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A Rare Clinical Portrait: Unveiling the Complexities of Foster-Kennedy Syndrome

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ABSTRACT

Introduction: Foster-Kennedy Syndrome (FKS) is an uncommon neuro-ophthalmological disorder characterized by unilateral blindness linked with an intracranial tumor. Its defining features, such as unilateral optic disc atrophy and contralateral papilledema, are identifiable through ophthalmoscopy. We present a case of FKS caused by a meningioma.

Case Presentation: A 45-year-old woman reported progressively deteriorating vision over the past year, accompanied by severe headaches, nausea, vomiting, and recurrent convulsions. Ophthalmic examination revealed no light perception (NLP) in the right eye (RE) and finger counting ability in the left eye (LE). The Marcus Gunn Pupil was positive in the RE. Optic atrophy in the RE and papilledema in the LE were observed, consistent with type 1 FKS. A head computed tomography scan showed a sizable tumor suspected to be a meningioma in the right frontotemporal lobe. The patient received a combination of medical and surgical treatments.

Discussion: FKS is a collection of findings on fundoscopy associated with frontal lobe tumors and is marked by ipsilateral optic nerve atrophy caused by direct compression of tumor on the optic nerve and contralateral papilledema due to increasing intracranial pressure. The incidence of true FKS has decreased in recent years due to advancements in neuroimaging, allowing for earlier detection of intracranial masses.

Conclusion: Funduscopic examination is crucial in patients presenting with visual disturbances and headaches. When FKS is suspected, neuroimaging should be performed to confirm the presence of intracranial mass. Early diagnosis and appropriate management are essential for preserving visual function and addressing the underlying cause.

INTRODUCTION

Foster-Kennedy Syndrome (FKS) is a rare neuro-ophthalmological condition associated with an intracranial tumor causing unilateral blindness.(Walia et al., 2012) The optic nerve is the only visible extension of the central nervous system (CNS) that can be directly observed using ophthalmoscope. Despite the clinical benefits of fundoscopy, it remains underutilized in routine clinical practice.(He et al., 2022). The hallmark features of FKS involve the asymmetric involvement of the optic nerves: one side demonstrates optic atrophy, often as a result of direct compression from the mass, while the contralateral side develops papilledema due to increased intracranial pressure. Foster-Kennedy Syndrome (FKS) is first identified by an Irish neurologist

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Robert Foster Kennedy in 1911. His significant contributions to neurology led to the discovery of a distinctive set of clinical symptoms, which later became known as Foster-Kennedy Syndrome. This syndrome is marked by unilateral optic atrophy, which arises from the direct compression of the optic nerve by an intracranial mass. Additionally, it is accompanied by contralateral papilledema, caused by increased intracranial pressure. The description of FKS provided a significant advancement in the field of neuro-ophthalmology, particularly in the era before the advent of radiological neuroimaging. Early recognition is crucial to avoid irreversible visual loss and to address the underlying pathology before significant neurological deterioration occurs (Singh et al., 2019). We report a case of Foster-Kennedy Syndrome (FKS), an extremely rare condition, described by a collection of findings on fundoscopy associated with intracranial mass, such as a tumor, that compresses the optic nerve on one side, leading to optic atrophy, while simultaneously increasing intracranial pressure, which results in papilledema in the contralateral eye. (Musa et al., 2022; Visvanathan & Velmurugan, 2020)

CASE PRESENTATION

A 45-year-old woman experienced progressively worsening blurred vision over the course of a year, accompanied by severe headaches, nausea, vomiting, and multiple episodes of tonic-clonic seizures. Her hemodynamic status was stable. Ophthalmological assessment showed that visual acuity in the right eye (RE) was no light perception (NLP), while the left eye (LE) was 6/60. The relative afferent pupillary defect (RAPD) was positive in the RE. A fundoscopic examination of the right eye revealed a well-defined pale optic disc. Meanwhile, the left eye showed a poorly defined margin and hyperemic optic disc with surrounding edema. A computed tomography (CT) scan was performed (Figure 2.) and showed a mass in the right frontotemporal lobe with perifocal edema surrounding it exerting pressure on the right lateral ventricle, resulting in a midline shift to the left measuring 1.19 cm. The laboratory tests and hemostasis function results showed normal findings. The patient was treated with intravenous dexamethasone (3x5 mg), citicoline (2x1 g), and paracetamol (3x1 g), and oral levetiracetam (2x500mg). The patient was then evaluated and treated by the neurology and neurosurgery division and a craniotomy for tumor removal was planned. The description was consistent with type 1 FKS.

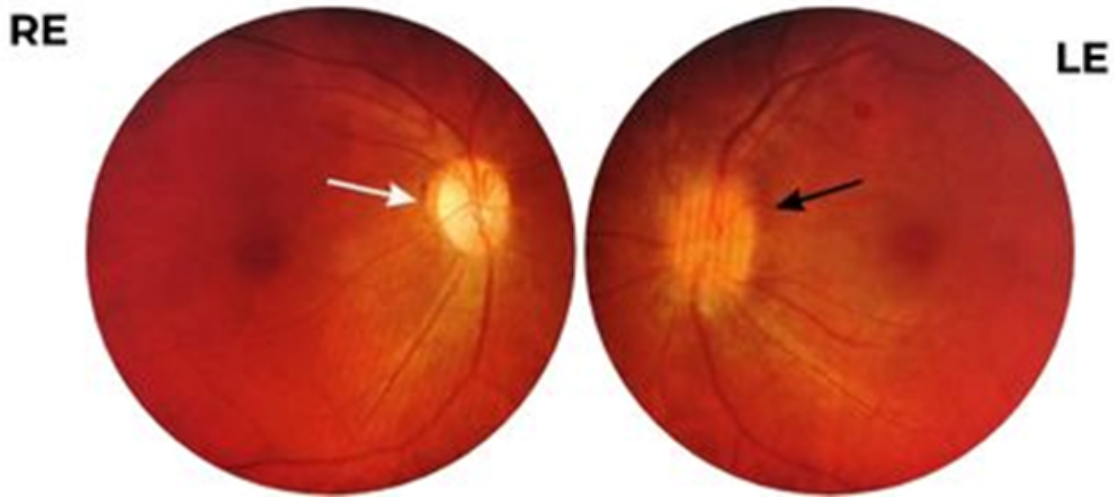

Figure 1. Fundus photographs of both eyes.
Pale optic disk of the RE (white arrow) and papilledema of the LE (black arrow)

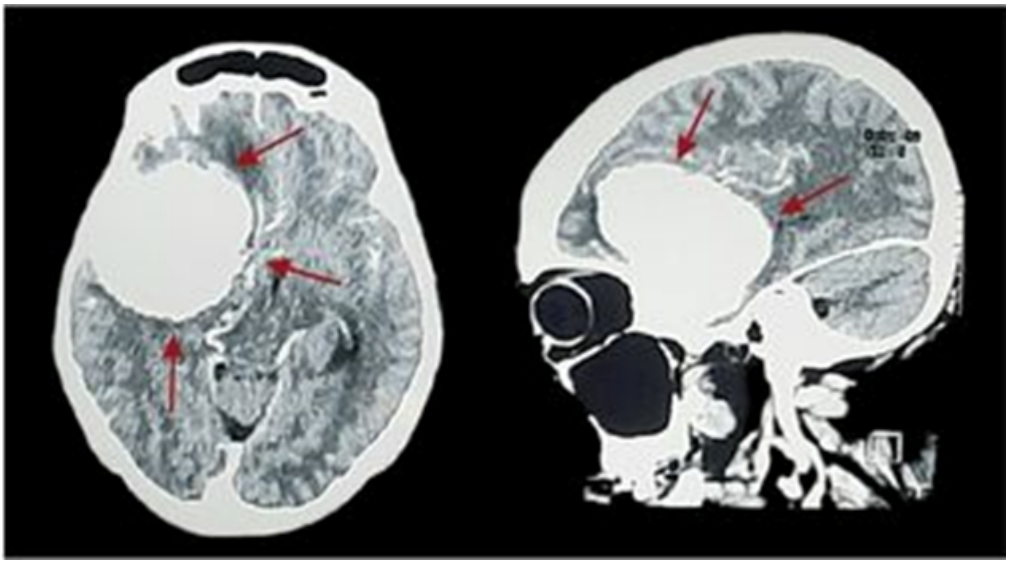

Figure 2. Head CT Scan Images in Axial and Sagittal View.
A mass (red arrow) in the right frontotemporal lobe with perifocal edema surrounding it exerting pressure on the right lateral ventricle, resulting in a midline shift to the left measuring 1.19 cm.

DISCUSSION

The prevalence of FKS in patients with intracranial neoplasms is notably low, accounting for less than 1%. The most extensive study on this topic was conducted by Tonnis in Germany in 1962, which identified 28 cases of FKS among 3,033 patients with intracranial tumors, resulting in an incidence rate of approximately 0.9%. The prevalence of FKS is influenced by the underlying causes, with meningiomas being the most common type of tumors associated with the syndrome (Lotfipour et al., 2011). There is no data reporting the number of cases in Indonesia. Globally, the number of reported cases of this condition is only 37 cases worldwide between 1909 and 1989 (Singh et al., 2019).

The hallmark of FKS is the presence of an intracranial mass lesion that must be confirmed by radiological imaging. Pseudo-Foster-Kennedy Syndrome presents with optic atrophy in one eye and papilledema in the other, similar to Foster-Kennedy Syndrome, but without an intracranial mass lesion. It can result from various conditions that lead to increased intracranial pressure or optic nerve damage. Accurate diagnosis and management require a thorough clinical examination, neuroimaging to rule out mass lesions, and addressing the underlying systemic or neurological conditions (Bansal et al., 2008).

Symptoms and signs of the presence of intracranial tumor may vary including headache, nausea, vomiting, decreased vision, anosmia, and personality changes. There are 3 types of True Foster-Kennedy Syndrome. Type 1 FKS is described as ipsilateral optic atrophy and contralateral papilledema. Type 2 FKS occurs when unilateral optic atrophy and bilateral papilledema. Meanwhile, type 3 FKS is a rare type that occurs when the intracranial mass is large enough to cause bilateral optic nerve compression, leading to optic atrophy in both eyes, along with increased intracranial pressure and papilledema (Lai et al., 2014; Zehden et al., 2021).

These findings occur due to the direct compression of intracranial tumors on the ipsilateral optic nerve and nearby blood vessels, leading to ischemia and subsequent optic atrophy. The elevated intracranial pressure causes papilledema in the eye opposite to the tumor. Additionally, if the tumor compresses the olfactory nerve, it can result in anosmia (Díaz-Vintimilla et al., 2021). Relative Afferent Pupillary Defect (RAPD), also known as Marcus Gunn pupil, is frequently encountered in the context of Foster-Kennedy Syndrome (FKS), particularly in the eye displaying optic atrophy. This neurological sign manifests as an abnormal pupillary response to light, wherein the affected eye demonstrates a diminished constriction or even dilation when light is directed into it compared to the unaffected eye. In FKS, the optic nerve of the affected eye experiences degeneration or compression due to the intracranial tumor, impairing its ability to transmit visual signals to the brain. Consequently, when light is shone into this eye, the pupillary response is reduced or absent, leading to the characteristic RAPD (Yari et al., 2018).

The management of Foster-Kennedy Syndrome (FKS) involves a multifaceted approach to address the underlying cause, manage visual deficits, and control increased intracranial pressure. If FKS is caused by a tumor, such as a meningioma or craniopharyngioma, surgical removal of the tumor is often the definitive treatment except for patients who have contraindications or are considered to be at high risk for surgery (Musa et al., 2022). For pituitary adenomas, medical treatment with dopamine agonists like cabergoline can be effective in reducing tumor size and improving visual function (Kanj et al., 2022). Regular monitoring of visual function, including visual acuity, visual fields, and funduscopic examination, is crucial to assess the progression of the disease and the response to treatment. In some cases, corticosteroids may be used temporarily to reduce intracranial pressure and optic disc edema and improve visual function (Kanj et al., 2022; Musa et al., 2022). Medications like acetazolamide may be used to reduce intracranial pressure in cases where FKS is caused by conditions like idiopathic intracranial hypertension. Supportive care, such as pain management with analgesics and anticonvulsant medications for seizures, may also be required (Ayele et al., 2020; Musa et al., 2022).

In this case, the patient was administered medications including dexamethasone, citicoline, paracetamol, and oral levetiracetam as symptomatic management before performing the definitive treatment, which is tumor removal. Dexamethasone is commonly utilized to treat vasogenic edema and elevated intracranial pressure in individuals diagnosed with brain tumors (Jessurun et al., 2019). Citicoline is a promising agent for the management of brain tumors, with a potential role in improving neurological function, reducing edema, and enhancing the effectiveness of other treatments in various brain ischemia and injury (Secades, 2021). Levetiracetam is an antiepileptic drug that has been studied for its potential use in treating seizures associated with brain tumors (Nasr et al., 2016). The surgery on this patient was successful. However, the patient did not return for a follow-up consultation with the ophthalmology department.

The prognosis for patients with Foster Kennedy Syndrome depends on various factors, including the underlying cause, the effectiveness of treatment, and the patient's overall health. Early diagnosis and treatment, as well as collaboration with neurosurgeons and regular monitoring, are essential in improving the prognosis for patients with FKS (Musa et al., 2022). Early diagnosis and prompt treatment are crucial to preserve visual function and prevent further complications in patients with FKS. In this case, the patient's prognosis is considered good because there are no underlying diseases and the surgery was successful without any

complications. General practitioners play a crucial role in conducting posterior segment eye examinations using ophthalmoscope in primary care to establish an early diagnosis, refer the patient to a specialist for further evaluation, and collaborate with specialists to ensure proper diagnosis and management.

CONCLUSION

Foster-Kennedy Syndrome is a rare neuro-ophthalmological condition associated with intracranial mass. Despite its rarity, FKS remains a crucial diagnostic consideration When suspected, neuroimaging should be performed to confirm the presence of an intracranial mass. Early diagnosis and prompt treatment are vital to preserving visual function and preventing complications.

CONSENT FOR PUBLICATION

The patient has consented to the publication of this case report with full awareness of the potential inclusion of their clinical details and images. Informed consent was obtained and all personal identifiers have been removed to ensure confidentiality.

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