

## Diagnose and Manage Pseudo–Foster Kennedy Syndrome

BY LAURA A. VICKERS, MD, M. TARIQ BHATTI, MD, AND MAYS A. EL-DAIRI, MD  
EDITED BY SHARON FEKRAT, MD, AND INGRID U. SCOTT, MD, MPH

**F**oster Kennedy syndrome was first described in 1911 by the neurologist Robert Foster Kennedy, who characterized the disorder as compression of one optic nerve by a subfrontal meningioma, resulting in optic nerve head pallor, with increased intracranial pressure causing contralateral optic nerve head edema.<sup>1</sup>

True Foster Kennedy syndrome is rare, but other causes of unilateral disc edema with contralateral optic pallor are more common. These have been termed pseudo–Foster Kennedy syndrome, or PFK. This review will present a recent case report of PFK and discuss differential diagnosis and management of the condition.

### Case Report

A 67-year-old woman presented to our institution, complaining of a 30-day history of headache that registered 10 on a 10-point scale of severity and a two-week history of decreased vision in her left eye. Her left optic nerve was swollen (Figs. 1A and 1B), and her erythrocyte sedimentation rate (ESR) was 9.

Initially, she was started on oral prednisone. The swelling of the left optic nerve resolved in four weeks and was replaced by pallor. At the five-week mark, when results from a left temporal artery biopsy were normal, the prednisone was tapered off.

One year later, the patient presented with scalp tenderness and jaw claudication. The optic nerve of her right



eye was mildly swollen, and the optic nerve of her left eye was pale superiorly (Figs. 2A and 2B). At this time, her ESR was 20. Because of her well-documented history of unilateral optic nerve head edema replaced by pallor in four weeks, neuroimaging was not performed. However, because of our persistent high clinical suspicion for giant cell arteritis (GCA), a right tem-

poral artery biopsy was performed. This demonstrated healing arterial wall inflammation with intimal elastic degeneration. The diagnosis: GCA presenting as PFK.

### Diagnosis and Management

PFK is a diagnosis of exclusion. When a clinician encounters a patient with one swollen and one pale optic nerve,

neuroimaging should routinely be obtained, especially if the history is vague or not typical for anterior ischemic optic neuropathy (AION).

Bilateral sequential anterior ischemic optic neuropathies, either arteritic (AAION) or nonarteritic (NAION), are the usual culprits in PFK, with NAION the most common etiology.

**NAION.** This occurs most frequently in middle-aged patients, with men and women affected equally.<sup>2</sup> On examination, the swollen nerve is likely to display hyperemic edema with disc hemorrhage, segmental pallor, and attenuated blood vessels. ESR, C-reactive protein (CRP), and platelet levels are normal.

**Management.** In coordination with the patient's primary care provider, testing and modification of vascular risk factors should be undertaken. No other interventions, including optic nerve sheath fenestration, steroids, aspirin, or intravitreal injections of anti-vascular endothelial growth factor medications, have proved effective.

**AAION.** In cases of AAION, the swollen nerve—the one affected more recently—often displays pallid edema. On average, patients with AAION are older, more commonly female, and have worse visual outcomes than NAAION patients with rare late improvement. These patients commonly have elevated ESR, CRP, and platelet levels. A 2- to 3-cm temporal artery biopsy with pathologic examination of consecutive slices spaced at 0.25 cm is the gold standard for diagnosis.

**Management.** In all patients older than 50 years of age presenting with signs of ischemic optic neuropathy, laboratory testing for GCA should be obtained. (This should include a complete blood count as well as the ESR and CRP tests.) If the patient is also complaining of headaches or other symptoms of GCA, or if the patient has an abnormal ESR or CRP level, systemic steroids should be started immediately, and a temporal artery biopsy performed within two weeks.

When the clinician has persistent high clinical suspicion for AAION despite negative laboratory testing and a

negative temporal artery biopsy on the more symptomatic side, biopsy of the contralateral temporal artery is recommended. When a patient presents with edema of the contralateral nerve after an initial AION, the diagnosis of AAION should be reconsidered even if testing was previously negative.<sup>3</sup> There is a false-negative rate of up to 5 percent for biopsy of the initially affected side. False-negative results have been attributed to inadequate length of biopsy specimen, skip lesions, errors in specimen sectioning, and prebiopsy treatment with steroids.<sup>4</sup>

## Uncommon Etiologies

PFK also may be observed less commonly in the following settings.

**Retrobulbar neuritis.** In this instance, the optic nerve head usually appears normal on examination. However, bilateral sequential optic neuritis may present with optic nerve head edema in the acutely affected eye.

**Chronic unilateral optic atrophy.** PFK may be seen in patients with this condition, regardless of its cause, and who later present with acute optic nerve swelling on the contralateral side. Potential etiologies include traumatic optic neuropathy, prior optic neuritis, compressive optic neuropathy, and a history of meningitis.

**Additional etiologies.** There are some rare reports of PFK occurring in younger individuals. In these patients, the etiology of PFK may include pachymeningitis related to microvasculitis that tests positive for perinuclear antineutrophil cytoplasmic antibodies (p-ANCA), idiopathic intracranial hypertension, or unilateral optic nerve hypoplasia. Additionally, there are reports of patients with increased intracranial pressure and papilledema who develop unilateral optic atrophy and thus present with PFK.

## A Note on Follow-up

Follow-up is essential; the clinician should observe the affected optic nerve for four to eight weeks to be sure that the swelling is resolving and is being replaced by pallor.<sup>2</sup>

There have been case reports of

patients who were initially diagnosed with bilateral sequential NAION and later proved to have a compressive lesion with development of neurologic symptoms.<sup>5</sup> As a result, clinicians should have a very low threshold of suspicion and should order additional testing, including neuroimaging, if there is any question about the etiology of PFK.

## Conclusion

The differential diagnosis of the clinical finding of a pale optic nerve with contralateral optic nerve head edema is broad. Most commonly, PFK is due to bilateral sequential NAION, but clinicians should have a low threshold for ordering neuroimaging and a laboratory workup for GCA and for performing an assessment for increased intracranial pressure. ■

*Dr. Vickers is a third-year ophthalmology resident, Dr. Bhatti is associate professor of ophthalmology and neurology and director of the neuro-ophthalmology service, and Dr. El-Dairi is assistant professor of ophthalmology; all are at Duke University Eye Center in Durham, N.C. The authors report no related financial interests.*

1 Kennedy F. *Am J Med Sci.* 1911;142:355-368.

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4 Pless M et al. *J Neuroophthalmol.* 2000;20(3):216-218.

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