Pseudo Foster Kennedy Syndrome - A rare case of vision loss in child

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Abstract

Pseudo Foster Kennedy Syndrome [PFKS] is a rare presentation in paediatrics age group which creates a hindrance for the daily wellbeing. Herein, we report a case of a 11-year-old child with unnoticed vision loss in both eyes preceding febrile illness. Medical and clinical history with fundus findings of disc oedema in one eye and other eye optic atrophy, with no evidence of intracranial mass in neuroimaging, leaded us to diagnosis of PFKS. Visual acuity of child improved on treatment with corticosteroids and was successfully discharged.

Introduction

Foster Kennedy Syndrome is a unilateral optic disc swelling with contralateral optic atrophy, usually due to a frontal lobe tumour compressing the optic nerve on one side and resulting in papilledema contralaterally. In the absence of an intracranial mass these findings may be labelled as Pseudo Foster Kennedy Syndrome [PFKS].

Bilateral sequential anterior ischemic optic neuropathies, either arteritic (AAION) or nonarteritic (NAION), are the usual culprits in PFK, with NAION the most common etiology[1]. Other causes of PFK that have been reported include optic neuritis, trauma, idiopathic intracranial hypertension and syphilis. Herein we present a rare case of PFK in a child.

Case Report

A 11-year-old female child 2nd in order born out of 2nd degree consanguineous marriage, presented with complaints of fever since 7 days, headache since 4 days and loss of vision since 4 days, initially in left eye followed by right eye which was sudden onset progressively associated with pain.

On presentation she was hemodynamically stable with normal blood pressures, but with complaints of fever and headache. She had vitals of Temperature 100.2 degree C, HR 116bpm, RR 24cpm, SpO2 98%, BP 98/64mmhg. Fever subsided with symptomatic treatment, but headache persisted. Ophthalmology evaluation was done which showed perception of light positive in right eye, and counting fingers close to face in left eye. Fundus examination showed right eye disc oedema and left eye optic atrophy. Baseline investigations done which showed septic screen negative. Probably the aetiology being viral pathology. Blood investigation showed normal counts [table1] with ESR of 20mm and CRP of 1.62mg/l. Neurological examination was found to be normal.

Table 1: Laboratory Investigations

Haemoglobin	12.5gm/dl
White blood cells	8800cells/cumm
Platelet count	196000/cumm
PCV	37.5%
ESR	20mm
CRP	1.62mg/l
Creatinine	0.5mg%
Urea	21.4mg/dL
Na/K/Cl	137/3.8/111meq/L

An MRI of the brain showed bilateral optic nerves vertical tortuosity with perineural prominent CSF spaces suggesting idiopathic raised ICT, with no evidence of intracranial mass, haemorrhage or infarct. [Figure 1]

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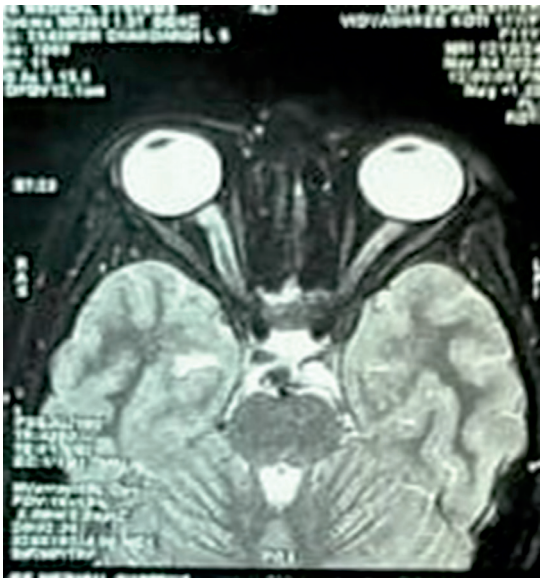


Figure 1: MRI showing bilateral vertical tortuosity of optic nerve with prominent CSF spaces

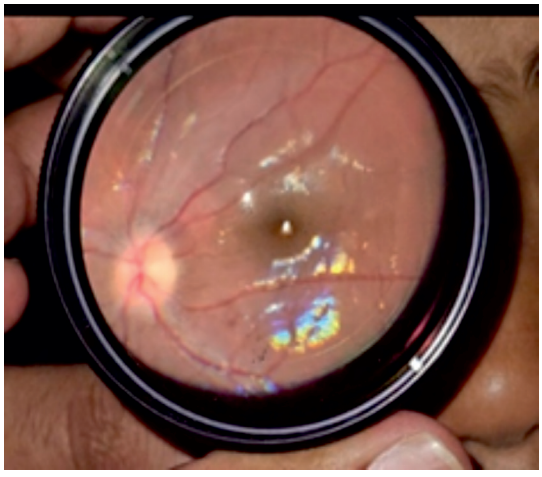


Figure 2: Disc oedema in Right eye

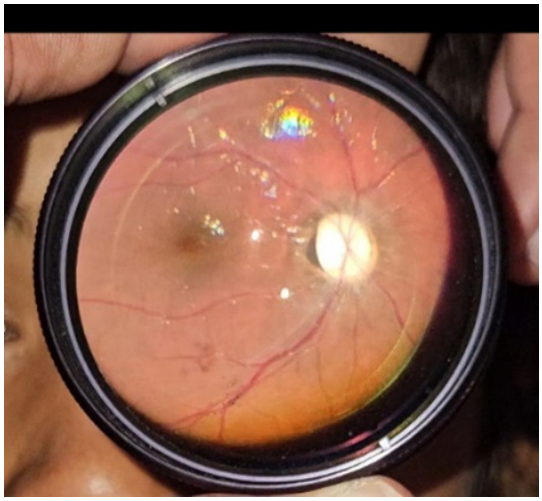
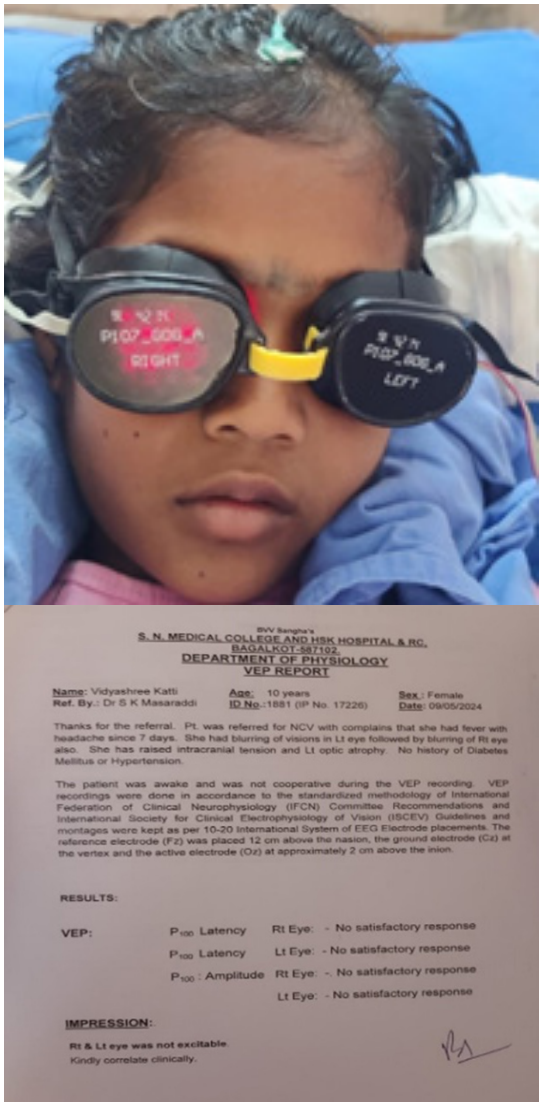


Figure 3: Optic Atrophy in Left eye

A lumbar puncture was performed in the left lateral decubitus position and CSF manometry showed normal pressures of 14 cm water with no infective aetiology from CSF analysis [total WBC 02/cumm with lymphocytes 100%, sugar 108mg% protein 46mg% ADA 3.8U/L and negative for AFB]. Visual evoke potential was not elicitable in both eyes. [Figure 4]



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VEP REPORT

Name: Vidyashree Katti Age: 10 years Sex: Female
Ref. By.: Dr S K Masaraddi ID No.:1881 (IP No. 17226) Date: 09/05/2024

Thanks for the referral. Pt. was referred for NCV with complains that she had fever with headache since 7 days. She had blurring of visions in Lt eye followed by blurring of Rt eye also. She has raised intracranial tension and Lt optic atrophy. No history of Diabetes Mellitus or Hypertension.

The patient was awake and was not cooperative during the VEP recording. VEP recordings were done in accordance to the standardized methodology of International Federation of Clinical Neurophysiology (IFCN) Committee Recommendations and International Society for Clinical Electrophysiology of Vision (ISCEV) Guidelines and montages were kept as per 10-20 International System of EEG Electrode placements. The reference electrode (Fz) was placed 12 cm above the nasion, the ground electrode (Cz) at the vertex and the active electrode (Oz) at approximately 2 cm above the inion.

RESULTS:

VEP:	P_{100} Latency	Rt Eye: - No satisfactory response
	P_{100} Latency	Lt Eye: - No satisfactory response
	P_{100} : Amplitude	Rt Eye: - No satisfactory response
		Lt Eye: - No satisfactory response

IMPRESSION:

Rt & Lt eye was not excitable.
Kindly correlate clinically.

Figure 4: Visual Evoke Potential test

Child was started on systemic corticosteroids i.e. injection methylprednisolone with dosage of 30mg/kg/dose and continued for 5 days. She was discharged successfully under the treatment coverage of oral steroids. Later she was started with oral prednisolone tablets 2mg/kg/dose for 1 week followed by tapered dose next week.

Post treatment visual acuity improved especially in the eye with the disc swelling.

Ophthalmic examination was done after completion of dosage, which showed an improved vision of 6/9 in atrophied left eye and 6/6 in right eye, which made her day-to-day activities easier with average academics in seventh standard.

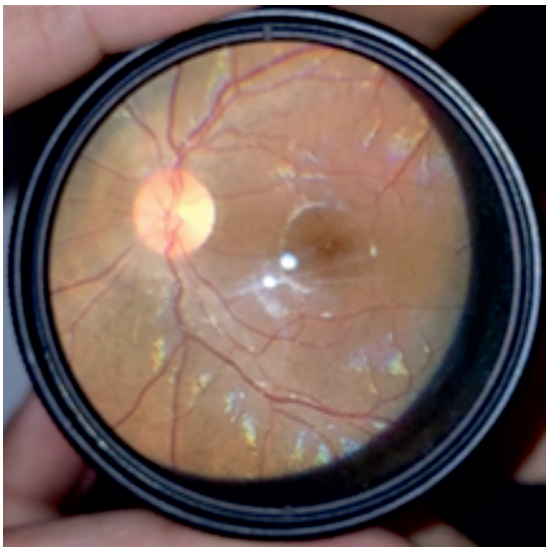

Figure 5: Right Eye Post Treatment Showing Resolution of Disc Edema

Discussion

Pseudo-Foster Kennedy Syndrome (PFKS) is characterized by unilateral optic atrophy with contralateral optic disc oedema, mimicking true Foster Kennedy Syndrome (FKS) but without an intracranial mass lesion. While FKS typically results from a frontal lobe tumour compressing the optic nerve, PFKS occurs due to sequential optic neuropathies, commonly non-arteritic anterior ischemic optic neuropathy (NAION[1].

In paediatric cases, PFKS is extremely rare, with most occurrences reported in middle-aged individuals. NAION is often linked to systemic conditions like hypertension, hypercholesterolemia, and sleep apnoea, which are less common in children[4]. Normal levels of ESR, C-reactive protein (CRP) and platelet counts are often noticed.

The nerve affected recently often displays pallid oedema in cases of arteritic anterior ischemic optic neuropathy [AAION]. In comparison patients with AAION are older and more commonly female. AAION have worse visual outcomes than NAAION patients with rare late improvement. An elevated ESR, CRP, and platelet levels are seen. Pathologic examination of consecutive slices spaced at 0.25 cm and a 2- to 3-cm temporal artery biopsy is the gold standard for diagnosis However, as in this case, viral pathology may contribute to transient optic nerve ischemia, leading to PFKS.

Approximately one fourth of patients, such as the one described in this case, are asymptomatic and diagnosed on routine eye examination[5].

The differential diagnosis of PFKS includes optic neuritis, idiopathic intracranial hypertension, and compressive optic neuropathy. Optic neuritis, commonly associated with demyelinating diseases, presents with pain, colour vision changes, and an afferent pupillary defect features absent in this patient[6]. Idiopathic intracranial hypertension may cause papilledema but lacks sequential optic nerve damage seen in PFKS. Autoimmune optic neuropathy is one of the other differential diagnoses of PFKS, which is characterised by acute or subacute bilateral vision and colour vision loss, retinal degeneration and seropositivity for one or more retinal autoantibodies which are against our case findings. Vitamin B12 deficiency has been implicated in optic neuropathy, yet normal levels in this case ruled out nutritional etiology[7].

Treatment approaches remain controversial, as no definitive therapy exists for NAION. Systemic corticosteroids, as administered in this case, have shown efficacy in reducing optic disc oedema and improving visual outcomes. The patient's successful recovery with improved visual acuity underscores the potential benefits of early intervention. Ophthalmological evaluation plays a crucial role in distinguishing PFKS from other optic neuropathies and guiding appropriate management.

According to Bansal et al,[2] a two-year boy with history of developmental delay, panhypopituitarism, hydrocephalus and obesity presented with vision deterioration. MRI showed Chiari malformation with small pituitary gland with no other symptoms of raised intracranial pressure. Ophthalmic examination revealed right severe papilledema and left hypoplastic optic disc.

Thus, in the absence of an intracranial mass, unilateral disc swelling with contralateral papilledema is termed as Pseudo Foster Kennedy Syndrome.

As mentioned in Laura et al,[1] a 67-year-old woman who initially presented with 30day history of headache and decreased vision with ESR of 9 was treated with one week of oral steroids. There after 1 year later she had come with a healing arterial wall inflammation in temporal artery biopsy, with ESR 20, left optic nerve pale and right swollen which led to the diagnosis of GCA presenting as PFK.

Here in our case, the child had symptoms of headache and fever with sequential loss of vision with MRI brain showing no evidence of intracranial mass. Along with her fundus examination of right eye disc oedema and left eye optic atrophy clearly formulate the diagnosis of PFK Syndrome. No evidence of demyelinating lesions was identified which might have supported to optic neuritis.

It may be that the atrophic optic nerve had the most severe optic disc oedema initially, leading to optic

atrophy, with relative preservation of less affected optic nerve, which continues to manifest optic disc swelling. Normal levels of vitamin B12 [400pg/mL] excludes the possibility of optic neuropathy caused by the same. An elevated ESR and variable loss of vision in both eyes adds clue to the diagnosis.

Pretreatment visual acuity was just counting fingers in front of face. Later following initiation of treatment vision in both eyes had improved. Relative afferent pupillary defect was not identified in fundus examination, which will be specific in optic neuritis patients. There were no signs of increased intraocular pressure and no colour vision changes and pain as seen in optic neuritis.

The medical history of vision loss preceding fever and headache with the above-mentioned fundus findings with normal CSF pressures point towards the presumptive diagnosis of anterior ischemic optic neuropathy (AION). Furthermore, a temporal artery biopsy was recommended.

Conclusion

There is no accepted treatment for AION. With successful treatment, the disc swelling in one eye may resolve, but optic atrophy in the other eye may be permanent. Systemic corticosteroid therapy and prednisone therapy on higher doses have shown improvement in visual acuity. Pseudo Foster Kennedy Syndrome is rare in paediatric age group and usually asymptomatic and ophthalmology evaluation contribute much more in the diagnosis.

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