not sufficiently explain a patient's clinical presentation. Being familiar with the typical clinical presentation of each entity has the potential to decrease morbidity associated with delayed diagnosis and treatment.

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Idiopathic intracranial hypertension in a transgender female

A 39-year-old morbidly obese African-American male-to-female transgender patient presented with acute-on-chronic vision loss in the right eye. She reported a 1-year history of a dim temporal field OD initially at night, which progressed acutely to include daytime as well. The patient was seen by an optometrist, who found bilateral optic disc edema and referred the patient to an outside hospital. A computerized tomography (CT) of the head was normal.

She reported continued intermittent headaches, transient visual obscurations lasting seconds, bilateral tinnitus, and an episode of diplopia the night before presentation. She denied any recent weight change or new medications. She had no past ocular history. Past medical history included human immunodeficieny virus (HIV) with a CD4 count of 856, viral load undetectable, on abacavir/dolutegravir/lamivudine. During the gender transition process, she took estradiol, ethinylestradiol, spironolactone, and conjugated estrogen tablets, and has not taken these medications for an estimated 4–6 years. She had hypertension treated with hydrochlorothiazide and valsartan, and chronic atrial fibrillation treated with amiodarone. The patient was taking rivaroxaban for a prior deep venous thrombosis. She had a stable fusiform aneurysm of the left carotid artery terminus and morbid obesity (body mass index 45 kg/m^2). Past surgical history included cosmetic facial procedures. She denied smoking, alcohol, or drug use, recent sexual activity, and recent travel.

On presentation the patient was hypertensive to 155/85 mm Hg, and her pulse was 81 beats per minute and had a regular rate. The remainder of the nonocular physical examination was unremarkable.

Best corrected visual acuity was 20/25 OD and 20/20 OS. Intraocular pressures were 14 mm Hg OU. There was a right relative afferent pupillary defect. Ishihara color plates were 7/14 OD and 14/14 OS. Extraocular movements were intact and the slit lamp exam was normal OU. Automated perimetry (Humphrey visual field testing 24-2) revealed a dense superior and inferior altitudinal field defect OD and a markedly enlarged blind spot with a superior and inferior nasal step OS (see Fig. 1). Optical coherence tomography revealed a retinal nerve fiber layer thickness of 157 microns OD and 173 OS (normal range 97.3 ± 9.6 microns) with evidence of macular edema OU.[1] A fundus exam revealed Frisen grade IV optic disc edema OU.

Bloodwork, including complete blood count (for a female reference range), hemoglobin A1c, vitamin B12, folate, and thyroid stimulating hormone levels were all normal. The estimated glomerular filtration rate (GFR) was artificially low at 47 mL/min/1.73 m^2, given that it was calculated for a female (rather than a chromosomal male); upon recalculation for a patient of male sex, it was

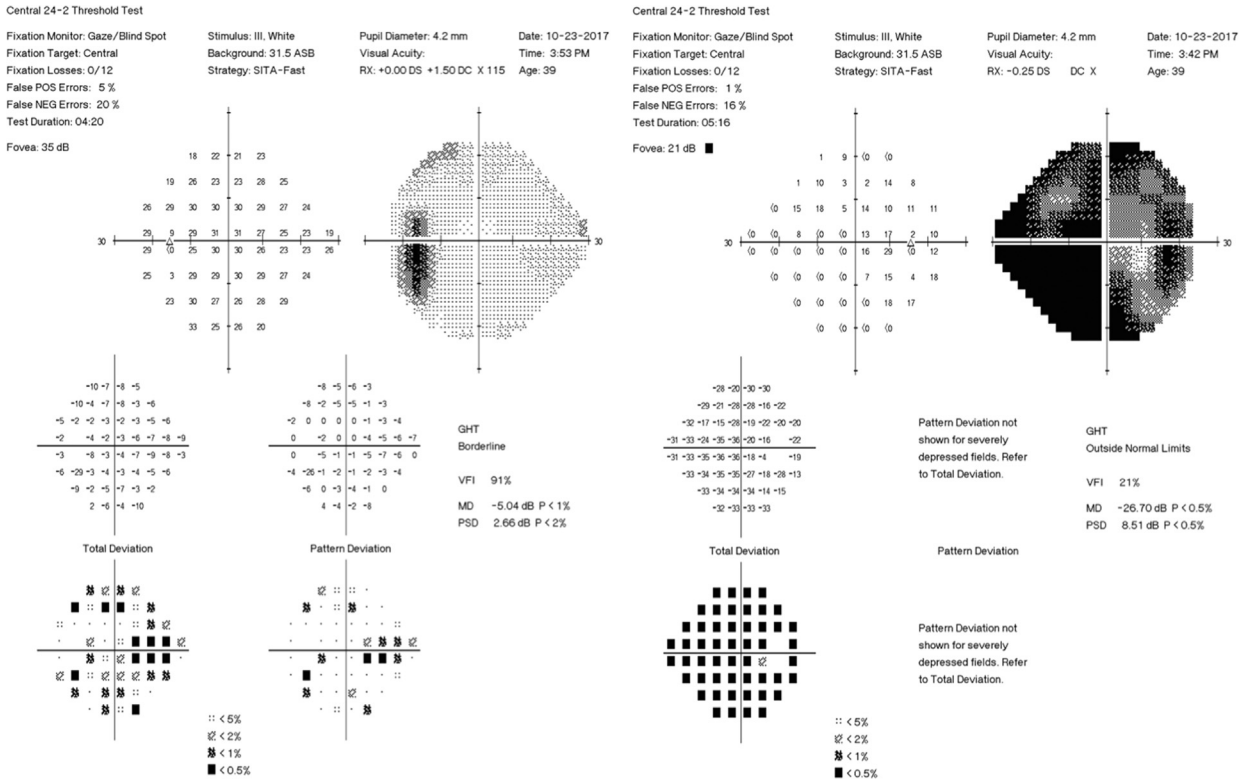

Fig. 1—Humphrey Visual Field Testing 24-2 preformed at the time of consult. This testing depicts a generalized restriction in the right eye and enlarged blind spot in the left eye.

normal at 63 mL/min/1.73 m^2. The basic metabolic panel was otherwise normal.

Magnetic resonanance imaging (MRI) of the brain revealed a partially empty sella, compression of the globes, and perineural optic nerve fluid accumulation bilaterally (Fig. 2). Severe narrowing of the distal right transverse sinus was noted on magnetic resonance venography, which was deemed congenital on subsequent CT angiogram; no sinus thrombosis was noted. Lumbar puncture (LP) opening pressure was 40 cm H_2O; cerebrospinal fluid (CSF) IgG synthesis rate, glucose, white blood cells, and protein were all normal. CSF cultures and cryptococcal antigen testing were negative. A diagnosis of idiopathic intracranial hypertension (IIH) was made based upon the modified Dandy criteria.

The patient underwent a right optic nerve sheath fenestration, tolerated the procedure well, and was started on 2 grams of acetazolamide per day by mouth. On post-operative day 1 the patient had subjective improvement in vision and has not since returned to the ophthalmology clinic.

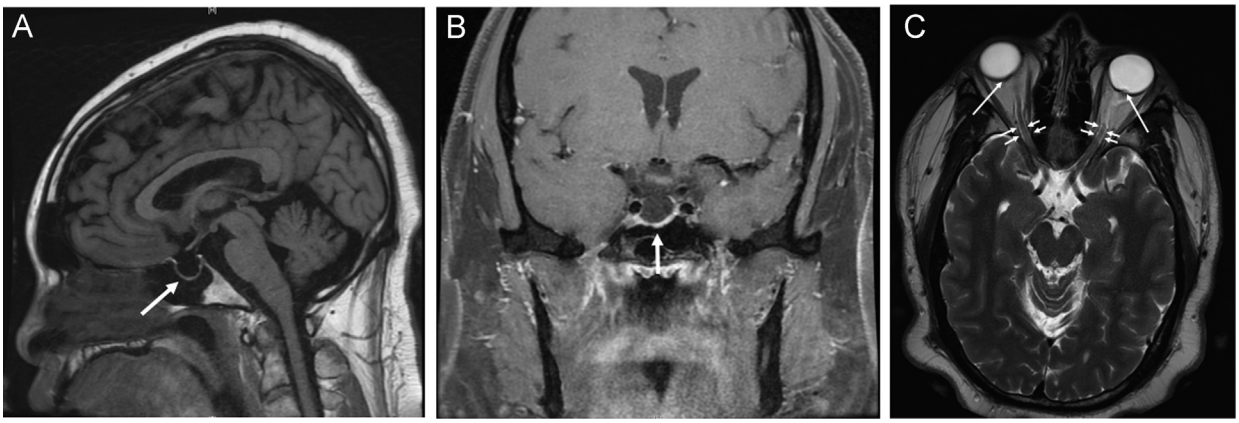

Fig. 2—MRI images illustrating the radiographic findings indicative of increased intracranial pressure. A) T1-weighted image of the sagittal view illustrating an empty sella turcica (white arrow) from excess CSF accumulation and compression of the underlying pituitary gland. B) T1-weighted image of the coronal view of the empty sella (white arrow). C) T2-weighted image of the axial view with fluid accumulation in the optic nerve sheath (short white arrows) and flattening of the globes bilaterally (long white arrows).

Discussion

IIH is largely a condition of obese women of childbearing age. Given the disease's unique association with this population, it has been postulated that sex hormones contribute to the development of IIH.[2] Patients with physiologically elevated estrogen levels, such as pregnant women whose estrogen levels are increased from pregnancy and those taking estrogens, are known to be at higher risk for IIH.[3] It is proposed that elevated estrogren levels result in thrombophilia and thrombosis in the dural venous sinuses, thus resulting in decreased CSF absorption and elevated intracranial pressure (ICP).[3,4]

Additionally, androgen levels (including testosterone, dehydroepiandrosterone sulfate, and androstenedione) have also been studied in the development of IIH. Elevated androgen levels were found to correlate with an earlier onset of IIH.[5] Polycystic ovarian syndrome (PCOS), a disease of hyperandrogenism known to also exist alongside obesity, was found in 39% of women with IIH in one case series.[6] Increased circulating androgen levels were similarly found in 4 reported cases of female-to-male transgender patients with IIH who were receiving intramuscular testosterone to aid in their gender transition.[7–10] A detailed comparision of reported female-to-male transgender patients who received hormonal therapy and developed IIH can be found in Hornby et al.[10] It may be that IIH in transgender and PCOS patients represents a neuro-metabolic complication of an abnormal circulating androgen range (elevated for females, but decreased for males).[10] Utilizing the Naranjo algorithm for evaluating adverse drug reactions, another author identifed a "possible" relationship between testosterone and IIH development in transgender individuals.[11]

The role that obesity plays in the development of IIH may be similarly mediated through hormonal influences. Excess adipose tissue allows for the increased peripheral aromatization of testosterone to estrogen, increased levels of cortisol, and aldosterone production, all of which raise ICP by stimulating CSF production via mineralocorticoid receptors in the choroid plexus.[12] Adipocytes produce the hormone leptin, which stimulates a receptor on the epithelial choroid plexus resulting in sodium and water movement into the CSF.[13] Obesity is a known association with IIH in men, though not as often as when compared to women with IIH.[14] This study interestingly suggested that among IIH patients, African-American men with hypertension are at a greater risk for vision loss and may need to be treated surgically more acutely than others, as in the case of our patient.[14]

Conclusion

To our knowledge this is the first known case of IIH in a male-to-female transgender patient in the English-language ophthalmic literature. In transgender patients, sex hormone disequilibrium can disrupt the normal balance of CSF production and absorption, resulting in elevated ICP and development of IIH. In this patient, there is a complex relationship between altered hormone levels, obesity, African-American race, chromosomal male sex, and existing hypertension. The special clinical challenges of diagnosing and treating transgender patients with IIH include: 1) adjustment of gender-based normal control data for the transgender state (e.g., hematocrit, GFR calculations); 2) the inadvertent misclassification of demographic risk factors for IIH (e.g., a transgender obese male mistakenly not known to have been obese female, PCOS in a transgender female); and 3) use of exogenous hormone therapy. We encourage clinicians to report the development of IIH in transgender patients to better understand and address the unique medical challenges faced by this demographic.

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Adult T-cell leukemia/lymphoma with conjunctival chemosis from infiltration and raised intraocular pressure

Dear Editor,

Ophthalmic manifestations of adult T-cell leukemia/lymphoma (ATLL) are uncommon and have been considered as markers of a poor prognosis for this rare malignant lymphoproliferative neoplasm of mature T cells, which is triggered by retroviral human T-lymphotrophic virus type 1 (HTLV-1).[1–11] While corneal involvement,[12] conjunctival infiltration,[13] intraocular uveitis,[14] and episcleritis[15,16] have been reported in cases of ATLL, this clinico-pathological case report describes bilateral conjunctival infiltration with lymphoma cells associated with raised intraocular pressure (IOP) in the absence of intraocular inflammation.

A patient with no personal or family history of glaucoma was referred for glaucoma assessment with a 3-month history of bilateral red eyes and raised IOP. The patient was initially treated elsewhere for allergic conjunctivitis with Patanol (olopatadine hydrochloride ophthalmic solution) 0.1%. Systemic inquiry revealed an abdominal skin rash, 10 pounds of weight loss, lower limb pain, and malaise. Medications included a steroid inhaler for asthma and occasional use of a steroid skin cream. Ocular examination revealed mild conjunctival hyperemia and chemosis bilaterally (Fig. 1). Visual acuity was 20/25 OD and 20/40 OS. IOP measurements were 24 mmHg OD and 23 mmHg OS. Pachymetry was 584 μm OD and 572 μm OS. There was no proptosis, bruit, or lid abnormality, and eye movements were intact. Anterior chamber depth was normal bilaterally with no intraocular inflammation. Gonioscopy revealed wide open angles. Posterior segment evaluation revealed cup to disc ratios of 0.45 OD and 0.4 OS with normal retina. Visual field testing was normal bilaterally. The patient was referred to the Cornea and External Diseases Service for a conjunctival biopsy, which revealed evidence of T-Cell lymphoma/leukemia infiltration (Fig. 2) and was positive for T-cell markers, including CD4 and Forkhead box P3 (FOXP3) (Fig. 3). The patient was also referred to Internal Medicine for further evaluation. Blood investigations showed leukocytosis, elevated c-reactive protein levels, and anti-HTLV-1 antibody. Subsequent bone marrow examination, including flow cytometry analysis, confirmed ATLL. The patient was referred for further management by the Ocular Oncology Service. With two cycles of chemotherapy, oral steroids, topical ophthalmic steroid drops, and topical hypotensive agents, the conjunctival hyperemia and chemosis resolved and IOP normalised. Despite systemic

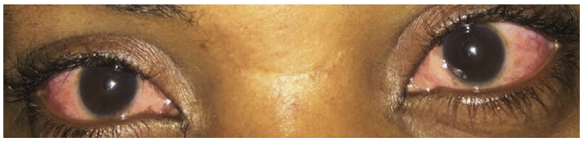

Fig. 1—Photograph revealing bilateral conjunctival hyperemia and mild chemosis at initial presentation.

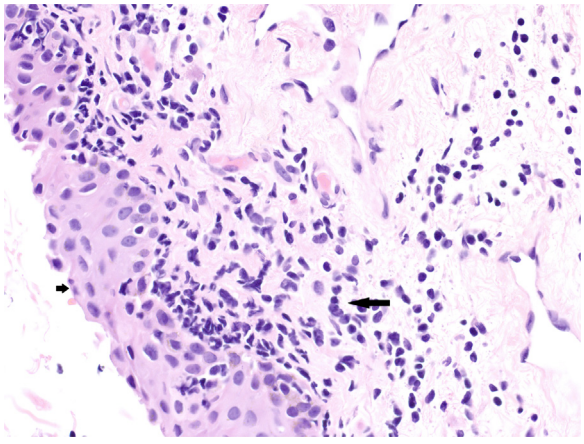

Fig. 2—H&E-stained section of the conjunctival biopsy. The epithelium (short arrow) is intact. The submucosa is infiltrated with scattered lymphoma cells (long arrow). Minimal exocytosis of lymphoma cells is seen in the epithelium (magnification X400).