Pseudo-Foster Kennedy Syndrome due to Idiopathic Intracranial Hypertension Associated with Empty Sella Syndrome and Hyperprolactinaemia: A Rare Case Report

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Abstract

Pseudo-Foster Kennedy syndrome is characterized by unilateral papilledema with contralateral optic atrophy in the absence of intracranial space occupying lesions. A 40-year old obese Asian-Indian female presented with painless progressive diminution of vision in her right eye since one month and constant headache for two months. Fundoscopy showed optic disc pallor in the right eye and papilledema in the left eye. Serum prolactin level was raised. Cerebrospinal fluid manometry revealed high opening pressure value of 38 cm of water with normal composition. MRI scan of the brain detected empty sella syndrome. Pseudo-Foster Kennedy syndrome due to IIH was diagnosed and treated with oral acetazolamide 250 mg, four times a day which resulted in complete resolution of papilledema and headache over a period of two months. Here we report a case of pseudo-Foster Kennedy syndrome which is directly attributed to idiopathic intracranial hypertension and associated with empty sella syndrome and hyperprolactinaemia.

Keywords: benign intracranial hypertension, idiopathic intracranial hypertension, empty sella syndrome, hyperprolactinaemia, Pseudo-Foster Kennedy syndrome, Foster Kennedy syndrome, pseudotumour cerebri

Introduction

Foster Kennedy syndrome was first described in 1911 by a British neurologist Robert Foster Kennedy.1 It is a rare neurological syndrome and characterized by same sided optic atrophy, produced by direct pressure on the optic nerve by an intracranial mass and opposite sided papilledema secondary to raised intracranial pressure (ICP) due to the same intracranial mass.1 The presence of optic atrophy in one eye and papilledema in the contralateral eye in the absence of an intracranial mass is called pseudo-Foster Kennedy syndrome.2 Different reported causes of pseudo-Foster Kennedy syndrome are idiopathic intracranial hypertension (IIH), pachymeningitis, optic nerve hypoplasia, meningioma infiltrating superior sagittal sinus, non-basal glioma, malignant hypertenion, simultaneous or non-simultaneous ischemic or non-ischemic optic neuropathy, traumatic optic neuropathy, congenital syphilis, and optic neuritis.2-10 Up to date, only two cases of pseudo-Foster Kennedy syndrome secondary to IIH have been reported.2,3 Hereby we describe a rare case of pseudo-Foster Kennedy syndrome due to IIH and associated with empty sella syndrome and hyperprolactinaemia.

Case Report

A 40-year old woman with a body mass index of 31 kg/m² presented to us complaining of a gradual painless progressive diminution of vision in her right eye, and throbbing headache for two months. She also had galactorrhea. There was no history of trauma, fever, weakness, seizure and any drug intake. She underwent a hysterectomy three years ago for dysfunctional uterine bleeding. Visual acuity in the right eye was counting fingers at two feet with accurate projection of rays in all four quadrants and 20/40 in the left eye. The swinging light test showed a relative afferent pupil defect in the right eye. Extraocular movements were normal in both the eyes. Intraocular pressure by applanation tonometry was 12 mm of Hg in both the eyes. Dilated fundus examination showed optic disc pallor in the right eye and papilledema in the left eye (Figure 1). The physician performed a lumbar puncture which revealed high opening pressure of 36 cm of water. A Humphrey visual field (30-2) revealed visual field loss in the right eye and peripheral constriction and enlargement of blind spot in the left eye (Figure 2).

Figure 1: (a) Fundus photograph of the right eye shows secondary optic atrophy; (b) well established papilledema in the left eye; (c) partially resolved papilledema with oral acetazolamide at one month; (d) and complete resolution of papilledema in the left eye at two month follow up.
Lab testing revealed normal rheumatoid factor, C-reactive protein, antinuclear antibody, thyroid profile, serum cortisol, and cerebrospinal fluid analysis. Serum prolactin level was 31.18 ng/ml (normal value 3.34-26.72ng/ml). ELISA for HIV was negative. Magnetic Resonance Imaging of brain showed dilated infundibular cistern compressing the pituitary suggestive of incidental finding of empty sella syndrome (Figure 3). The rest of ophthalmic and neurological examinations were normal. The patient was diagnosed with pseudo-Foster Kennedy syndrome due to IIH associated with empty sella syndrome and hyperprolactinaemia. She was started on oral acetazolamide 250 mg QID, bromocriptine 2.5 mg, OD and was advised to lose weight. She experienced relief from headache and improvement of vision in her left eye with this treatment at the end of four weeks. At the end of two months, she completely recovered from headache, papilledema and vision in her left eye improved to 20/20. However, vision in the right eye remained counting fingers at two feet. Her serum prolactin level was returned to normal and she had no galactorrhea. She was advised to have oral acetazolamide 250 mg twice a day and review after one month but she did not return back to us.

**Discussion**

Idiopathic intracranial hypertension, also known as pseudotumor cerebri, is a neurological disease of unknown etiology that primarily affects obese females of 20-44 years age. The classical presentation of IIH is headache and diminution of vision. The reported incidence of pseudotumor cerebri is about 0.9 cases per 100000 populations, with male female ratio of 8:1. Modified Dandy criteria for IIH includes:

1. Signs and symptoms of raised ICP;
2. absence of neurological abnormalities or impaired level of consciousness;
3. High cerebrospinal fluid opening pressure with normal composition;
4. No etiology for increased ICP on neuroimaging;
5. No other cause for intracranial hypertension.

Our patient had all the features essential for diagnosing her as a definitive case of IIH.

Ophthalmic manifestation of IIH includes papilledema, decreased visual acuity and occasionally abducens and facial nerve palsy. Pseudo-Foster Kennedy syndrome may occur secondary to IIH. Persistently high ICP in pseudotumor cerebri may lead to optic atrophy in an eye having mild papilledema within days or weeks. Once optic atrophy has set in, chances of papilledema are almost nil because of non-viable neurons. Association of IIH and empty sella is well known and is thought to be due to long standing raised ICP which leads to herniation of the subarachnoid cistern into the sella turcica when the diaphragma sellae is absent as seen in our case. Our patient also had hyperprolactinaemia which is a rare complication of IIH and is thought to be developed due to compression of the pituitary stalk. Defective CSF absorption, abnormal vitamin A metabolism, increased CSF production, increased estrogenicity, cerebral edema and increased intracranial venous pressure are amongst the widely accepted mechanisms for development of IIH.

MRI brain and orbit in IIH patient may show empty sella or partially empty sella, flattened posterior globe, enlarged perioptic subarachnoid space, enlargement or increased tortuosity of optic nerve, intraocular protrusion of optic nerve head and slittlike ventricles. Restoration of vision and resolution of papilledema are primary goals in management of IIH. Treatment options for IIH are conservative approach which includes acetazolamide, a carbonic anhydrase inhibitor and topiramate. Repeated therapeutic lumbar puncture, gastric bypass surgery and lumboperitoneal shunt are invasive modes of treatment. Gastric bypass surgery acts by reducing weight which is the main risk factor for IIH. Optic nerve sheath fenestration is also a well established mode of treatment of papilledema in IIH.

This is the 3rd case of pseudo-Foster Kennedy syndrome due to IIH reported worldwide and the first documented case in

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**Figure 1:** Magnetic Resonance Imaging of brain showing empty sella.

**Figure 2:** (a) A Humphrey visual field (30-2) visual field showed visual field loss in the right eye; (b) and peripheral constriction and enlargement of the blind spot in the left eye.
India. This case is different from previously reported two cases of pseudo-Foster Kennedy syndrome due to IIH due to association of empty sella and hyperprolactinaemia. This case is reported here for its rarity. The neurologist should keep in mind possibility of hyperprolactinaemia in pseudo-Foster Kennedy syndrome due to IIH.

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