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Foster Kennedy Syndrome: A Case Report

Dr. Komal S. Godase
Junior Resident,
Dept. of Ophthalmology,
Vilasarao Deshmukh Government Medical College, Latur.

Dr. Uday S. Mohite
Professor and Head of Department ,
Dept. of Ophthalmology,
Vilasarao Deshmukh Government Medical College, Latur.

Dr. Vivek S. Gosavi
Assistant professor,
Department of Ophthalmology,
Vilasarao Deshmukh Government Medical College, Latur.

Dr. Mayur Kulkarni
Assistant professor,
Department of Ophthalmology,
Vilasarao Deshmukh Government Medical College, Latur.

Correspondence to:
Dr. Komal S. Godase

Abstract:-

➢ ***Purpose***

To describe a presentation of Foster Kennedy syndrome (FKS) with a distinct pattern: one eye shows optic nerve atrophy due to compression by an intracranial tumor, while the other eye exhibits optic disc edema caused by heightened intracranial pressure. This atypical presentation highlights how commonplace clinical signs can mask a rare condition or disease phenotype.

➢ ***Case Report***

A 26-year-old woman visited an ophthalmologist reporting sudden vision loss in her left eye over the past two months, accompanied by untreated headaches. She had no other health concerns. Her best corrected visual acuity was 6/36p in the right eye and PL+PR Faulty in the left eye. A relative afferent pupillary defect of grade 2 was observed in the left eye. Intraocular pressures were normal. Extra ocular movements were normal. Fundus examination revealed optic atrophy in left eye and frank optic disc edema in right eye. MRI brain scan was advised to patient to look for space occupying lesions. MRI imaging showed a 3.1x3.4x3.8cm sized predominantly cystic lesion involving sella and supra-sellar region appearing hyper intense on both T2/FLAIR and T1 with peripheral blooming and peripheral enhancement of solid component with extensions s/o adamantinomatous Craniopharyngioma with Superiorly-compression and elevation of optic chiasma, bilateral optic tracts. And hence patient is diagnosed as case of Foster Kennedy syndrome. The patient was given Nepafenac 0.1% eye drop once daily for two weeks. The patient was referred for a neurosurgical consultation and Craniotomy with aspiration was done by neurosurgeon and sample is sent for cytology.

➢ ***Conclusion***

When encountering a patient with optic atrophy in one eye along with optic disc edema in the other eye, suspicion should arise regarding the presence of a space-occupying lesion. A concise case history , extensive ocular work up and cranial magnetic resonance imaging showing sudden visual loss and raised intracranial pressure which give rise to the diagnosis of sight threatening, permanent and fatal condition such as Foster Kennedy Syndrome.

Keywords:- *Foster Kennedy Syndrome (FKS), Vision Loss, Optic Atrophy, Intracranial Hypertension.*

I. INTRODUCTION

Foster Kennedy syndrome (FKS) manifests through neurological symptoms, including loss of smell (anosmia) and vision impairment (central scotoma). The vision loss can occur in one or both eyes, depending on the stage of the syndrome. This condition is characterized by optic nerve damage (atrophy) in one eye due to compression, accompanied by swelling of the optic nerve (papilledema) in the opposite eye. These changes result from heightened pressure within the skull caused by a space-occupying lesion in the brain.[1]

FKS usually presents in three distinct types. [2] Type 1, the most common form, exhibits optic atrophy in one eye and papilledema in the opposite eye. Type 2 is marked by papilledema in both eyes and optic atrophy in one eye. [3] Type 3 is distinguished by the progression from bilateral papilledema to bilateral optic atrophy. These three types of FKS result from distinct stages of metastasis of brain tumors. Meningiomas are tumors that arise within the central nervous system and occupy space within the cranial cavity. [4] Common locations for meningiomas to develop include the sphenoid wing, olfactory groove, and frontal lobe within the cranial cavity. [5,6]

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Meningiomas represent the most prevalent form of non-malignant brain tumors. [7] Their progression is typically insidious, characterized by subtle and gradual changes that may go unnoticed as the condition advances.

There are several intracranial masses that can lead to Foster Kennedy Syndrome, including meningiomas of the olfactory groove or sphenoidal wing, craniopharyngiomas, pituitary adenomas, neuroblastomas, plasmacytomas, aneurysms, and tuberculous brain abscesses. These masses can cause compression or damage to the optic nerve or increase intracranial pressure, leading to the syndrome's characteristic features.

II. CASE REPORT

A 26-year-old woman visited an ophthalmologist, reporting a two-month history of vision loss in her left eye. She also mentioned experiencing headaches during this time, although she hadn't sought treatment for them. Aside from these symptoms, she had no other health concerns. There was no history of head trauma, and both her medical and family histories were normal.

- *On Ocular Examination:*

Lid of both eyes were normal with clear conjunctiva and cornea. Depth of anterior chamber was normal. Iris showed normal color pattern in both eyes. Pupils of the right eye were normal in size and reacting to light while there was presence of relative afferent pupillary defect (RAPD) in the left eye. Visual acuity was 6/36p for right eye and PL+, PR was faulty for left eye. Extra ocular movements were normal with normal intraocular pressures in both eyes. Fundoscopic examination demonstrated right optic disc edema and left optic nerve atrophy (Fig.1).

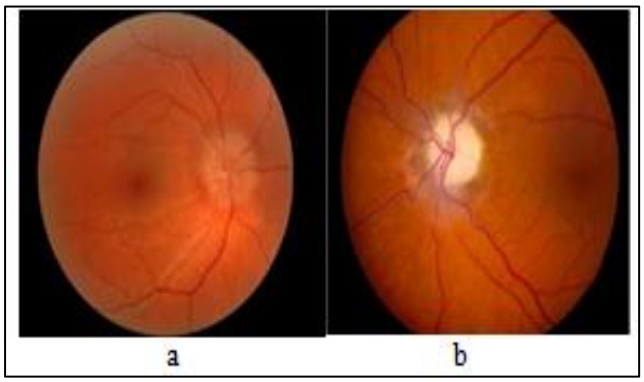

Fig 1:- a: RE Optic Disc, b: LE Optic

- *Magnetic Resonance Imaging*

Brain magnetic resonance imaging showed a meningioma located at the left sphenoid wing, which was exerting pressure on the left optic nerve. (Fig. 2).

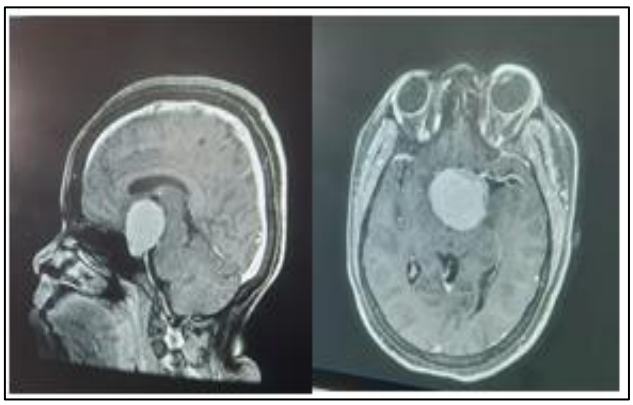

Fig 2 Magnetic Resonance Imaging of Brain

A 3.1 x 3.4 x 3.8 cm sized predominantly cystic lesion involving sella and supra-sellar region appearing hyper intense on both T2/FLAIR and T1 with peripheral blooming and peripheral enhancement of solid component with extensions s/o Adamantinomatous Craniopharyngioma. Superiorly-compression and elevation of optic chiasma, bilateral optic tracts.

The patient underwent preoperative embolization of the tumor through the left middle meningeal artery, followed by a left frontotemporal craniotomy. Histopathological analysis confirmed a transitional meningioma (WHO grade I).

Postoperatively, the patient recovered without complications. At the six-month follow-up, their visual acuity remained stable, with resolution of optic disc edema and headache on the right side.

III. DISCUSSION

Ocular conditions with presentations resembling FKS include pseudo-Foster Kennedy syndrome, non-arteritic anterior ischemic optic neuropathy (NAION), and optic nerve hypoplasia, featuring hallmark signs such as unilateral optic atrophy or unilateral papilledema. NAION ranks as the second leading cause of optic neuropathy. [8]

Optic nerve hypoplasia is a congenital condition characterized by incomplete development of the optic nerve, resulting in the typical small and pale appearance observed in affected individuals. [9,10] Optic neuritis refers to inflammation affecting the optic nerve, and when the inflammation specifically involves the optic nerve head, it is termed optic papillitis. [11]

FKS commonly presents with large tumors located in the frontal lobe and olfactory groove meningiomas. While smaller tumors can be equally serious, they may not produce the same orbital compression effects as larger ones. Meningiomas, originating from abnormal growth of the meninges in the central nervous system, are the primary culprit, with tumors in the sphenoid wing and pituitary gland also contributing to the condition. Clinicians typically identify FKS through signs such as optic atrophy in one eye and papilledema in the opposite eye, along with other symptoms like reduced visual acuity, diplopia, and visual

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field loss. Contralateral eye vision usually remains unaffected until later stages. Proptosis may occur if the tumor extends into the orbit, particularly in the anterior cranial fossa. Neuroimaging studies, such as head and orbital CT scans or MRI with or without contrast, are essential for confirming the diagnosis. [12]

IV. MANAGEMENT OF FKS IN THIS CASE

Patient was diagnosed predominantly cystic lesions with Adamantinomatous Craniopharyngioma and was referred to neurosurgery for further management. Along with routine blood investigations, patient was also advised for endocrinal deficiencies: GH, FSH, ACTH, and TSH. Surgery is the first line of treatment for most of symptomatic patients. Craniotomy with aspiration was advised to the patient and sample was sent to cytology.

V. CONCLUSION

FKS is a rapidly evolving, insidious condition. Routine examinations, such as dilated fundoscopy and ophthalmoscopy, in addition to a proper case history can make a great difference in the detection and prognosis of the condition. An MRI is the best diagnostic option for suspected cases. Ocular signs and symptoms may be accompanied by systemic presentations, such as anosmia, nausea, and emotional imbalance. This fatal condition results in ocular morbidity and can progress to total blindness in both eyes. In extreme cases, the condition is life-threatening. The management of FKS requires a coordinated multi-disciplinary approach. The prognosis of FKS depends largely on the extent of the intracranial space-occupying lesion.

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