Junctional visual field loss: a reappraisal of nomenclature

Historically, junctional visual field loss (JVFL) has been associated with a lesion at the junction of the optic nerve and chiasm, and the most common etiologies are neoplasm and aneurysm.[1,2] We present 2 cases of JVFL and review the nomenclature on the various types of scotomas associated with lesions at the junction of the optic nerve and chiasm (e.g., junctional scotoma and junctional scotoma of Traquair). Based on our review of the English-language ophthalmic literature, we believe that our cases are unique and serve to demonstrate the need for expansion of the classification of JVFL.

The first case is a 32-year-old Peruvian man who presented with a 6-week history of headache and progressive blurred vision predominantly in his right eye that began 1 month after returning from a trip to Peru. Past medical, surgical, social, and ocular histories were noncontributory, and the patient was on no prior medications.

Examination revealed visual acuity of 20/80 OD and 20/50 OS, a right-sided relative afferent pupillary defect, and bilateral optic disc edema. Automated perimetry revealed diffuse depression OD and a temporal hemianopic visual field defect OS (Fig. 1), concerning for a lesion at the junction of the right optic nerve and optic chiasm. Magnetic resonance imaging (MRI) of the brain and orbit revealed abnormal leptomeningeal enhancement at the midbrain

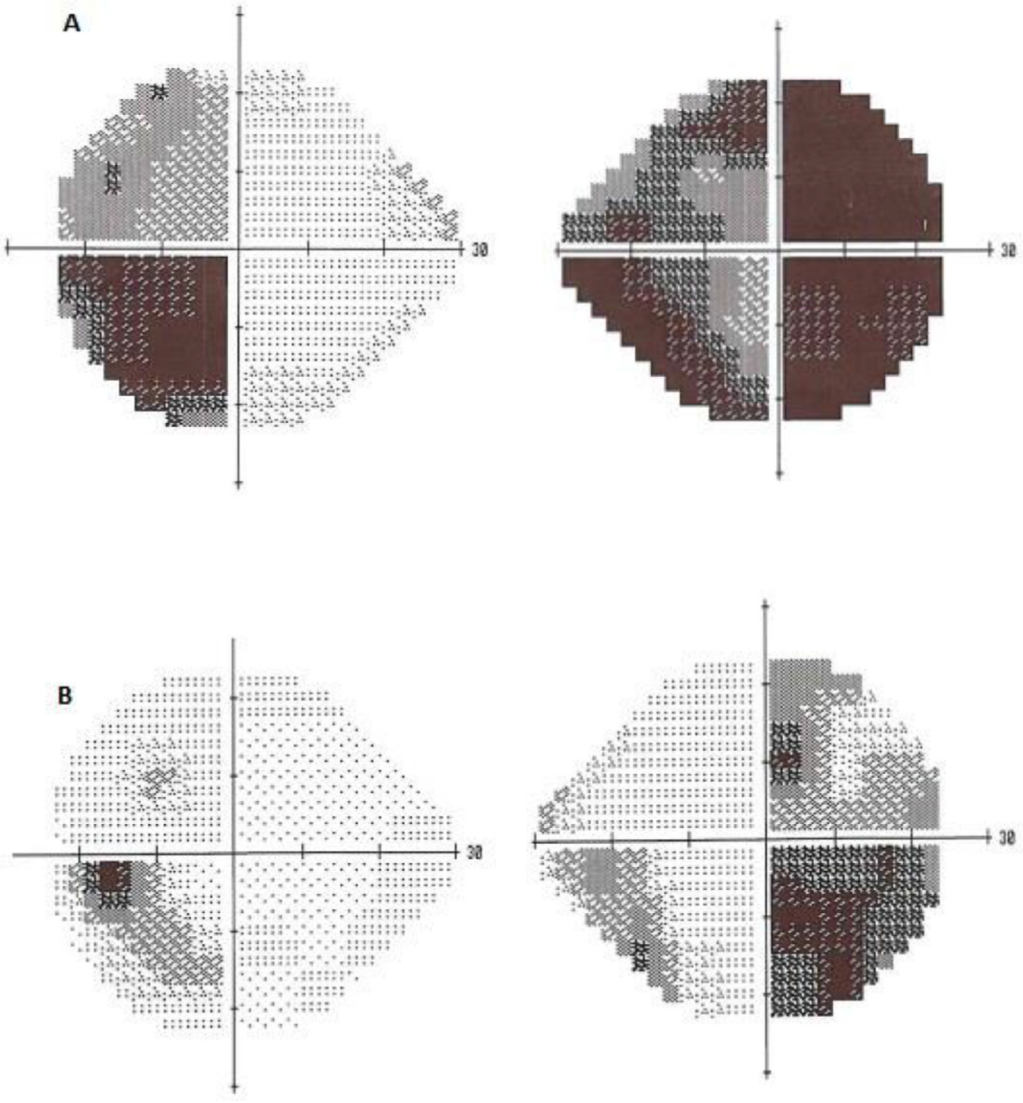

Fig. 1—Case 1. Humphrey visual field 24-2: (A) initial-onset junctional visual field loss with generalized depression OD and a dense temporal hemianopic loss OS; (B) 29-month follow-up showing overall improvement in the visual field results.

https://doi.org/10.1016/j.jcjo.2022.07.009
ISSN 0008-4182

Published by Elsevier Inc. on behalf of Canadian Ophthalmological Society.

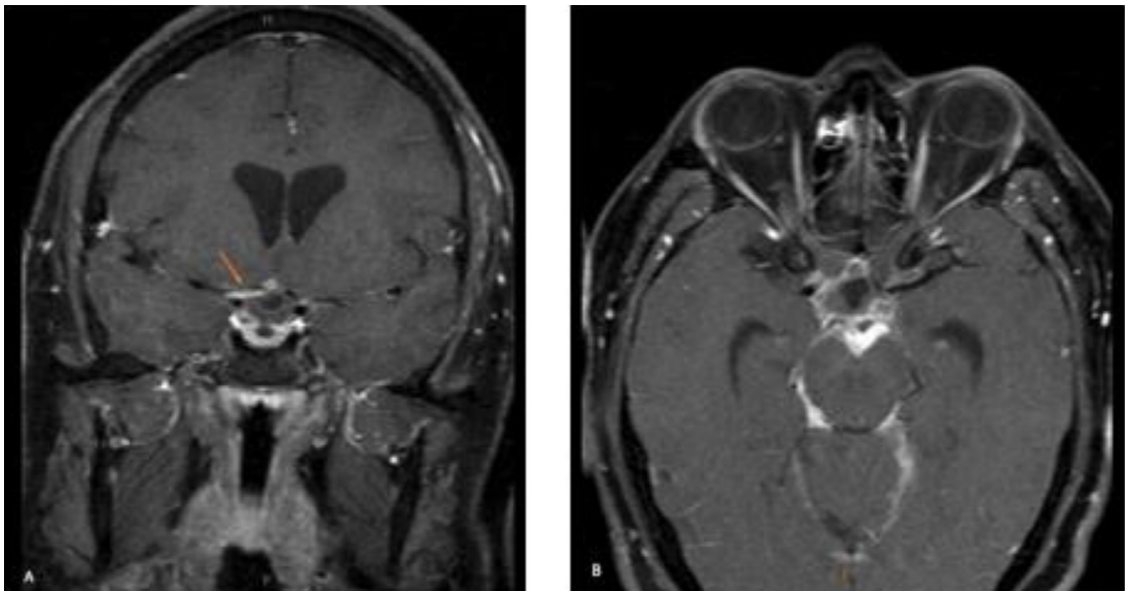

Fig. 2—Case 1. (A) Postcontrast coronal T_1-weighted magnetic resonance image with abnormal enhancement around the right optic nerve near the optic chiasm (arrow). (B) Postcontrast axial image depicting leptomeningeal enhancement and edematous changes in the surrounding midbrain area.

and right optic nerve close to the optic chiasm. There was mild ventriculomegaly and a suprasellar cystic lesion with mild mass effect on the optic chiasm as well as an area of calcification in the left parietal lobe (Fig. 2). Initial cerebrospinal fluid (CFS) studies revealed a white blood cell count of 295 cells/μL (normal, <5 cells/μL) with zero neutrophils, 76% lymphocytes (normal,40%−80%), 23% monocytes (normal, 15%−45%), and 1% eosinophils (normal, <1%). CSF glucose was 23 mg/dL (serum, 107 mg/dL), and CSF protein was 130 mg/dL (normal, 15−45 mg/dL). The CSF was positive for cysticercosis antibody IgG at 7.59 (normal, <0.75). The diagnosis of neurocysticercosis was made, and the patient was treated accordingly. His medication regimen included albendazole, praziquantel, acetazolamide, and corticosteroids. He underwent a ventriculoperitoneal shunt. Repeat testing revealed improvement of visual acuity to 20/25 OD and 20/20 OS, and MRI brain showed resolution of the ventriculomegaly, leptomeningeal enhancement, and suprasellar cyst seen on initial imaging.

The second case was a 59-year-old white woman who developed rapidly progressive bilateral vision loss. Examination revealed visual acuity of 20/20 OD and counting fingers OS. Automated perimetry revealed a central scotoma OS with an inferotemporal depression and a contralateral temporal hemianopic visual field defect OD (Fig. 3). Neuroimaging findings revealed extensive diffuse demyelinating plaques with multiple acute enhancing plaques (Fig. 4). No specific lesions were identified at the junction of the optic chiasm and optic nerve. A diagnosis of multiple sclerosis was made. Intravenous corticosteroids were administered with improvement. Disease-modifying treatments were offered to the patient, but she was lost to follow-up.

JVFL can be caused by a lesion at the junction where the optic nerve meets the optic chiasm. The classic *junctional scotoma* refers to an ipsilateral central scotoma with a contralateral superior temporal hemianopic field defect caused by a lesion affecting the crossing inferonasal fibres from the contralateral eye. In the past, this crossing nasal fibre was referred to as the *Wilbrand knee*, but the existence of this anatomic inferonasal crossing fibre remains controversial.[2] Regardless of whether the Wilbrand knee exists anatomically or not, the localizing value of JVFL to the junction of the optic nerve and chiasm remains valid, and neuroimaging should be directed to this location. The etiologies for JVFL include aneurysm, neoplasm, an infiltrative process, an inflammatory process, an infectious process, demyelination, traumata, and degenerative disease. In contrast to the junctional scotoma (above), the junctional scotoma of Traquair is a monocular hemianopic visual field defect, typically from compression of the crossing nasal fibre at the junction of the optic nerve and chiasm. Some patients, however, as in our 2 cases, have ipsilateral optic neuropathy−type visual fields (including central and arcuate or altitudinal visual field loss) and contralateral complete temporal hemianopic loss. These types of JVFL, while neither junctional scotoma nor junctional scotoma of Traquair, also localize to the junction of the optic nerve and chiasm. We propose the term *junctional visual field loss* to encompass the possible combinations of optic neuropathy and contralateral temporal hemianopic loss.

Neurocysticercosis is a parasitic infection caused by the laraval stage of the tapeworm *Taenia solium*. Cysticercosis is transmitted by ingestion of *T. solium* eggs that are shed in the stool of a human tapeworm carrier; after ingestion, the

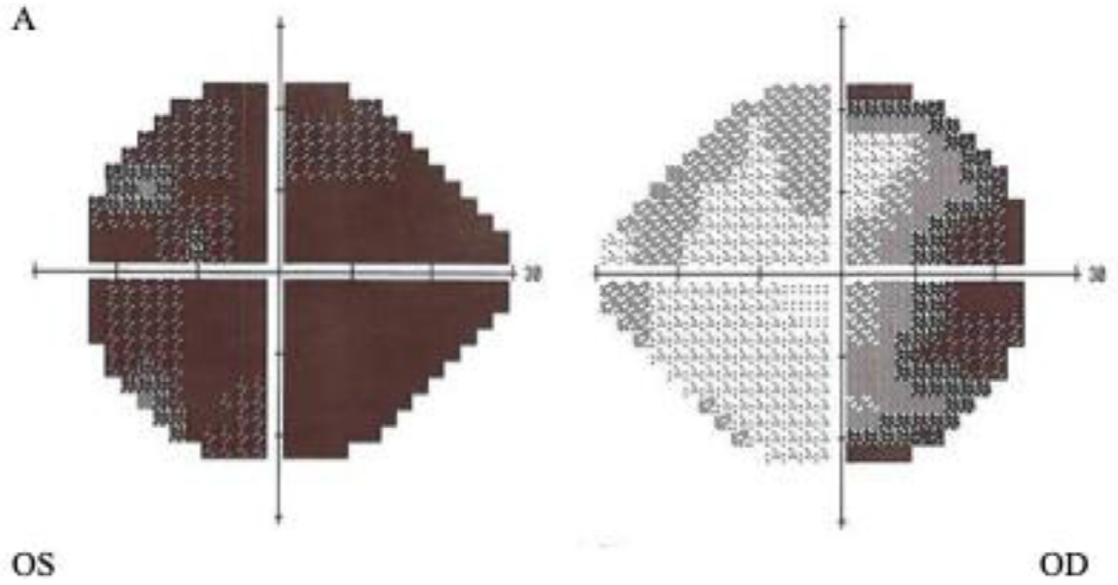

Fig. 3—Case 2. Humphrey visual field 24-2 showing dense generalized depression OS with a contralateral temporal hemianopic defect OD consistent with junctional visual field loss.

embryos hatch in the small intestine and are subsequently able to disseminate hematogenously. Neurologic manifestations can occur if the blood–brain barrier is disrupted. Ocular and orbital cysticercoses have been described leading to proptosis, ptosis, lid edema, diplopia, impaired visual acuity, and orbital cellulitis.[3] Neurocysticercosis, which is the most prevalent parasitic infection of the nervous system, is endemic to most of the world and contributes significantly to the burden of epilepsy and other neurologic morbidity.[4]

Multiple sclerosis (MS) is the most prevalent immune-mediated inflammatory demyelinating disease of the CNS. The majority of patients with MS have a relapsing-remitting disease course with a typical presentation of a clinically isolated syndrome such as optic neuritis, dysesthesias, paresis, or brainstem or spinal cord syndromes. When considering the neuro-ophthalmologic manifestations of MS, the more common abnormalities include vision loss due to optic neuritis, diplopia owing to internuclear ophthalmoplegia, skew

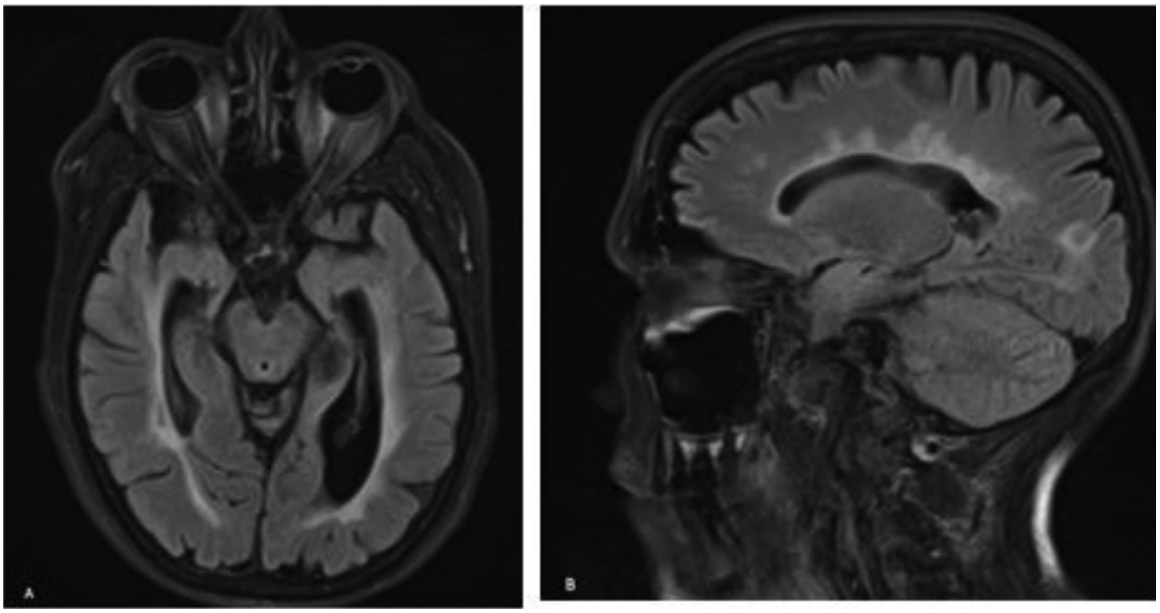

Fig. 4—Case 2. Magnetic resonance imaging of the brain with and without contrast material: (A) Fluid-attenuated inversion recovery axial image depicting bilateral optic atrophy; (B) fluid-attenuated inversion recovery sagittal image depicting periventricular and juxtacortical demyelinating lesions.

deviation, or other ocular motor nerve palsies, nystagmus, and paroxysmal disorders such as ocular flutter, opsoclonus, and square-wave jerks.[4]

Clinicians should be aware of both junctional scotoma and junctional scotoma of Traquair. In addition, any combination of ipsilateral optic neuropathy—type visual loss (with or without loss of central visual acuity) and a contralateral temporal hemianopic visual field defect also constitutes a form of JVFL localizing to the junction of the optic nerve and chiasm. Neuroimaging should be directed to this location in any case of JVFL.

Jasmine Chopra,* Alan Arismendez,* Jay Mahajan,* Aroucha Vickers,* Andrew G. Lee[†,‡,§,‖,¶,**,††,‡‡]

*Valley Hospital Medical Center, Las Vegas, Nev.; [†]Blanton Eye Institute, Houston Methodist Hospital, Houston, Tex.; [‡]Weill Cornell Medicine, New York, NY; [§]University of Texas Medical Branch at Galveston, Galveston, Tex.; [‖]Texas A&M University School of Medicine, Bryan, Tex.; [¶]University of Texas MD Anderson Cancer Center, Houston, Tex.; **Baylor College of Medicine, Houston, Tex.; [††]University of Buffalo, Buffalo, NY; [‡‡]University of Iowa Hospitals and Clinics, Iowa City, Iowa.

Originally received Jun. 15, 2022. Accepted Jul. 28, 2022.

Correspondence to
Jasmine Chopra, DO; jasminechopra@yahoo.com.

References

1. Bird A. Field loss due to lesions at the anterior angle of the chiasm. Proc R Soc Med 1972;65:519–20.
2. Garcia HH, Gonzalez AE, Gilman RH. *Taenia solium* cysticercosis and its impact in neurological disease. Clin Microbiol Rev 2020;33 e00085-19.
3. Dhiman R, Devi S, Duraipandi K, et al. Cysticercosis of the eye. Int J Ophthalmol 2017;10:1319–24.
4. Olek MJ, Frohman TC, Frohman EM, Narayan RN.*Manifestations of multiple sclerosis in adults*. UpToDate [Internet]. N.d. Available at: https://www.uptodate.com/contents/manifestations-of-multiple-sclerosis-in-adults#! (accessed June 1, 2022).

Footnotes and Disclosure

The authors have no proprietary or commercial interest in any materials discussed in this correspondence.