

Case Study

Third Ventricular Ependymoma mimicking Foster Kennedy Syndrome- A Case Report

ABSTRACT

Aims: To report a case of Foster Kennedy Syndrome arises from third ventricular ependymoma in a young adult.

Presentation of Case: 21-year-old female presented with bilateral blurring of vision with preceding generalized headache, nausea and vomiting. Fundoscopy revealed optic atrophy of the right eye with a swollen optic disc on the left in keeping with features of Foster Kennedy Syndrome. MRI of the brain revealed a third ventricular tumour extending into the suprasellar region with associated hydrocephalus. Following craniotomy and excision of her tumour, the histopathological findings were consistent with 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 described the triad of unilateral optic disc swelling with contralateral optic atrophy and ipsilateral anosmia. The clinical signs of Foster Kennedy syndrome were postulated to direct compression from the mass on the optic nerve with indirect effect from the 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 comprises a triad of unilateral optic atrophy with contralateral optic disc swelling and ipsilateral anosmia [1]. Despite being well documented, this syndrome is rare and manifests

19 in only in 1-2% of all intracranial mass lesions [1]. It is believed to have a localization value
20 that indicates a lesion arising from the floor of the anterior cranial fossa. The pathological
21 entities responsible are usually tumours related to the frontal lobe, olfactory groove or optic
22 chiasm and are usually meningiomas or gliomas [2]. We report a rare case of Foster
23 Kennedy syndrome secondary to a tumour originating within the third ventricle that resulted
24 in a clinical picture mimicking this syndrome.

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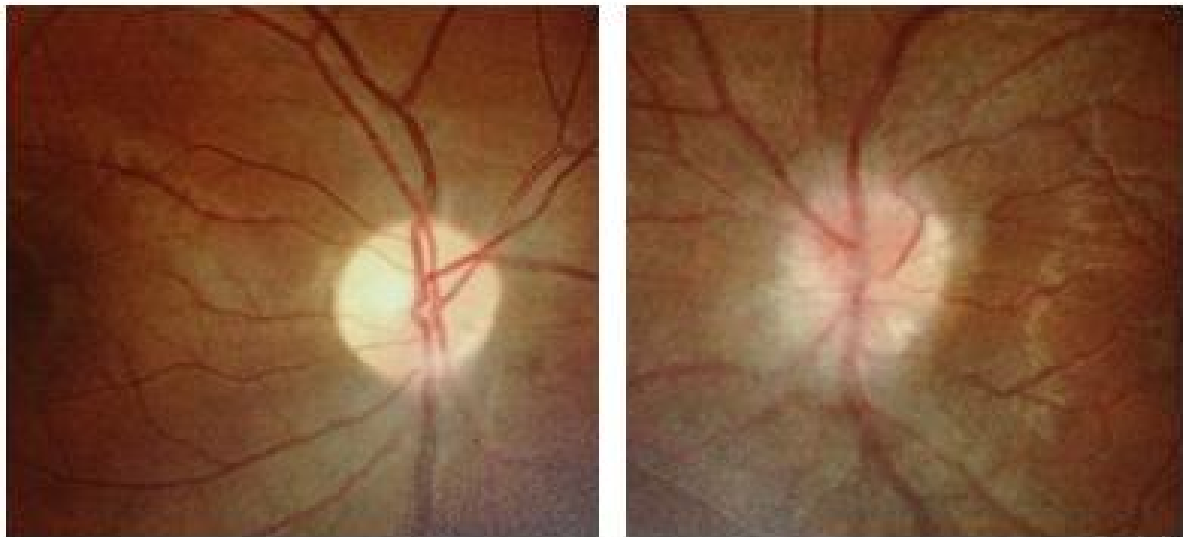
26 **2. CASE REPORT**

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

36

37 On examination, the patient was alert and conscious and orientated to time place and
38 person. Her vital signs were stable. Assessment of higher mental function revealed
39 impairment in her short term memory. Ocular examination noted that the right eye was only
40 able to perceive light while the left eye was 6/12. Relative afferent pupillary defect was
41 profound on the right. The anterior segments of both eyes examination were unremarkable
42 and the intraocular pressure was normal bilaterally. There was bilateral sixth nerve palsy.
43 Fundusoscopic examination of the left eye revealed a hyperaemic swollen optic disc with
44 blurred margins, and right eye showed optic atrophy with temporal pallor (Fig 1).
45 Neurological examination was otherwise unremarkable.

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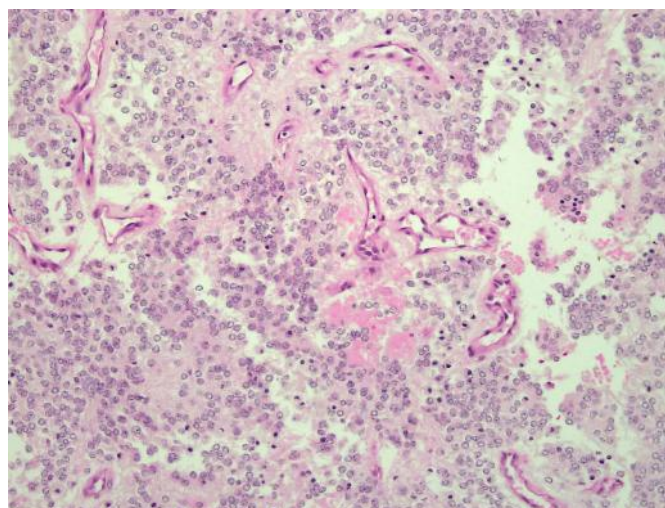
47
48 Figure 1: Right eye optic atrophy and Left optic disc swelling

49
50 Patient underwent a Magnetic Resonance Imaging (MRI) of her brain which revealed a
51 heterogeneous enhancing lesion arising within the third ventricle and extending into the
52 suprasellar region. The mass was ill defined, lobulated and measured 3.1 (W) x 3.7 (AP) x
53 3.7(CC) cm. It appeared hypointense on T1, hyperintense on T2 and was not suppressed by
54 FLAIR. The mass was also noted to be associated with obstructive hydrocephalus (Fig 2)
55



56
57 Figure 2: MRI showing heterogeneous enhanced lesion at the third ventricle extending into
58 suprasellar region with accompanied hydrocephalus

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



62
63 Figure 3: Histopathological examination revealed clear cell ependymoma (WHO Grade II)
64

65 One month after surgery, the vision in her eyes remained unchanged but fundoscopic
66 examination noted resolution of the left optic disc swelling. She is currently on follow up and
67 is undergoing adjuvant therapy.

68

69 3. DISCUSSION

70 Foster Kennedy syndrome was an uncommon clinical sign which was first described in 1911
71 by the British neurologist Robert Foster Kennedy [1-2]. It described the triad of unilateral
72 optic disc swelling with contralateral optic atrophy and ipsilateral anosmia. Majority of Foster
73 Kennedy Syndrome result from intracranial mass. Mass lesion leading to Foster Kennedy
74 Syndrome often located in olfactory groove, frontal lobe and sphenoidal region [1]. However,
75 cases have been reported in parasellar or sellar tumours, optic nerve tumours and occipital
76 tumours. Pseudo Foster Kennedy Syndrome is term used to defined patients with unilateral
77 optic disc swelling and contralateral optic atrophy in the absence of intracranial mass [2-3]. It
78 has been reported in patients with bilateral sequential consecutive ischemic optic neuropathy
79 or optic neuritis. It is also reported in patients with meningitis, carotid aneurysms, optic nerve
80 hypoplasia and intracranial inflammation and retinitis pigmentosa [3-6]. There was also a
81 case reported on optic disc swelling secondary to uncontrolled hypertension with
82 contralateral ischemic optic neuropathy [3].

83

84 The pathogenesis of Foster Kennedy syndrome was not well elucidated [3, 7]. Foster
85 Kennedy postulated the causes of such clinical signs were due to the direct compression of
86 the lesion on the optic nerve and an indirect cause secondary to raised intracranial pressure
87 [8,9]. However, only 22% of cases of Foster Kennedy Syndrome were supported by this
88 mechanism. Other postulated mechanisms include bilateral optic nerve compression without
89 raised intracranial pressure in 33% of cases, and chronic elevation of intracranial pressure
90 without compression in 5% of cases. About 40% of cases, the mechanism of Foster
91 Kennedy Syndrome were unclear [7-9].

92

93 In this patient, the cause of Foster Kennedy syndrome is most likely due to the hypothesis as
94 suggested by Foster Kennedy. From the imaging, the mass at the third ventricle had
95 involved the visual pathway and causing obstructive hydrocephalus. There was direct
96 compression of the right optic nerve by the mass, leading to right optic nerve atrophy. The
97 contralateral optic disc swelling resulted from the raised intracranial pressure as evidenced
98 by the presence of obstructive hydrocephalus and dilatation of third ventricle. The preceding
99 headache prior to the visual symptoms suggested the chronic elevation of intracranial
100 pressure prior to the direct compression of the optic nerve from the mass. Clinically, there
101 was bilateral sixth nerve palsies suggesting focal localizing signs results from raised
102 intracranial pressure. The presence of memory loss suggested the involvement of the
103 hippocampus. Anosmia, one of the triad of Foster Kennedy syndrome, which is due to direct
104 compression of olfactory tract, was not elicited in this patient.

105

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

109

110 Ependymomas are very rare; they arise from ependymal cells lining the ventricular system,
111 including the spinal cord. They constitute 7% of all intracranial tumors in children aged 15
112 years old or younger [10,11]. It is relatively rarer in adult, accounting for 2-5% of all
113 intracranial tumors [12]. Most of the intracranial ependymoma arise in the posterior fossa,
114 where fourth ventricle being the most common site of occurrence[11]. It can also occur within
115 the brain parenchyma. In adult, most of the ependymoma arise in spinal cord [12].
116 Supratentorial ependymomas are rare; especially those arise from third ventricle [13, 14].
117 The clinical manifestation depends on the structure affected.

118

119 Surgery remains the mainstay of therapy of ependymomas. However, complete resection is
120 not possible as most of the time it has involved surrounding critical structures. The other
121 options of treatment for residual or recurrent tumor include radiotherapy and radiosurgery
122 [15,16]. The role of chemotherapy is limited however, it is reported children with near total
123 resection may benefit from chemotherapy and to avoid the deleterious effect of irradiation
124 [17].

125

126 **4. CONCLUSION**

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

130

131 **COMPETING INTERESTS**

132 Authors have declared that no competing interests exist.

133

134 **CONSENT**

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

137

138 **ETHICAL APPROVAL**

139 Non applicable

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