Orbital Myositis in Churg-Strauss Syndrome

A case of orbital myositis in a patient with Churg-Strauss syndrome (CSS) is reported herein. To our knowledge, this association has been reported only once previously. We also describe the previously reported ophthalmic manifestations of CSS.

Report of a Case. A 55-year-old woman was referred to our unit with a 1-week history of redness and pain behind the right eye, which worsened on eye movement.

Her medical history included adult-onset, steroid-dependent asthma, diagnosed at 30 years of age and currently stabilized with inhaled corticosteroid use. She also had, from allergic rhinitis, nasal polyps and severe sinus disease requiring repeated surgery. She had been noted to have peripheral blood eosinophilia on several occasions for the last 7 years and had a recurrent rearr for the last 4 years involving her trunk and back, responsive to corticosteroid treatment and found to be intratumoral granuloma annulaire on tissue biopsy results obtained 4 years earlier. A repeated biopsy 1 year later revealed an intense urticarial inflammatory reaction with perivascular eosinophilic and lymphocytic inflammatory infiltrates. Peripheral blood eosinophil levels, 4 and 3 years previously, were raised at 2300/µL and 2060/µL, respectively. A bone marrow biopsy obtained 3 years earlier revealed a marrow eosinophilia and an elevation of eosinophil precursor populations (eosinophil and eosinophil precursors, 10%) with normal erythropoiesis, myelopoiesis, marrow cells, and marrow architecture. Findings from bone marrow lymphocyte surface marker analysis and gene rearrangement studies were normal. Allergies included sulfur, aspirin, and certain foods, for which she had undergone desensitization therapy.

On examination, she had swelling and tenderness of the right upper lid and bony tenderness over the right eye. Her right conjunctiva was inflamed and edematous with a
raised lesion on the inferior aspect (Figure 1A). She had 2 mm of right axial proptosis (Hertel exophthalmometer readings of 16 mm OS and 14 mm OD, respectively) and a red, swollen, tender, right superior rectus muscle insertion (Figure 1B). The right medial and lateral recti insertions also appeared prominent. Visual acuity (using meters) was 6/6 OD and 6/5 OS. Findings from a full, dilated ocular examination of both eyes was otherwise unremarkable.

A computed tomography scan of the orbits showed significant enlargement of the right superior rectus muscle along with some enlargement of the other recti. There was also opacification of all paranasal sinuses bilaterally (Figure 2A). Biopsy findings of the right conjunctival lump were confirmed histologically to be a focal eosinophilic inflammatory cell infiltrate with no granulomata or evidence of vasculitis (Figure 2B). Complete blood cell count findings were normal, with no evidence of eosinophilia. Rheumatoid factor was borderline at 20 kIU/L (reference, <20 kIU/L). Other unremarkable results included serum electrolyte levels; liver function tests; serum angiotensin-converting enzyme, antinuclear antibody, and antineutrophil cytoplasmic antibody levels; chest x-ray; echocardiography; barium swallow; and stool examination.

She began a reducing course of oral prednisolone (initial dose, 60 mg), 1 day prior to undergoing conjunctival biopsy, and her myositis resolved during the following 2 weeks.

Five months after her episode of myositis, she underwent a biopsy of nasal tissue during functional endoscopic sinus surgery for ongoing sinusitis. The biopsy findings revealed nonspecific inflammatory nasal polyps with prominent eosinophilia. Again, no granulomatous inflammation or vasculitis was identified. These results were considered consistent with CSS, however nondiagnostic in a pathological sense.

Comment. Churg-Strauss syndrome is an uncommon, systemic, vasculitic disorder and is largely typified by a history of asthma, allergic disease not including drug allergy, and eosinophilia (≥10%) on differential white blood cell count.1 Churg-Strauss syndrome classically affects the lung and paranasal sinuses, but extrapulmonary manifestations, namely skin, cardiac,
and gastrointestinal, are also commonly described. Ocular features are unusual. Early recognition of the ocular features of this syndrome, and early institution of appropriate treatment, may minimize complications and potentially completely reverse the disease process.

We report a case of orbital myositis in a patient with CSS. Our patient meets the American College of Rheumatology 1990 criteria for CSS, with a documented history of asthma, eosinophilia (25% differentiated white blood cells), and allergic disease, namely, allergic rhinitis, pansinusitis, and food allergy. Conjunctival biopsy revealed florid extravascular infiltration of eosinophils; however, there were no granulomas or vasculitis. Myositis was diagnosed on clinical and radiological grounds. The conjunctival lump and the superior rectus myositis arose simultaneously and, therefore, can be considered the same disease process, particularly so because both the conjunctival lesion and myositis resolved concurrently with the administration of oral steroids.

The eosinophilic tissue infiltrative phase of CSS is well described, and the lack of histological evidence of granulomas or vasculitis may occur with biopsy techniques such as fine-needle aspiration or bronchoalveolar lavage. However, other explanations include disease suppression by corticosteroid use, or a prevasculitic phase of the illness. A forme fruste type of CSS is described, in which the disease has been partially or completely suppressed by systemic or inhaled corticosteroid therapy for asthma and only appears clinically when changes in steroid therapy are made. This is illustrated by our case, in which the biopsy did not reveal any evidence of granulomata or vasculitis in a patient taking inhaled corticosteroids, with a history of oral steroid use, and 1-day use of oral prednisolone. The rapid response to corticosteroids, as seen in this case, is typical of CSS.

Ocular features of CSS in the literature are relatively sparse but varied. These include conjunctival nodules, corneal ulcer, episcleritis, uveoscleritis, ischemic optic neuropathy, amaurosis fugax, central retinal vein occlusion, central and branch retinal artery occlusion, retinal vasculitis, retinal hemorrhage, cranial nerve palsies, and orbital inflammatory syndrome.

A review by Takanashi et al describes 15 cases in the literature that meet the American College of Rheumatology 1990 criteria for the classification of CSS. This includes a case of dacryoadenitis and myositis. On initial examination, this patient had proptosis and radiologically confirmed symmetrical lacrimal gland and superior and lateral rectus muscle swelling. Biopsy findings of the lacrimal gland and lateral rectus muscle revealed extravascular infiltration of eosinophils.

Churg-Strauss syndrome may be difficult to diagnose. The combination of asthma, eosinophilia, and a history of allergies must raise the suspicion of CSS and the possibility of vasculitis. However, biopsy of ocular tissue may give somewhat indeterminate results.
To our knowledge, orbital myositis has only once been previously reported. In conclusion, we present a case of superior rectus muscle myositis due to CSS. The concurrent association with an eosinophilic conjunctival nodule further illustrates the heterogeneity of the ocular manifestations of CSS.

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Relapsing Diffuse Lamellar Keratitis After Laser In Situ Keratomileusis Associated With Recurrent Erosion Syndrome

Diffuse lamellar keratitis (DLK) is a well-described complication of laser in situ keratomileusis (LASIK) that generally occurs within the first week after surgery. Late-onset cases of DLK have been reported to occur many months after surgery and are sometimes associated with recurrent erosions. We describe 3 patients who had intraoperative epithelial defects and who subsequently developed DLK multiple times in the same location of the same eye, always following an episode of recurrent erosion.

Report of Cases. Case 1. A 33-year-old woman underwent bilateral LASIK in May 2001 for high myopia. Preoperative evaluation revealed clear corneas with no evidence of anterior basement membrane dystrophy. The procedure was uneventful in the right eye. In the left eye, however, a 2.0 × 2.0-mm corneal epithelial defect was noted in the superior paracentral location after creation of the flap, and the epithelium surrounding the defect was noted to be generally poorly adherent to the Bowman layer. A bandage soft contact lens was placed on the left eye, and the patient was instructed to use a combination of 0.1% fluorometholone and 0.3% ofloxacin eye drops, 4 times daily, in both eyes. On the first postoperative day, the patient's uncorrected visual acuity (UCVA) was 20/70 OS. On ophthalmic examination, the contact lens was in place, and there was mild flap edema, but no epithelial defect or lamellar keratitis was noted. The soft contact lens was removed, and on the next day, the patient manifested acutely with reports of decreased vision, foreign body sensation, photophobia, tearing, and pain in the left eye. Visual acuity was still 20/70 OS, but a recurrent 2.0 × 1.9-mm epithelial defect was found in the same location as when the defect had first been noted. No lamellar keratitis was found, and a bandage soft contact lens was placed on the eye. On the third postoperative day, the patient's visual acuity was decreased to 20/400 OS, and moderately severe DLK was found. Treatment with 1% prednisolone acetate was instituted every hour around the clock in place of the 0.1% fluorometholone. By 2 days later, the DKL began to recede, and the epithelial defect was healed. Eight months postoperatively, the patient's best-spectacle-corrected visual acuity (BSCVA) was 20/20 OS.

At this time, the patient underwent an uneventful LASIK recurrent erosion (with relifting of the original flaps). Five months after the retreatment, while the patient was away on vacation, she went to an emergency department with reports of decreased vision, foreign body sensation, photophobia, tearing, and pain in the left eye. She was diagnosed as having a corneal abrasion and prescribed 0.3% ofloxacin, 4 times daily. On seeing an ophthalmologist 2 days later, her visual acuity was 20/60 OS, and she had a healing epithelial defect in a location corresponding to that of her previous 2 epithelial defects, as well as interface inflammation consistent with DLK. The patient was instructed to use both 1% prednisolone, every 2 hours, and 0.3% ofloxacin, 4 times daily. Two days later, the patient returned to our care and was found to have responded well to topical corticosteroid therapy in the left eye with only trace DLK noted and with a BSCVA of 20/30 OS.

Ten and a half months after retreatment, the patient was seen with reports of discomfort in her left eye for 2 days and had a BSCVA of 20/15 OD and 20/25 OS. Once again, an area of epithelial irregularity was found in the same area of the superior cornea in which epithelial defects had previously existed. An underlying moderate DLK was present in the superior flap interface. Treatment with 1% prednisolone acetate was used every 2 hours initially, with a rapid tapering course. The patient responded well to therapy, and the BSCVA is 20/25 OS.

Case 2. A 54-year-old woman underwent sequential bilateral LASIK in September 2001 for moderate myopia. Because the left eye sustained an intraoperative epithelial defect in the inferotemporal para-central location, a bandage soft contact lens was placed on the eye, and the patient was instructed to use both 0.1% fluorometholone and 0.3% ofloxacin eye drops, 4 times daily. The next day, UCVA was 20/80 OS. Although there was still a 2.0 × 2.0-mm epithelial defect present, no interface inflammation was noted. The defect was healed by the third postoperative day with a UCVA of 20/20 OS.

Two and a half months later, the patient was seen with a 2-day his-