Ocular syphilis occurs most commonly during secondary infection but may present at any stage. Manifestations include conjunctival chancre, keratitis, iridocyclitis, optic neuritis, chorioretinitis, vasculitis, and retinal vessel occlusion, with panuveitis as the most frequent. Syphilis screening therefore should be considered in all patients presenting with uveitis, retinitis, or vasculitis, particularly in high-risk individuals. HIV infection, male homosexuality, and age greater than 40 years are associated with ocular syphilis in urban North American centres. In our all-male cohort, 3 of 5 patients were HIV positive, 3 endorsed sex with men, and all but 1 were older than 40 years. However, patients may be reticent to disclose sexual history or unaware of partner-incurred exposure. In 2 patients, ocular symptoms occurred months to years following initial infection, highlighting the importance of syphilis testing even in cases without known or recent exposure.

Investigations should include enzyme immunoassay, followed by RPR and TP-PA titres. Negative serology despite strong clinical suspicion should prompt retesting in 2–4 weeks to identify early seroconversion. One patient displayed persistent seronegativity despite biopsy-proven syphilitic gumma. Gumma is an exceedingly rare presentation of late tertiary syphilis and likely reflects inadequate treatment for a primary infection.

Ocular syphilis is a subtype of neurosyphilis. Cerebrospinal fluid pleocytosis and positive RPR titres occur in up to 79% of ocular syphilis patients. Lumbar puncture is universally recommended because further treatment is required if cerebrospinal fluid abnormalities persist beyond 2 years. Lumbar puncture was deferred in 3 patients at our centre because they lacked neurologic symptoms.

Ocular syphilis treatment parallels neurosyphilis protocols. Aqueous crystalline penicillin G 3–4 million units should be administered intravenously every 4 hours for 10–14 days. Penicillin desensitization is strongly preferred over use of alternate agents in patients with penicillin allergy. Visual prognosis is favourable following treatment: a recent study reported 65% recovery at 1 month, with early improvement as the strongest prognosticator. In our patients, 4 demonstrated a final BCVA of 20/30 or better, whereas 1 demonstrated persistent vision loss owing to optic atrophy and chronic vasculitis.

Our series illustrates the heterogeneous nature of syphilitic uveitis and emphasizes the importance of syphilis screening when faced with unexplained or refractory ocular inflammation. Rapid recognition of syphilis can save vision, prevent systemic sequelae, and reduce transmission of a growing epidemic.

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Footnotes and Disclosure
The authors have no proprietary or commercial interest in any materials discussed in this correspondence.
a hazy view OS. Ultrasound of the left eye demonstrated a flat retina with vitreous opacities. The patient was diagnosed with interstitial keratitis with an anterior and intermediate uveitis. The differential diagnosis was broad and included infectious agents, systemic inflammatory conditions, and masquerade syndromes. Because of the history of varicella-zoster virus in the left eye, this presentation was deemed likely secondary to zoster, and treatment was initiated accordingly.

Within 1 week, the patient was referred to the uveitis service. There was mild improvement in the vision OS, and the anterior segment demonstrated questionable iris neovascularization (Fig. 1). Gonioscopic examination demonstrated diffusely engorged iris vessels at the pupil margin, possible neovascularization of the angle inferiorly, and increased iris pigmentation. The patient had also developed cystoid macular edema OS. Preliminary investigations demonstrated acute on chronic kidney disease with worsening creatinine but was negative for HLA-B27, syphilis, sarcoid, and tuberculosis. Less than 1 week later, vision OS dropped to hand motion accompanied by large intraretinal hemorrhages in all 4 quadrants, mild vascular tortuosity, and worsening macular edema (ME; Fig. 2).

Fluorescein angiography demonstrated nonspecific focal areas of leakage in the right eye. The left eye demonstrated multifocal areas of choroidal leakage as well as blockage from vitreous condensations. The differential diagnosis was expanded to include ocular ischemic syndrome, but carotid Doppler examination showed no carotid stenosis. Blood work for a vasculitic etiology was negative, but complete blood count demonstrated anemia with mild thrombocytopenia and elevated inflammatory markers. An anterior chamber tap was done, and herpes zoster virus was detected with >7 million copies. The patient was assessed by the glaucoma service and noted to have zonal iris transillumination defects and blood in Schlemm’s canal, raising the suspicion for elevated episcleral venous pressure. This was ruled out by computed tomography of the orbits and the absence of any cavernous sinus pathology. An ocular oncology assessment ruled out an intraocular tumour that might explain the increased pigmentation noted on gonioscopy. Given the reduced dose of oral antivirals owing to worsening kidney function, the patient was given an intravitreal injection of 2 mg ganciclovir. Nepafenac was also started to reduce the cystoid macular edema.

Concurrently, the patient was followed by his nephrologists, who noted worsening kidney function and uncontrolled blood pressure despite increased use of diuretics. The patient developed extensive peripheral edema, a weight gain of 10 lb, and shortness of breath with crackles on auscultation. An abdominal ultrasound demonstrated nonobstructive nephrolithiasis with hyperechoic nodules in the spleen. A Doppler examination of the lower extremity ruled out venous thrombosis. On follow-up examination, there was no significant improvement in the patient’s ocular findings. A second injection of intravitreal ganciclovir was administered along with systemic prednisone 50 mg per os qd. Minimal improvement was noted in the ME with systemic steroids, and they were rapidly tapered over 2 weeks. Over the subsequent 8 weeks, the patient received 2 intravitreal injections of bevacizumab to target the ME. Further investigations undertaken by nephrology demonstrated an elevated urine protein creatinine ratio of 0.53 g/g. The
patient also had elevated serum free light chains, and serum protein electrophoresis demonstrated elevated gamma-globulins, raising the suspicion of a plasma cell dyscrasia. Finally, a bone marrow biopsy was done that was consistent with multiple myeloma.

Multiple myeloma can affect nearly all ocular structures in the form of crystalline deposits in the corneal stroma, retinopathy, and hyperviscosity syndrome. Given the ocular history of herpes zoster OS and positive herpes zoster virus detection in the aqueous fluid, treatment was targeted toward management of the viral infection. However, the clinical evolution and development of systemic symptoms were not consistent with a viral etiology. The presence of nonspecific changes on fluorescein angiography in the uninvolved right eye raised the suspicion of a systemic disease. The evolving systemic symptoms in addition to close communication with allied treating physicians led to the appropriate diagnosis. It is extremely rare for multiple myeloma to present as panuveitis and even more so for it to have iris involvement, thus raising the suspicion that there was an element of active herpes virus. The patient also demonstrated typical findings of zoster uveitis, including unilateral inflammation, elevated intraocular pressure, iris atrophy, and pupil distortion. Studies examining the association between clinical manifestations of ocular herpes zoster virus and viral load have concluded that viral load in the aqueous humor correlates with the intensity of ocular findings, including iris atrophy and pupil distortion. Thus, the high viral load detected further supports a role for active virus in this patient’s case. Several studies have demonstrated an increased incidence of zoster reactivation in the context of multiple myeloma. We speculate that the hyperviscosity retinopathy in the left eye was likely secondary to multiple myeloma. Although hyperviscosity syndrome in association with multiple myeloma is rare, it has been reported.

This case emphasizes the need to maintain a broad differential diagnosis in patients presenting with intraocular inflammation, the importance of a multidisciplinary approach in patient management, and consideration of infectious etiologies in immunocompromised patients.

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Varied management of idiopathic intracranial hypertension in female-to-male transgender patients

Idiopathic intracranial hypertension (IIH) is characterized by elevated intracranial pressure, visual disturbances, papilledema, and headache. There is an association between sex hormone excess and IIH, particularly in patients undergoing hormonal therapy for gender transition. The increased androgen concentration during testosterone therapy for gender reassignment presents an opportunity for studying different treatment modalities. First-line treatment of IIH involves dietary changes and a carbonic anhydrase inhibitor such as acetazolamide. Surgical treatment options include shunts (ventriculoperitoneal [VP] or lumboperitoneal), optic nerve sheath fenestration (ONSF), and endovascular stenting. Though both ONSF and VP shunting remain standards of care when medical therapy fails, there are little data on this procedural combination. We present the first patient to undergo a successful simultaneous ONSF and VP shunt treatment for IIH.

A 23-year-old morbidly obese (body mass index = 54 kg/m²) female-to-male transgender patient on testosterone therapy with a history of migraine and asthma but no ocular history presented to the emergency department with worsening left-sided headache and blurry vision. Vital signs in the emergency department revealed temperature of 36.4°C, blood pressure of 146/102 mm Hg, pulse of 62, and respiratory rate of 14. The patient’s headache, nausea, and photophobia began 3 weeks before presentation. The blurred vision OS started 2 weeks before presentation, whereas vision began to decline OD 2 days prior.