete discussion of this case, see the Clinicopathologic Reports, Case Reports, and Small Case Series section in the September ARCHIVES (Ghaiy R, Meyer DR, Farber MA. Epithelial downgrowth complicating evisceration with orbital implant exposure. Arch Ophthalmol. 2005;123:1268-1270).

Be sure to visit the Archives of Ophthalmology Web site (http://www.archophthalmol.com) and try your hand at our Clinical Challenge Interactive Quiz. We invite visitors to make a diagnosis based on selected information from a case report or other feature scheduled to be published in the following month’s print edition of the ARCHIVES. The first visitor to e-mail our Web editors with the correct answer will be recognized in the print journal and on our Web site and will also be able to choose one of the following books published by AMA Press: Clinical Eye Atlas, Clinical Retina, or Users’ Guides to the Medical Literature.