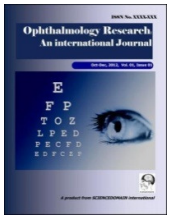

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Third Ventricular Ependymoma Mimicking Foster Kennedy Syndrome- A Case Report

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Authors' contributions

This work is carried out in collaboration of all authors. Authors ELT and RK designed the study, wrote the first draft of the manuscript and managed the literature search. Authors RK, WHWH and LSAT revised it critically for intellectual contents. Authors ELT, RK, WHWH and TLK were involved in the assessment and treatment of the patient. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Aims: To report a case of a third ventricular tumour mimicking Foster Kennedy Syndrome in a young adult.
Presentation of Case: A 21-year-old female presented with bilateral blurring of vision with preceded by generalized headache, nausea and vomiting. Fundoscopy revealed optic nerve atrophy of the right eye and a swollen optic disc on the left in keeping with features of Foster Kennedy Syndrome. MRI of the brain revealed a third ventricular tumor extending into the suprasellar region with hydrocephalus. Surgical excision of the tumour was done and the subsequent histopathological report confirmed it to be a clear cell ependymoma.
Discussion: Foster Kennedy syndrome is a rare clinical constellation describing a pattern of ocular findings typically related to extraaxial tumours involving the anterior skull base. It is characterized

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by the triad of unilateral optic disc swelling, contralateral optic atrophy and ipsilateral anosmia. The clinical signs of Foster Kennedy syndrome are a result of direct compression of the mass on the optic nerve and an indirect effect from raised intracranial pressure.
Conclusions: We conclude from this report that intraventricular or intraaxial lesions in the vicinity of the optic apparatus may also produce features mimicking Foster Kennedy syndrome in clinical practice.

Keywords: Foster Kennedy syndrome; third ventricle; ependymoma; optic atrophy; optic disc swelling.

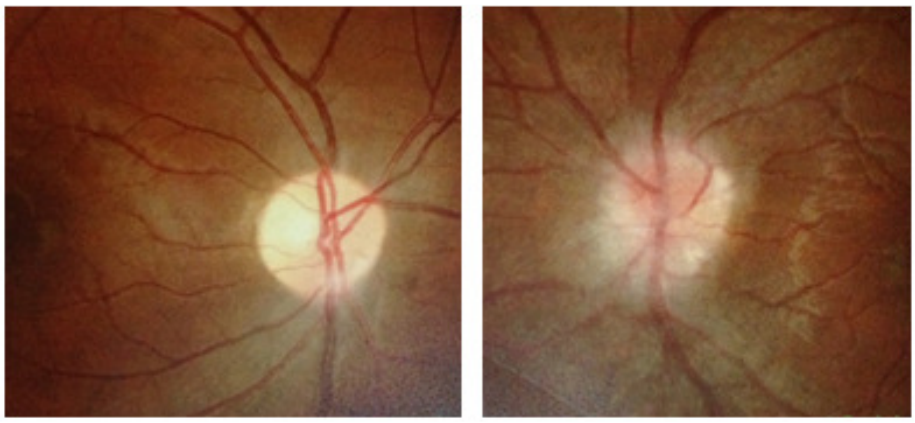
1. INTRODUCTION

Foster Kennedy syndrome is a rare clinical entity which was first described in 1911. It is characterized by a triad of unilateral optic atrophy, contralateral optic disc swelling and ipsilateral anosmia [1]. This is a rare clinical syndrome and manifests in only in 1-2% of all intracranial mass lesions [1]. It is believed to have a localizing value which indicates that it arises from the floor of the anterior cranial fossa. The clinical signs are usually caused by tumors related in the frontal lobe, olfactory groove or optic chiasm and are usually meningiomas or gliomas [2]. We therefore report a rare case of third ventricular tumour that resulted in a clinical picture mimicking this syndrome.

2. CASE REPORT

A 21-year-old lady with no prior medical illness, presented with progressive blurring of vision in both eyes for a period of 6 months. The blurring of vision was more profound in right eye and it had worsened over the last one month. The patient's husband noted that she was less aware of objects coming from right side unless she turned her head in that direction. Her visual symptoms were also preceded by progressively worsening intermittent generalized headaches. The headache was throbbing in nature and more severe in the morning upon awakening and was associated with nausea and vomiting. In the days prior to admission the patient was also noted to be lethargic and withdrawn. She denied any anosmia.

On examination, the patient was alert and conscious and orientated to her surroundings. Her vital signs were normal. Assessment of higher mental function revealed impairment in her short term memory. Ocular examination noted that the right eye was only able to perceive light while the left eye was 6/12. Relative afferent pupillary defect was profound on the right. The anterior segments of both eyes examination were unremarkable and the intraocular pressure was normal bilaterally. There was bilateral sixth nerve palsy. The visual field testing noted temporal field defect in the left eye. Funduscopic examination of the left eye revealed a hyperaemic swollen optic disc with blurred margins, and right eye showed optic atrophy with temporal pallor (Figs. 1A and B). There was no spontaneous venous pulsation noted on both optic discs. Neurological examination was otherwise unremarkable.



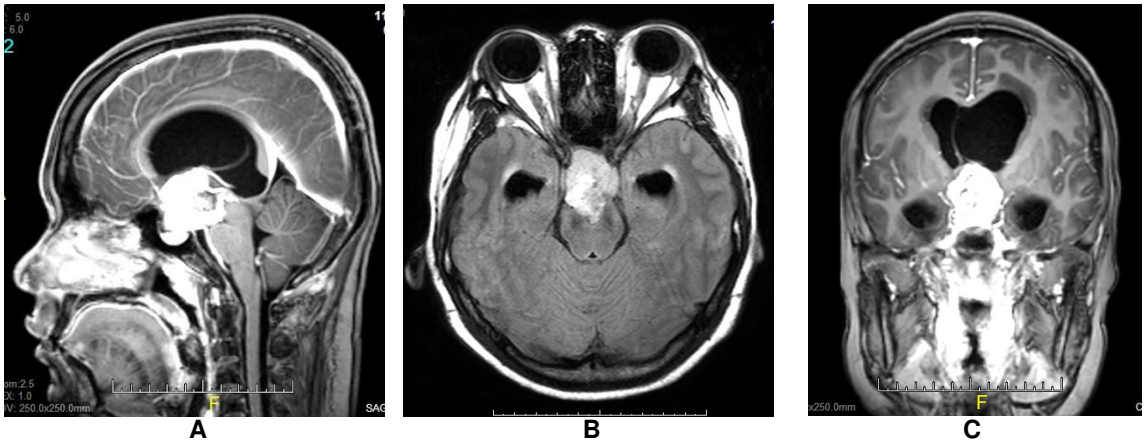
A **B**

Fig. 1A and B. Right eye optic atrophy and Left optic disc swelling

MRI was done which revealed a heterogeneous enhancing lesion arising within the third ventricle and extending into the suprasellar region. The mass was ill defined, lobulated and measured 3.1 (W) x 3.7 (AP) x 3.7(CC) cm. It appeared hypointense on T1, hyperintense on T2 and was not suppressed by FLAIR. The mass was also noted to be associated with obstructive hydrocephalus Figs. 2A-C).

The patient underwent a frontal craniotomy and excision of her tumour via a subfrontal translamina terminalis approach. Histopathology examination revealed features of clear cell ependymoma (WHO Grade II) (Fig. 3).

One month after surgery, the vision in her eyes remained unchanged but fundoscopic examination noted resolution of the left optic disc swelling. Right eye optic disc remained same. The patient was planned for further MRI imaging, however she defaulted follow up care. The patient presented again after 1 year at which time she suffered from recurrent headaches. An urgent MRI of her brain revealed evidence of tumour recurrence Figs. 4A-B). She was advised for further surgery but refused. Subsequently she was referred to the oncologist who advised a course of Radiation therapy totaling 50Gy in 30 fractions.



A B C

Fig. 2A-C. MRI showing heterogeneous enhanced lesion at the third ventricle extending intosuprasellar region and impinging the right optic nerve with accompanied hydrocephalus

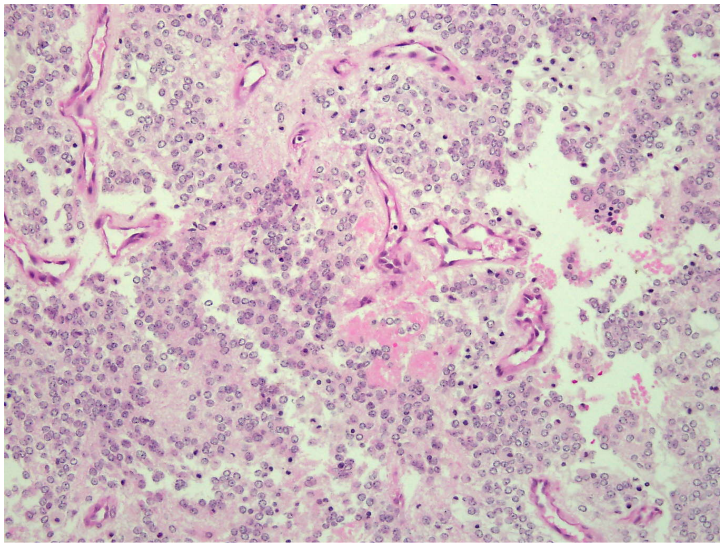
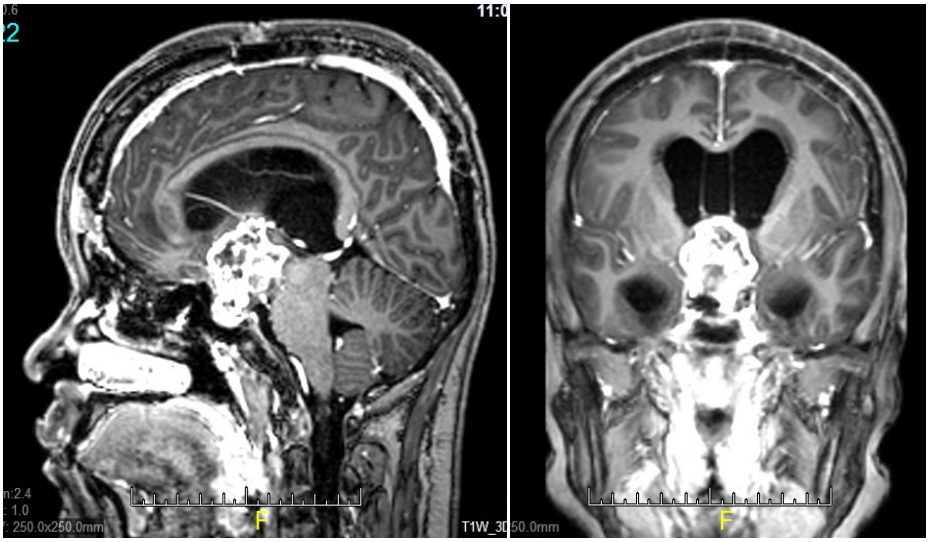


Fig. 3. Histopathological examination revealed clear cell ependymoma (WHO Grade II)



A B

Fig. 4A-B. MRI of the brain showing recurrence of her tumour one year after surgery

3. DISCUSSION

Foster Kennedy syndrome is an uncommon clinical constellation which was first described in 1911 by the British neurologist Robert Foster Kennedy [1-2]. He described the triad of unilateral optic disc swelling with contralateral optic atrophy and ipsilateral anosmia. A majority of patients with features of Foster Kennedy Syndrome have intracranial masses originating from the olfactory groove, frontal lobe or sphenoidal region [1]. There exist some isolated case reports of this syndrome being seen in tumours originating in the sellar or parasellar region, optic nerve or even the occipital lobe. Pseudo Foster Kennedy Syndrome is an alternate term used to describe patients with unilateral optic disc swelling and contralateral optic atrophy in the absence of an intracranial mass [2-3]. It has been reported in patients with bilateral sequential consecutive ischemic optic neuropathy or optic neuritis as well as in conditions such as meningitis, carotid aneurysms, optic nerve hypoplasia, intracranial inflammation and retinitis pigmentosa [3-6]. One case report has even noted optic disc swelling secondary to uncontrolled hypertension with contralateral ischemic optic neuropathy [3].

The exact pathogenesis of Foster Kennedy syndrome has not been well elucidated to date [3,7]. Foster Kennedy himself postulated that the causes of such clinical signs were due to the direct compression of the lesion on the optic nerve and an indirect cause secondary to raised intracranial pressure [8,9]. However, only 22% of cases of Foster Kennedy Syndrome were supported by this mechanism. Other postulated mechanisms include bilateral optic nerve compression without raised intracranial pressure in 33% of cases, and chronic elevation of intracranial pressure without compression in 5% of cases. In about 40% of cases, the mechanisms of Foster Kennedy Syndrome were unclear [7-10].

In this patient, we are of the opinion that her clinical features mimicking Foster Kennedy syndrome are most explainable by the original hypothesis of direct compression as well as secondarily raised intracranial pressure. From the imaging, the mass at the third ventricle has extended beyond the third ventricle into the suprasellar and sellar region. There was direct compression of the right optic nerve by the mass, leading to right optic nerve atrophy. The contralateral optic disc swelling resulted from the raised intracranial pressure as evidenced by the presence of obstructive hydrocephalus and dilatation of third ventricle. The preceding headache prior to the visual symptoms suggested the chronic elevation of intracranial pressure prior to the direct compression of the optic nerve from the mass. Clinically, there were also bilateral sixth nerve palsies which are likely a false localizing sign from raised intracranial pressure. The presence of memory loss suggested the involvement of the hippocampus.

Anosmia, one of the triad of Foster Kennedy syndrome, which is due to direct compression of olfactory tract, was not elicited in this patient.

We reported this rare case of Foster Kennedy syndrome secondary to ependymoma arising from third ventricle. To our best knowledge, this is the first case of third ventricular ependymomas in young adult leading to clinical features of Foster Kennedy syndrome.

Ependymomas are very rare; they arise from ependymal cells lining the ventricular system, including the spinal cord. They constitute 7% of all intracranial tumors in childrenaged 15 years old or younger [10,11]. It is rarer in adults, accounting for 2-5% of all intracranial tumors [12]. Most of the intracranial ependymoma arise in the posterior fossa, where fourth ventricle being the most common site of occurrence [11]. It can also occur within the brain parenychma. In adults, most of the ependymoma arise in spinal cord [12]. Supratentorial ependymomas are rare; especially those arise from third ventricle [13,14]. The clinical manifestation depends on the structure affected. Surgery remains the mainstay of therapy of ependymomas. However, complete resection is not possible as most of the time it has involved surrounding critical structures.

4. CONCLUSION

Foster Kennedy syndrome may not be necessary due to lesion arising from anterior cranial fossa. Any intraventricular or intraaxial lesions in the vicinity of the optic apparatus may also produce features mimicking Foster Kennedy syndrome in clinical practice.

CONSENT

All authors declare that 'written informed consent was obtained from the patient for publication of this case report and accompanying images.

ETHICAL APPROVAL

Not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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