Diffusion-weighted imaging in a case of malignant optic glioma

Angela W. Kim, MD

Malignant gliomas of the anterior visual pathways are rare, rapidly progressive tumors that usually present in middle-aged adults, rapidly progress to complete blindness and death within a year,¹ and often initially simulate optic neuritis clinically and radiographically.¹ Mild proptosis, ophthalmoplegia, and lid edema are seen less commonly, usually late in the disease.¹

We report a case of malignant optic nerve glioma with diffusion-weighted imaging (DWI) findings on MRI that have not previously been reported in this disease, to the best of our knowledge, and highlight the cytotoxic edema/infarction often seen in this condition.

Case report

A 54-year-old diabetic man experienced isolated, progressive visual loss OS for 6 weeks then suddenly developed intractable pain, eyelid swelling, subconjunctival hemorrhage, limitation of eye movements, and mild proptosis. Visual acuity was 20/20 OD and no light perception (NLP) OS. Fundusoscopic examination was normal OD, and showed florid disc edema with both central retinal vein occlusion (CRVO) and central retinal artery occlusion (CRAO) OS.

Orbit CT showed left optic nerve enlargement, sheath enhancement, and stranding in the fat suggestive of inflammation. Extensive blood testing was unrevealing. An orbital inflammatory syndrome was suspected. Pain rapidly responded to oral corticosteroids.

MRI obtained the following day showed nonenhancing enlargement of the left optic nerve and left side of the chiasm, hyperintensity on T2 and DWI, and hypointensity on T1 and apparent diffusion coefficient mapping, consistent with infarction (figure 1). Within hours, he developed a temporal hemianopsia OD. A malignant process was suspected. Lumbar puncture (LP) revealed 7 leukocytes and atypical inflammatory pleocytosis, and was otherwise normal. A 19-mm biopsy of the left intraorbital optic nerve showed necrosis, chronic inflammation, and congested vessels with mural acute inflammation and organizing thrombi, without evidence of neoplasm. A hypercoagulability workup was normal.

He was referred to neurosurgery for intracranial biopsy given the risk to the remaining hemi-field of vision. A 19-day interval MRI showed considerable reduction in previous enlargement and T2 signal, replaced with new enhancement and disappearance of restricted diffusion. Neurosurgery deferred biopsy because of resolved pain and radiographic improvement. The patient lost his medical insurance.

Ten weeks after initial presentation, the patient was NLP OU, despite taking prednisone. Eighteen days later, he presented with mental status changes. MRI showed an infiltrating mass centered in the optic chiasm with extension along both optic tracts (figure 2). A repeat

The Permanente Medical Group, Inc., Ophthalmology, Fresno, CA.

Funding information and disclosures are provided at the end of the article. Full disclosure form information provided by the author is available with the **full text of this article at Neurology.org/cp**.

Correspondence to: akim58@hotmail.com

Figure 1 Initial MRI of optic nerve and optic chiasm

(A) Axial T1 postcontrast study shows shaggy enhancement of left optic nerve sheath, (B) left chiasmal enlargement, patchy enhancement of right chiasm and prechiasmatic nerve. (C) The entire left optic nerve and (D) anterior chiasm are hyperintense on diffusion-weighted imaging and (E, F) hypointense on apparent diffusion coefficient mapping.

high-volume LP showed elevated protein but no malignant cells. An intracranial biopsy revealed glioblastoma, WHO grade IV. The Ki-67 index was 40%. The patient died 2 days later.

DISCUSSION

Proximal optic nerve involvement with malignant optic glioma is associated with painful, severe visual loss with distinctive funduscopy findings of progressive optic disc edema, venous stasis/occlusion, followed by arterial occlusion and infarction.¹ The neoplastic cells tend both to aggregate around blood vessels, causing occlusion, and to envelop the optic nerve beneath the pia mater, causing impairment of capillary perfusion with progressive occlusion of the central retinal vessels distally, resulting in ischemic or hemorrhagic infarction.¹

The differential diagnosis of optic nerve enlargement is extensive and includes optic glioma, optic nerve sheath meningioma, optic neuritis, infiltration by leukemia, lymphoma, medulloepithelioma, histiocytic and granulomatous processes, carcinoma, pseudotumor, Krabbe disease, and infarction.

Although the CT appearance in our patient resembled periopic neuritis, the optic nerve and chiasmal infarction, along with normal brain MRI and the LP results, were not consistent with typical demyelinating optic neuritis. Tolosa-Hunt syndrome (THS) was briefly considered. However, visual loss preceded pain and decreased motility by 6 weeks, the orbital apex/cavernous sinus revealed no imaging abnormalities, and involvement of the chiasm has not been reported in THS. Corticosteroids decreased postischemic pain and edema. On repeat MRI, edema had resolved, and the optic nerve/chiasm demonstrated luxury perfusion, which reversed the previous findings.

The majority of patients with restricted diffusion in the optic nerves have been reported in cases of ischemia associated with surgery, trauma, nonarteritic vascular compromise, embolism, and infection. The only case of restricted diffusion reported in inflammatory optic neuropathy occurred in a young child who developed a CRAO and CRVO due to atypical optic neuritis, presumably from inflammatory vascular occlusion in the anterior intraorbital optic nerve.²

Figure 2 Follow-up MRI prior to intracranial biopsy

T1 axial image shows a heterogeneously enhancing mass centered on the optic chiasm with extension along both optic tracts, measuring 5 cm × 3 cm × 2 cm, with invasion of both medial temporal lobes.

Restricted diffusion in the optic nerve has been reported in a single case each of presumed meningioma³ and lymphoma.⁴ Chiasmal infarction is rare given extensive vascular collateralization. Two cases of restricted diffusion in the chiasm with multiple brain infarctions have been reported. One case included the optic nerve and was due to a posttraumatic internal carotid artery dissection,⁵ and the other occurred after cardiac surgery.⁶

Recognition and correct interpretation of the radiographic findings in our patient, which resulted from vascular occlusion and infarction due to malignant glioma, may have led to a diagnostic biopsy prior to extensive infiltration. We add this case to the rapidly growing literature on optic nerve infarction identified on DWI.

REFERENCES

1. Hoyt WF, Meshel LG, Lessell S, Schatz NJ, Suckling RD. Malignant optic glioma of adulthood. *Brain* 1973;96:121–132.
2. Spierer O, Sira LB, Leibovitch I, Kesler A. MRI demonstrates restricted diffusion in distal optic nerve in atypical optic neuritis. *J Neuroophthalmol* 2010;30:31–33.
3. Lope LA, Hutcheson KA, Khademian ZP. Magnetic resonance imaging in the analysis of pediatric orbital tumors: utility of diffusion-weighted imaging. *J AAPOS* 2010;14:257–262.
4. Sudhakar P, Rodriguez FR, Trobe JD. MRI restricted diffusion in lymphomatous optic neuropathy. *J Neuroophthalmol* 2011;31:306–309.
5. Ko SB, Lee K. Acute optic-nerve infarction in carotid dissection. *N Engl J Med* 2010;363:765.
6. Fabian ID, Greenberg G, Huna-Baron R. Chiasmal stroke following open-heart surgery. *J Neuroophthalmol* 2010;30:219–221.

ACKNOWLEDGMENT

The author thanks Wayne T. Cornblath, MD, for reviewing the manuscript.

STUDY FUNDING

No targeted funding reported.

DISCLOSURES

The author reports no disclosures. Full disclosure form information provided by the author is available with the **full text of this article at Neurology.org/cp**.