

Case Report

A Hypertensive Early-Middle-Aged Man with Loss of Vision: a Rare Case of Primary Empty Sella Syndrome and Central Retinal Vein Occlusion

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Abstract:

Primary empty sella (PES) syndrome is a rare and benign condition usually with no symptoms, and, often revealed incidentally. But at times the syndrome manifests with a wide and variable range of ocular and endocrine features. Central retinal vein occlusion (CRVO) is in and of itself a rare cause of blindness, and its presence in the context of primary empty sella is even rarer. In this article, the authors report a case of a hypertensive man in his late thirties presenting to the medicine inpatient with headache and blindness of the left eye. Investigations came up with the diagnosis of PES syndrome with CRVO with macular edema. However, this patient had no overt endocrine problems.

Key words: Empty sella, primary empty sella, empty sella syndrome, central retinal vein occlusion, loss of vision

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Introduction

Empty sella syndrome (ESS), a benign condition, results from an abnormal extension of the subarachnoid space into the sella turcica containing the pituitary gland, through an incompetent diaphragm sella.¹ ESS can be primary or secondary.²

presence of certain risk factors. The most well-known are the weak diaphragm sella and raised intracranial pressure.³ In the majority of the cases with primary empty sella (PES), patients remain asymptomatic, the condition being revealed only incidentally on radiology.⁴ However, on rare occasions, PES can produce hormonal dysfunctions due to compression of the pituitary gland and, ocular manifestations due to traction and/or compression on the optic nerve.⁵ Common ocular features include loss of visual acuity, oculomotor nerve dysfunction, and glaucoma-like tunnel vision.^{3,4} Central retinal vein occlusion (CRVO) is per se a rare ocular condition with a prevalence of $0.11\% \pm 0.05\%$ per subject among Indian people and 0.1-0.7 % according to other population-based studies, more rarely it can be a complication of PES.^{5,6} This article is about a hypertensive man developing loss of vision in one eye due to CRVO in the presence of PES.

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Secondary ESS may result from surgery, radiotherapy, or medical treatment of tumors of the sellar regions or from infarction or hemorrhage of the pituitary gland.^{1,2} Though the primary ESS is idiopathic, it may occur in the

Figure 1: a) Red left eye with pterygium b) Dilated left pupil

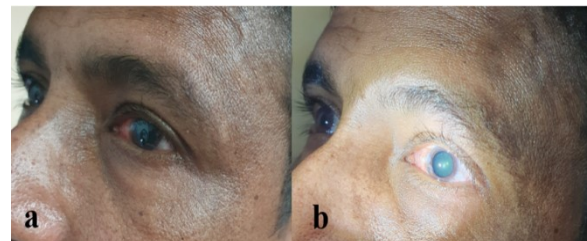


Figure 2: Color fundus photograph of the left eye showing the 'stormy sunset appearance'

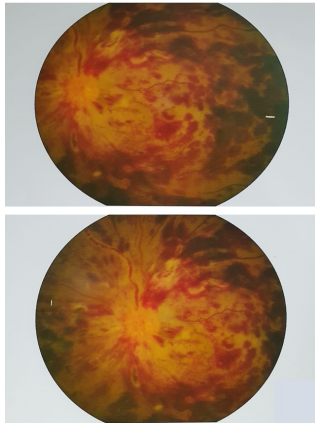


Figure 3: OCT showing macular edema of the left eye

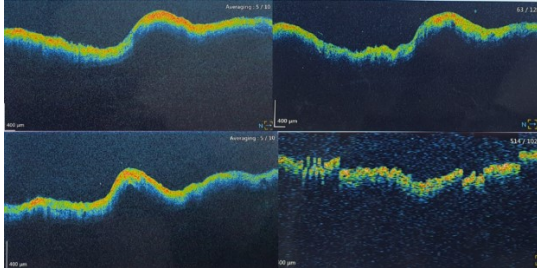


Figure 4: MRI of brain showing empty sella

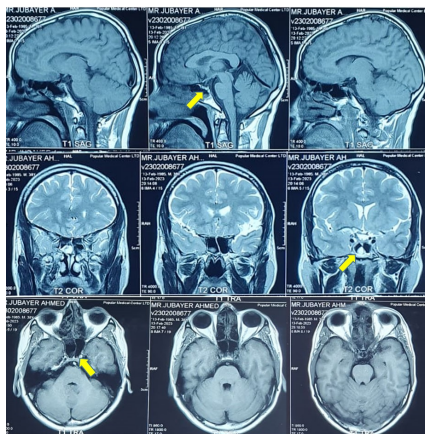
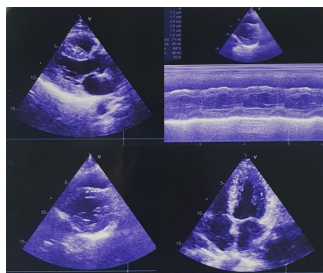


Figure 5: Normal echocardiography



Case Report

A 38-year-old, hypertensive, non-diabetic, and non-smoker man presented to the Medicine inpatient with a unilateral left-sided dull aching headache that began one month ago. The pain was not associated with nausea or vomiting and was partially relieved by taking paracetamol. He had concomitant redness in his left eye and within a short course, he experienced gradual loss of vision in the same eye which was painless. He had been on daily doses of Amlodipine 5mg, Olmesartan 40mg, and, Rosuvastatin 10mg. He gave no past history of any other major medical illness, surgery, irradiation, or trauma. On examination his blood pressure was 130/80 mmHg and, his pulse was regular, 84/ minute. He was afebrile (core temperature 98.6° F) and had no focal neurological deficit. On ophthalmological examination, his left eye was red and, had pterygium. His visual acuity was 6/6 on the right eye but reduced to mere 'perception-of-light' with dilated pupil on the left side (Figure 1). His intra-ocular pressure was normal. A routine ophthalmoscopic examination and, also the Color fundus photo of the left eye revealed a reddish optic disc with a poorly demarcated disc margin and, an obliterated physiological cup. Retinal blood vessels were very tortuous and, multiple widespread flame-shaped and blot hemorrhages were noted in all four quadrants and the macula. Cotton wool spots were also visible in all parts of the fundus. All these features were compatible with the classical 'stormy sunset appearance' impression of central retinal vein occlusion (Figure 2). The Optical Coherence Tomography (OCT) of the left eye showed a clear Vitreo Macular Interface. The neurosensory layer of the retina was elevated and, was irregularly shaped. There was optical empty space behind this layer and, features were suggestive of macular edema of the left eye (Figure 3). However, the Radial Peripapillary Capillary Complex (RPCC) was not well visualized in the OCT due to its 'out of field' location. As a part of the evaluation of the patient's persistent headache, Magnetic Resonance Imaging (MRI) of the brain was done which revealed otherwise normal brain features, except for the sella turcica being mostly occupied by CSF (cerebrospinal fluid) intensity

area and the pituitary appearing poorly defined and compressed. The images were compatible with the radiological diagnosis of an 'empty sella' (Figure 4).

His blood counts, serum creatinine, serum electrolytes, serum TSH (Thyroid Stimulating Hormone), blood glucose, and serum lipids- all were within normal ranges. His electrocardiogram was also normal and, echocardiography showed an ejection fraction of 64% (Figure 5).

So, with a diagnosis of CRVO of the left eye with macular edema (Macula Off), he was prescribed by the consulting ophthalmologist with anti-VEGF injections (3 doses of Bevacizumab) and anti-inflammatory Nepafenac eye drops. The patient was then referred to higher centers for neurosurgical and other evaluation and management.

Discussion

Empty sella, frequently an incidental radiological finding, is a misnomer and, a more appropriate term is 'intra-sellar arachnoidocele', as because here, the sella turcica is filled either fully or partially with CSF, due to herniation of the arachnoid through diaphragm sella.^{1,2,3} It is incidentally revealed in 8-38% during routine MRI of the brain and 5-10% at autopsy.⁷ ESS is the clinical condition arising from this pathology. The PES is idiopathic and, is clinically silent in most cases. It is common among obese and, multiparous women, with a female-to-male ratio of 5:1.^{7,8} Repeated pregnancies in the multiparous women, cause enlargement of the pituitary gland, gradually weakening the sellar diaphragm. Around half of the females with PES are obese and, at least two-thirds of them are overweight.⁷ Obesity-related intracranial hypertension and obstructive sleep apnea, which causes hypercapnia induced raised CSF pressure, are the presumed mechanisms of developing ESS in these patients.² The endocrine manifestations of PES, which are found in 19% of the patients, may be complex, subtle, or, only casually revealed during biochemical testing.^{7,9} In females with PES, menstrual abnormality in the form of amenorrhea is the most frequent (40%) feature. Galactorrhoea, hirsutism, sterility, and, gynecomastia are other endocrine problems

encountered.^{3,4} In the index case, a male person in his late thirties had no clinical evidence of endocrinopathy, but he could not undergo a thorough biochemical evaluation of endocrine function owing to time constraints from early and rapid referral. However, he had normal thyroid hormones on a routine investigation. He was a hypertensive man with well-controlled blood pressure with medication and, remained asymptomatic in regard to his PES before the appearance of his headache and blindness of one eye. Headache is the most frequently encountered problem among men with PES.¹ Though hypertension is an independent risk factor for ischemic CRVO¹⁰, in this case, the concomitant presence of empty sella might be a major contributor. The pituitary gland on MRI, which was done to exclude causes of headache, was grossly compressed.⁷ However, in the image, the sign of a compressed optic nerve was missing. He was a non-obese, non-diabetic, non-smoker, with normal lipid levels and, normal platelets, indicating that atherosclerotic mechanisms might not be the principal pathophysiologic factors of CRVO in this particular case. Miller et al and others reported cases of CRVO in COVID-19 pneumonia patients which might be due to a hypercoagulable state inferred by the condition.⁶ High intraocular pressure is sometimes responsible for CRVO and has been documented in several case reports since the twentieth century but the reported case in this article had normal ocular pressure.^{12,13,14} CRVO can be of two types. The mild type occurs due to leaky retinal vessels, manifesting with blurred vision on waking up in the morning and improving during the daytime. On the other hand, the severe type usually occurs due to thrombotic occlusion of the central retinal vein.^{9,10} Blindness in this severe type is due to the development of macular edema. Conventional treatment of macular edema by anti-VEGF and anti-inflammatory medication may theoretically improve edema and vision.^{14,15} But CRVO and macular edema in the presence of an underlying etiology of optic nerve compression requires neurosurgical intervention because case reports show promising results with surgery.¹⁶ Keeping this background information in mind, the patient

was referred for further neurosurgical evaluation.

Conclusion

Primary empty sella syndrome is a rare entity and, a concomitant CRVO is even rarer. Though empty sella is a benign condition, it can at times give rise to serious endocrine or ocular conditions. The physicians and the surgeons should be alert about this piece of knowledge.

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