

Foster Kennedy Syndrome: Revisiting a Classical Phenomenon

Síndrome de Foster Kennedy: Revisando un Fenómeno Clásico

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Abstract

Foster Kennedy Syndrome is a classic, yet rare, neuro-ophthalmologic syndrome due to an intracranial mass, most often a tumor, that consists of optic atrophy on the same side of the lesion and contralateral papilledema. We present the case of a 48-year-old female patient with decreased visual acuity and the typical clinical features described above due to a sphenoid wing meningioma. Although not a common condition, Foster Kennedy Syndrome should always be kept in mind in a patient with visual disturbances secondary to an intracranial mass.

Key words: *Foster Kennedy Syndrome, optic atrophy, papilledema, meningioma, case report.*

Resumen

El síndrome de Foster Kennedy es un síndrome neuro-oftalmológico clásico, pero poco común debido a una masa intracraneal, por lo general un tumor, que consiste en atrofia óptica del mismo lado de la lesión y papiledema contralateral. Presentamos el caso de una paciente femenina de 48 años con disminución de la agudeza visual y las manifestaciones clínicas típicas descritas anteriormente, debido a un meningioma del ala esfenoidal. Aunque no es un trastorno común, el síndrome de Foster Kennedy siempre debe ser considerado en un paciente con alteraciones visuales secundarias a una masa intracraneal.

Palabras clave: *síndrome Foster Kennedy, síndrome neuro-oftalmológico, papiledema, meningioma, reporte de caso.*

Introduction

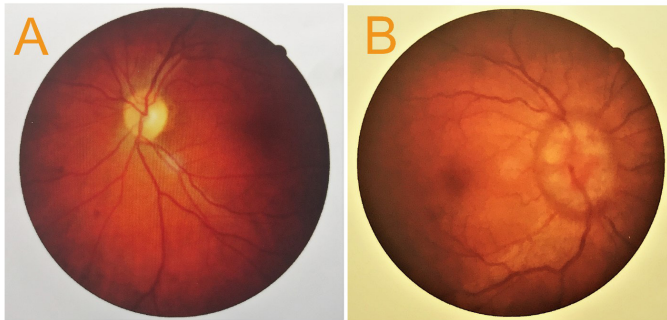
Foster Kennedy syndrome was first described in 1911 by Robert Foster Kennedy, a Northern Irish neurologist, in a 37-year-old woman with headaches, vomiting, anosmia and decreased visual acuity in her left eye. After funduscopic examination, left optic atrophy and right-side papilledema were observed by Foster Kennedy. The patient had a large olfactory groove meningioma, more on the left than on the right side, as the autopsy later revealed.¹ It has been proposed that the optic atrophy is caused by either direct compression of the optic nerve by the intracranial mass or ischemic damage to its axons. Papilledema in this context would be a consequence of increased intracranial pressure.²⁻⁴ Ever since, only a handful of cases displaying the classic neuro-ophthalmologic features due to an intracranial mass have been reported, most of them secondary to meningiomas.¹⁻³ We present the case of a 48-year-old female patient who presented with the typical clinical features described above with a contrast-enhanced head computed tomography revealing a left sphenoid wing meningioma.

Case Report

A 48-year-old woman with previous history of arterial hypertension presented with a three-month clinical picture of decreased visual acuity affecting both eyes, particularly the left eye, and headache.

On ophthalmologic examination, the right eye (OD) could perceive only finger motions, with no light perception on the left eye (OS). She also had a left afferent pupillary defect. Fundusoscopic examination revealed papilledema on the right side, with left optic disc atrophy (Figure 1).

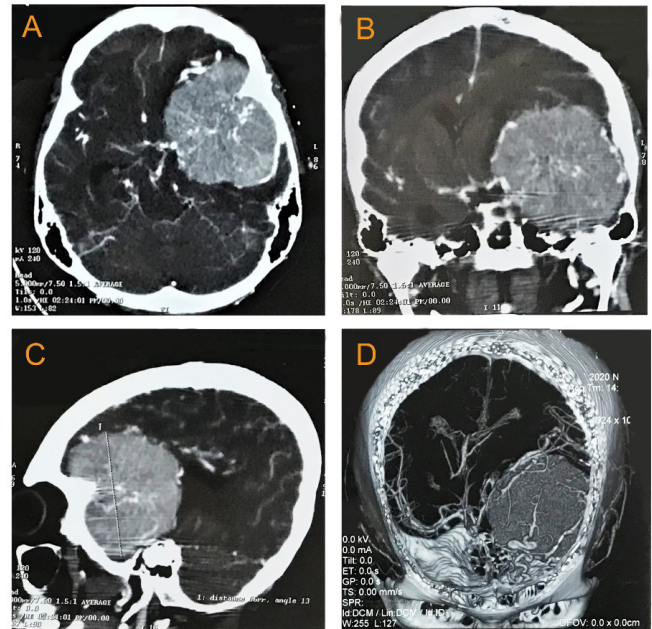
Figure 1. Classic ophthalmoscopic findings of Foster Kennedy Syndrome. **A)** Left optic disc atrophy. **B)** Right optic disc edema.



The rest of the neurologic examination was unremarkable. A provisional diagnosis of Foster Kennedy syndrome was made and a contrast-enhanced head computed tomography (CT) was performed. It showed a rounded, well circumscribed, extra-axial homogenous and hyperdense mass lesion with regular borders over the left sphenoid wing, compromising the ipsilateral optic nerve. The mass had surrounding hyperdense cerebral edema with displacement of the midline structures, absence of sulci and compression of the frontal ventricular horns (Figure 2). The radiological findings were highly suggestive of left sphenoid wing meningioma.

Laboratory studies including complete blood count, serum chemistry, electrolytes, and liver function tests were normal. She was referred for neurosurgical evaluation, however, due to the COVID-19 pandemic she was not able to attend it. Two months after the initial evaluation the patient presented to the emergency department with decreased mental status, vomiting and a generalized convulsive tonic-clonic seizure. The patient was admitted for urgent advanced airway management and neurosurgical consultation. A new head CT scan was requested, revealing the previously described left sphenoid wing meningioma with increased mass effect and edema,

Figure 2. Imaging findings. **(A-C)** Contrast-enhanced head CT in axial, coronal and sagittal views showing a rounded, well circumscribed, extra-axial homogenous and hyperdense mass lesion with regular borders over the left sphenoid wing. **(D)** 3-D reconstruction demonstrating a left sphenoid wing meningioma.



plus left uncus and subfalcine herniation. Antiedema therapy was started with intravenous dexamethasone and mannitol. While awaiting urgent surgery in the Intensive Care Unit (ICU), the patient's condition deteriorated with absent corneal, palpebral and cough reflexes, despite sedation being withdrawn, and requiring vasopressor support. Eventually the patient passed away before surgical intervention could be carried out. No autopsy was performed due to COVID-19 pandemic biosafety protocols.

Discussion

Foster Kennedy Syndrome, named after neurologist Robert Foster Kennedy (1884–1952), refers to the combination of unilateral optic disc atrophy caused by an ipsilateral intracranial tumor, usually in the frontal lobe, and contralateral optic disc edema secondary to increased intracranial pressure.^{2,3}

Although described as a classic neuro-ophthalmologic syndrome, it is rarely seen in clinical practice, presenting in only 1% to 2.5% of intracranial masses.^{1,4} Out of 36 cases in the literature, only 8 (22%) had the true Foster Kennedy syndrome.¹

Regarding its pathophysiology, it has been postulated that the optic atrophy is due to direct compression of the optic nerve by the mass, or by vascular compression and subsequent ischemia (optic disc axons do not regenerate) and contralateral edema due to the raised intracranial pressure.^{2,4}

It can be classified into three types depending on the ophthalmologic signs seen in funduscopy: Type 1: optic atrophy on one side with contralateral papilledema, Type 2: bilateral papilledema developing unilateral optic atrophy and Type 3: bilateral papilledema developing bilateral optic atrophy.¹

The majority of the reported cases are caused by meningiomas of the olfactory groove, sphenoidal wing or sub-frontal regions. They are usually observed in patients between 40 and 60 years of age and are more frequent among women. Although focal symptoms and signs may be sparse, nonspecific symptoms such as nausea, headache and personality changes have been reported.^{2,4}

Our case displayed the classic clinical features of Foster Kennedy syndrome due to one of its most common causes: a sphenoid wing meningioma. Unfortunately, the patient could not attend to proper neurosurgical evaluation on time due to restrictions secondary to the COVID-19 pandemic and eventually deteriorated very quickly before a surgical intervention could be performed.

Conclusion

In this case reports highlights the importance of timely diagnosis and management of a classic, yet rare, neuro-ophthalmologic syndrome which in most instances its produced by treatable neurosurgical causes. It is important that both ophthalmologists and neurologists keep a low threshold for this condition when evaluating a patient with headache and visual loss who presents with optic atrophy and papilledema.

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Artículo sin conflicto de interés

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