Case Report

Primary Empty Sella Syndrome Presenting with Profound Bilateral Visual Loss

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Abstract

Primary empty sella syndrome (PESS) on background idiopathic intracranial hypertension (IIH) presenting with profound bilateral visual loss is a rare clinical occurrence. We report the case of PESS on background IIH in a 37-year-old obese Nigerian female. At presentation, she had a history of an insidious onset of loss of vision in both eyes with an associated history of headaches and tinnitus. Clinical examination and imaging studies revealed papilloedema and empty sella with a completely flattened pituitary gland. This report highlights that PESS could initially present to an ophthalmologist as a case of profound bilateral visual loss.

Keywords: Bilateral visual loss, diaphragma sellae, empty sella, idiopathic intracranial hypertension, primary empty sella syndrome

Introduction

Empty sella (ES) typifies an enlarged and/or deformed sella turcica filled with cerebrospinal fluid from the herniation of the subarachnoid space into the sella.[1-3] An ES is usually an incidental anatomic finding but can occasionally be symptomatic.[1,2] When symptomatic, reported distribution of clinical manifestations includes headaches 80%–88%, visual disturbances 20%, endocrine disturbances 19%–52%, and neurologic and psychiatric disturbances.[1]

ES could be primary or secondary.[1,2] Primary ES (PES) is characterized by one or more of the following: unknown cause[1] or no previous report of pituitary radiation/surgery/pathologies[1] or congenital absent (complete or partial) diaphragmatic sella[1,2] or lax/patulous diaphragmatic sella (as seen in intracranial hypertension [IIH])[3,4] or lax/patulous diaphragmatic sella following physiologic pituitary atrophy (such as after pregnancy, lactation, and menopause).[1] Secondary ES (SES) is defined in cases with iatrogenic pituitary atrophy (such as atrophic pituitary tumor occurring after medical treatment, surgery, radiotherapy, and apoplexy) or pathological pituitary atrophy (such as following postpartum pituitary necrosis, hypophysitis, and brain trauma).[1,2] Similarly, symptomatic ES is classified into PES syndrome (PESS) and SES syndrome.

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We present a rare case of PESS on a background of IIH in a 37-year-old Nigerian female with profound bilateral visual loss.

Case Report

A 37-year-old obese Nigerian female presented with 6-month history of gradual onset profound bilateral visual loss, which has progressively worsened since onset. There was a preceding 2-year history of intermittent, uncharacteristic headache, of mild-to-moderate severity, usually at the frontal part of her head. Seven months before presentation, the headache became of moderate severity, "pressure-like" with increased frequency, and associated new onset of tinnitus in her right ear. There is no history of head trauma, any type of surgery, or radiation therapy. Her menstrual flow and cycle were not altered. She was unmarried but a single mother of a male child. Other systemic history was not significant. On examination, visual acuity was nil perception of light on the right eye and hand

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movement on the left eye. The eyeballs were orthophoric, and ocular motility was full in all directions of gaze. Pupils were centrally located, clear, round, and 5 mm in size; direct and consensual pupillary reactions were sluggish. Fundoscopy and fundus photograph revealed on the right eye – edematous pale disc, indistinct disc margin, with scanty peripapillary hard exudates while on the left eye – edematous erythematous disc, indistinct disc margin, with marked peripapillary hard exudates extending on to the fovea with star configuration and macular edema [Figure 1]. Other ocular findings were essentially normal. Her weight was 74 kg, height was 1.54 m, a BMI of 31.2 kg/m^2 (Class 1 Obesity, WHO), and blood pressure was 110/80 mmHg. Brain magnetic resonance imaging (MRI) revealed an absent pituitary gland in the sella/pituitary fossa and displaced infundibulum [Figure 2]. She was commenced on tablet acetazolamide 250 mg qds and tablet neurobion bd. Blood workup was unremarkable except for marginal elevation in prolactin 21.8 ng/ml (8.39–20.15 ng/ml). She was counselled on the guarded visual prognosis and was scheduled for a chiasmopexy by the neurosurgical team, but she declined due to financial constraint and unguaranteed visual restoration. She has since commenced visual rehabilitation.

Discussion

A diagnosis of PESS on background IIH was made in our index patient. The impression of "background IIH" was based on the preceding clinical history of significant worsening of a chronic headache in a 37-year-old obese Nigerian female, with tinnitus and papilloedema [Figure 1]. These clinical features align with criteria A and C in the diagnostic criteria for IIH according to the International Classification of Headache Disorders 3.[5] Furthermore, IIH is ×10 more likely to occur in females, particularly those of childbearing age, than in males. The risk is increased by about 20 fold in obese females.[5,6] Furthermore, in accordance with both the Modified Dandy and Friedman diagnostic criteria for IIH,[5,6] the MRI findings [Figure 2] are in keeping with the neuroimaging findings in IIH.

IIH is relatively uncommon, with an incidence rate of 0.9–2.2/100,000 population.[1,7] ES is the most common imaging sign in IIH (70%–94%).[1,8] The presence of ES on MRI scan, profound bilateral visual loss, and typical features of IIH in our index patient; but without clinical findings suggestive of iatrogenic or pathological pituitary atrophy/shrinkage, configures a picture of PESS in our patient.

On literature search, the documented mechanism for ES in IIH includes longstanding raised intracranial pressure (ICP) leading to osseous erosions, remodeling and widening of sella turcica, stretching of diaphragma sellae, then lax diaphragma sellae and space between the edge of diaphragma sella and the pituitary gland stalk prone to herniation from the chronic high ICP, with resultant subarachnoid herniation into the sella, and pressure on the pituitary gland.[4,8] Obesity in our index patient may have synergistically accentuated the chronically elevated ICP. Morbid obesity is known to induce hypercapnia which causes chronic CSF pressure elevation.[9]

Visual disturbances are uncommon in ESS; only a few (20%) manifest visual compromise.[1] When visual disturbances occur, they are usually late manifestations.[1] Documented pathogenetic mechanisms in ESS for visual disturbances include (1) optic nerves or chiasmal compression by surrounding structures; (2) dehiscence of diaphragma sellae with a resultant insult to the optic nerves/chiasm; (3) anatomical distortion and traction caused by the displacement of the optic chiasm and/or of pituitary apparatus; and (4) vascular compromise following sagging of the optic nerves/chiasm into the enlarged, ES.[2,10] Evidently, visual compromise could be prevented or corrected by chiasmopexy.[10]

Conclusion

A case of PESS on background IIH with severe bilateral visual loss is reported. Obesity in a female with IIH likely increases the risk to develop PESS. Multiple pathogenic mechanisms may be responsible for visual compromise in PESS.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients

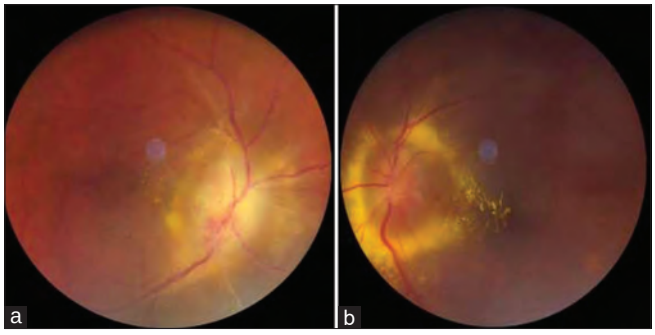

Figure 1: (a) Edematous pale disc with scanty hard exudates in the right eye. (b) Edematous erythematous disc with hard exudates extending toward the fovea with macular star configuration in the left eye

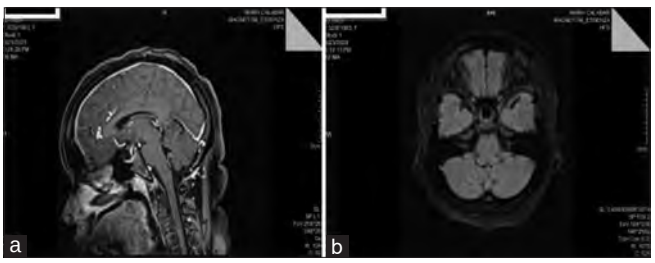

Figure 2: (a) Sagittal T1. T1-weighted images showing absent pituitary gland in the sella/pituitary fossa. The sella is filled with hypointense fluid, while the infundibulum is seen as a displaced hyperintense linear and dot-like structure within the sella. (b) Axial FLAIR. Axial FLAIR images showing absent pituitary gland in the sella/pituitary fossa. The sella is filled with hypointense fluid, while the infundibulum is seen as a displaced dotlike structure within the sella

understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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