

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.¹ 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.² The patient also demonstrated typical findings of zoster uveitis, including unilateral inflammation, elevated intraocular pressure, iris atrophy, and pupil distortion.³ 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.⁴ 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.⁵ 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.¹

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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Footnotes and Disclosure

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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.¹ 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.^{2,3} 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 km/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.

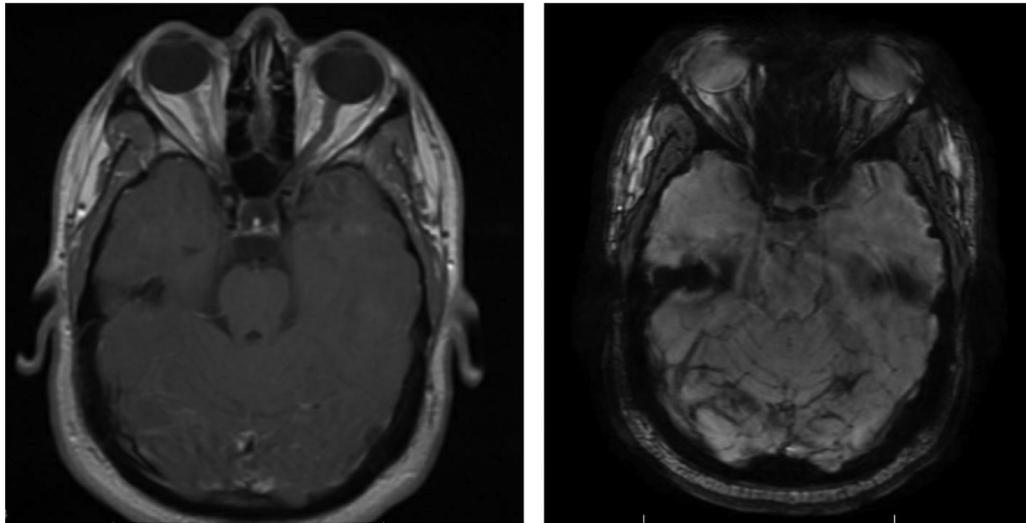


Fig. 1 – Preoperative magnetic resonance imaging of the brain and orbits without contrast medium. Notable findings include flattening of the posterior globes and slight prominence of the distal aspect of both optic nerves.

Ophthalmology was consulted to evaluate for optic disc edema. Uncorrected visual acuity was 20/400 OD and 20/300 OS with no improvement with pinhole in either eye. Pupils were equally round and reactive, with no evidence of relative afferent pupillary defect (RAPD). Intraocular pressures via Tono-Pen (Reichert Technologies, Depew, NY) were 23 mm Hg OD and 24 mm Hg OS. Motility examination revealed full motility OD and –4 abduction deficit OS. Visual fields in both eyes were constricted 360 degrees on confrontation testing. The patient could not identify the test plate during Ishihara color testing. Anterior segment examination was unremarkable bilaterally. Ophthalmoscopic examination revealed 360-degree edema of both optic nerves with obscuration of vessels, diffuse pallor, multiple peripapillary hemorrhages, and cotton wool spots; retinal vessels were tortuous with venous engorgement. Multiple flame hemorrhages were noted in the macula and periphery OU.

Neurology noted a normal neurologic examination aside from a left abduction deficit. Magnetic resonance imaging of the brain and orbits without contrast medium was only remarkable for flattening of the posterior globes and slight prominence of the distal aspect of both optic nerves (Fig. 1). Magnetic resonance angiography and venography of the head were without venous sinus thrombosis. Lumbar puncture yielded an opening pressure of 51 cm H₂O, negative cerebrospinal fluid (CSF) cultures and Gram stain, and normal CSF protein, glucose, and cell count. Based on these findings, the patient was diagnosed with IIH, started on 500 mg acetazolamide, and given the extent of vision loss, underwent bilateral ONSF and VP shunt placement.

One week postoperatively, the patient's cranial nerve VI palsy improved, but horizontal diplopia at distance was noted. His headaches improved. Uncorrected visual acuity had improved to 20/60 OD and 20/100 OS, with

pinhole to 20/70 OS, without RAPD. Motility examination was full OD with an abduction deficit OS. One month later, the patient's visual acuity improved to 20/30 OD and 20/40 OS with 1+ RAPD OD, residual –1 abduction deficit OS, continued 360-degree field constriction, and colors 8/14 OD and 10/14 OS. The patient was lost to follow-up.

Although the pathogenesis of IIH remains unknown, disruption of CSF circulation and metabolic or hormonal effects have been proposed.⁴ Hormonal influences are supported by the increased prevalence of IIH in overweight females and transgender patients undergoing hormonal therapy.³ Obesity was a common factor in both the female-to-male and male-to-female transitions. It has been proposed that the predilection for obesity may be owing to an increased load of circulating hormones.⁵

Our patient presented with headache, visual loss, and left sixth cranial nerve palsy 5 months following initiation of testosterone treatment. Because of the imminent permanent visual loss, surgical treatment was pursued. Visual acuity, abduction, and field deficits improved at 1-month follow-up. Treatment for IIH is specific to patient presentation and depends on the severity and time course of visual loss and structural considerations. However, combined ONSF and VP shunting procedures present a promising treatment option for persistent vision-threatening IIH.

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Corneal limbal xanthogranuloma in Erdheim-Chester disease



Adult orbital xanthogranulomatous disease is a rare non-Langerhans cell histiocytic condition with 4 clinical subtypes: adult-onset xanthogranuloma, adult-onset asthma with periocular xanthogranuloma, necrobiotic xanthogranuloma (NBX), and Erdheim–Chester disease (ECD). We report, to our knowledge, the first case of ECD with limbal infiltration.

A 52-year-old woman presented with xanthomatous cutaneous lesions around her eyes and flexural folds of submammary skin. Subsequent full body imaging revealed sclerotic lesions in her sternum, left rib, and bilateral femurs and tibias. There were no intracranial or orbital lesions. Bone biopsy of the sternal lesions confirmed ECD. The patient was treated with oral steroids and then cladribine by hematology. Two-monthly interval imaging with positron emission tomography showed stable systemic disease.

The patient was referred to the eye clinic with a progressively enlarging left ocular lesion that had been present since her initial diagnosis. Ocular examination revealed a 5 mm well-defined yellow deposit along the superonasal limbus (Fig. 1). There were multiple yellow deposits along her nasal lower eyelid bilaterally. She did not have any optic neuropathy, and her orbital examination was normal.

Biopsy of the lesion showed a subepithelial stromal deposit of foamy macrophages with bland nuclear features (Fig. 2). The cells showed pale eosinophilic cytoplasm with indistinct cytoplasmic borders and ovoid nuclei on highest magnification. Strong and diffuse immunoreactivity for CD68 and CD163 confirmed a histiocytic nature. The lesion also was positive for factor XIIIa but negative for Langerin, CD1A, S100, and BRAF V600E. These identical immunohistochemical features were demonstrated in a previous sternal bone biopsy for this patient.



Fig. 1—Clinical photograph showing xanthomatous lesion on limbus in left eye and multiple lower lid xanthelasmata.