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Grand Rounds

A 44-year-old woman with a 3-month history of bilateral, painless visual loss in the absence of other symptoms

Emily H. Shao, MBBS, BSC, Kevin Gallagher, MBBS, and **Nabeel Malik, MBBS**
Author affiliations: Department of Ophthalmology, Chelsea, and Westminster Hospital, London, United Kingdom

History

A 44-year-old woman presented at the Western Eye Hospital, London, with a 3-month history of gradual, painless, bilateral deterioration of her vision. She had no other ocular symptoms and no previous ocular history. Review of systems was unrevealing, and her medical history was unremarkable: she did not complain of headaches or anosmia, nor was she taking any regular medication. She was of normal body habitus. Family history was non-contributory.

Examination

On examination, visual acuity was hand movements in the right eye and no light perception (NLP) in the left eye. There was no relative afferent papillary defect. Pupils were round and reactive to light. The anterior segments were normal. On dilated fundus examination, she was found to have a grossly swollen right optic disc (Figure 1) and a pale left optic disc (Figure 2).

Imaging

Computed tomography (CT) of the head and orbits revealed a frontal mass extending into the planum sphenoidale and the pituitary fossa with the appearance of a large anterior cranial fossa floor meningioma (Figure 3). No abnormality was seen in the globes, optic nerves, extraocular muscles, or any intraconal structure in either orbit (Figure 4).

Treatment

The patient was referred to the neurosurgical team for review, and debulking of the lesion was recommended. Because she was a foreign national and thus ineligible for national health treatment, she had to return to her home country for treatment.

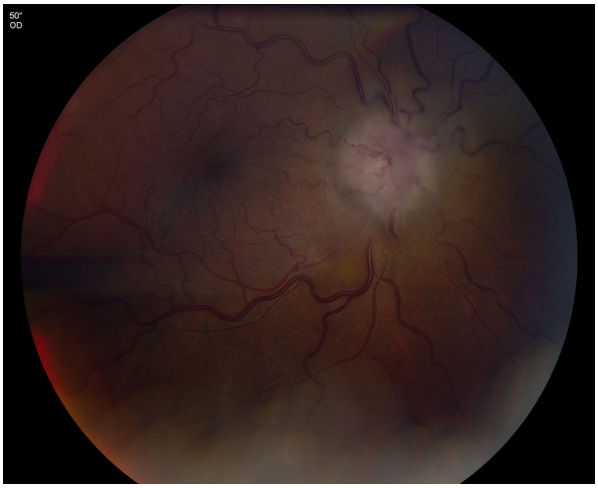

Figure 1. Papilledema of the right optic disc.

Differential Diagnosis

The patient was noted to have a swollen optic disc in one eye and an atrophic optic disc in the other eye. Such a combination of signs is rare and may be caused by a variety of pathological processes.

Foster Kennedy syndrome (FKS) consists of a clinical triad of optic atrophy in one eye, papilledema in the contralateral eye, and anosmia, caused by space-occupying anterior fossa masses.[1] Associated symptoms of raised intracranial pressure, including nausea, vomiting, and headaches, and frontal lobe signs may be present, including emotional lability and memory loss.

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Correspondence: Nabeel Malik MBBS, FRCOPHTH, Chelsea and Westminster Hospital, 369 Fulham Road, London SW10 9NH (email: nabeel-malik.eyes@gmail.com)

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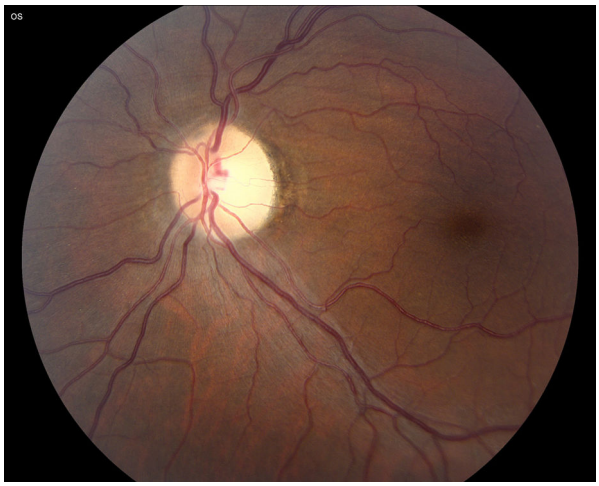

Figure 2. Left optic disc pallor.

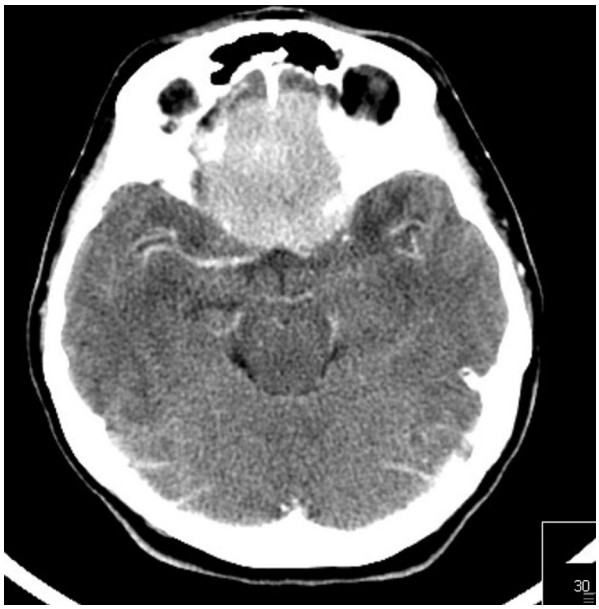

Figure 3. Computed tomography (CT) of the head showing a well-circumscribed homogenous lesion (4.6 × 4.4 × 2.5 cm) in the midline of the frontal region with surrounding edema consistent with meningioma.

In pseudo-FKS, optic disc pallor and contralateral disc swelling is present but is not caused by an intracranial mass. This may be caused by the same pathological process in each eye. For example, patients with bilateral sequential ischemic optic neuropathy or optic neuritis may have one pale, atrophic optic disc and a papillitis in the contralateral optic disc, mimicking the papilledema seen in FKS. Alternatively, two pathological processes may be responsible: any cause of unilateral optic disc

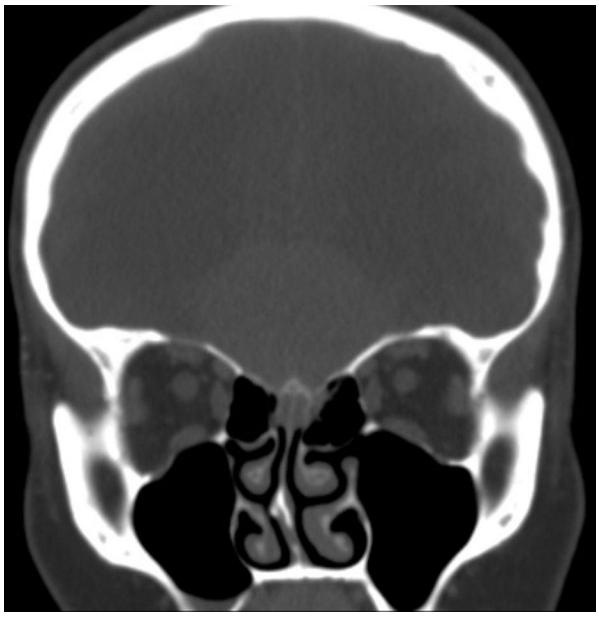

Figure 4. CT of the orbits showing no abnormality involving the globes, optic nerves, extraocular muscles, or any intraconal structures in either orbit; an intracranial mass is seen in the frontal region consistent with meningioma in the coronal view.

pallor (eg, previous unilateral ischemic optic neuropathy or optic neuritis; unilateral optic nerve hypoplasia) in conjunction with raised intracranial pressure causing papilledema in the contralateral eye will give rise to this combination of signs.[2] Papilledema does not occur in optic atrophy because the loss of neural fibers in optic atrophy results in a lack of neurons to swell.

Diagnosis

On presentation the patient had a 3-month history of visual loss, optic atrophy, and optic disc swelling. She did not have any symptoms or signs of raised intracranial pressure, or symptoms suggestive of demyelination or infection. CT scan of the head showed an intracranial mass. She was diagnosed with FKS, a rare neurological syndrome first described by Foster Kennedy in 1911.[1] Found in less than 1% of patients with intracranial tumors, the pathogenesis of FKS is still debated.[3] It was postulated to be caused by direct tumor compression, resulting in optic atrophy in one eye with raised intracranial pressure causing papilledema in the contralateral eye; however, bilateral optic nerve compression has been found subsequently in 33% cases, with only 22% satisfying Kennedy's original hypothesis.[4] The clinical course of FKS is frequently insidious in nature. Headache suggestive of raised intracranial pressure (ICP) may not be present, making early diagnosis difficult.[5]

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FKS may present with a relative afferent pupillary defect early on if one optic nerve is predominantly affected. Chronic papilledema may lead to nerve fiber atrophy, a degree of which may already have existed in our patient, which would explain the absence of relative afferent pupillary defect on examination. As in any cause of optic nerve dysfunction, reduced color vision is an early feature. Visual field defects, including a central scotoma, are typically present. In patients presenting with decreased visual acuity, testing of color vision and pupillary reflexes should be performed in addition to fundus examination. The clinical signs of FKS may be readily apparent in cases of gross optic disc swelling or pallor; however, optic disc pallor may be extremely subtle and easily missed.

Poorly understood, with a currently debated pathogenesis and often insidious onset, FKS remains a diagnostic challenge. Early diagnosis, with subsequent surgical resection may prevent further visual deterioration and may allow some recovery of vision in these patients.[6] Disease awareness with early intracranial imaging and surgery is necessary to minimize the disabling sequelae of FKS.

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