fication within the optic cup in the embryonic fissure, causing subsequent microphthalmia and anterior segment maldevelopment. The pathogenesis is uncertain; failure of a clone of pluripotential stem cells in the embryonic fissure to initiate normal retinal development may have occurred. This may be due to a somatic mutation in a gene involved in the retinal signaling pathway, such as CHX-10<sup>9</sup>, PAX6<sup>10</sup>, PAX2<sup>11</sup>, Rx<sup>12</sup>, or sonic hedgehog, SHH<sup>13</sup>, which are required for vertebrate retinal development. These mutations potentially could leave cortical development as a default pathway for retinal differentiation. Thus, a local cerebrocortical cell mass arising in the anterior retina/ciliary region may be analogous to experimental eye formation in ectopic locations when the Drosophila gene, eyeless (ey), homologous to mammalian PAX6, drives eye development wherever ectopic expression occurs. A germline deletion in SHH has been associated with iris and uveoretinal colobomas. We hypothesize in the present case that somatic mutation in such a gene may be causative.

The finding of leukocoria in a child requires thorough evaluation to exclude retinoblastoma or other intraocular malignancies. This neonatal development of an uncalcified, homogeneous intraocular mass in a microphthalmic eye was more consistent with a developmental anomaly than retinoblastoma. With no possible useful vision but potential malignancy, including atypical teratoid medulloepithelioma or retinoblastoma, enucleation of the eye was indicated. This child continues to thrive without needing further medical tests or interventions.

This study was supported in part by grants MT15014 (Dr Chan) and 012329 (Dr Gallie) from the National Cancer Institute of Canada, Toronto, a previous grant, 013136, on retinoblastoma from the Canadian Institutes of Health Research, Ottawa, Ontario (Dr Chan); and grants from the Canadian Genetic Diseases Network, Vancouver, British Columbia; the Retinoblastoma Family Association, Richmond Hill, Ontario; and the Royal Arch Masons of Canada, Hamilton, Ontario (Dr Gallie).

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10. Walther C, Gruss P. PAX-6, a murine paired box gene, is expressed in the developing CNS. Development. 1991;113:1435-1449.
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. Churg-Strauss syndrome classically affects the lung and paranasal sinuses, but extrapulmonary manifestations, namely skin, cardiac,
and gastrointestinal, are also com-
mmonly described. Ocular features are
unusual. Early recognition of the
ocular features of this syndrome, and
early institution of appropriate treat-
ment, may minimize complica-
tions and potentially completely re-
verse the disease process.

We report a case of orbital myo-
sitis in a patient with CSS. Our pa-
tient meets the American College of
Rheumatology 1990 criteria for CSS,
with a documented history of
asthma, eosinophilia (25% differen-
tiated white blood cells), and aller-
gic disease, namely, allergic rhini-
tis, pansinusitis, and food allergy. Con-
junctival biopsy revealed florid
extravascular infiltration of eosino-
phils; however, there were no granu-
lomas or vasculitis. Myositis was di-
agnosed on clinical and radiological
grounds. The conjunctival lump and
the superior rectus myositis arose si-
multaneously and, therefore, can be
considered the same disease pro-
cess, particularly so because both the
conjunctival lesion and myositis re-
solved concurrently with the admin-
istration of oral steroids.

The eosinophilic tissue infil-
trative phase of CSS is well de-
scribed, and the lack of histological
evidence of granulomas or vasculi-
tis may occur with biopsy tech-
niques such as fine-needle aspira-
tion or bronchoalveolar lavage.
However, other explanations in-
clude disease suppression by corti-
costeroid use, or a prevasculitic
phase of the illness. A forme fruste
type of CSS is described, in which
the disease has been partially or com-
pletely suppressed by systemic or in-
haled 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 granulomatata or vas-
culitis in a patient taking inhaled cor-
ticosteroids, 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 lit-
erature are relatively sparse but var-
ied. These include conjunctival nodule,
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 radio-
logically 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 eosin-
phils.

Churg-Strauss syndrome may be difficult to diagnose. The combi-
nation of asthma, eosinophilia, and a history of allergies must raise the
suspicions of CSS and the possibil-
ity of vasculitis. However, biopsy of
ocular tissue may give somewhat in-
determinate 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 eyedrops, 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. On ophthalmic examination, 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 UCVA 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 been previously. 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 bilateral sequential LASIK in September 2001 for moderate myopia. Because the left eye sustained an intraoperative epithelial defect in the inferotemporal paracentral 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 eyedrops, 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-