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UNILATERAL PAPILLEDEMA DUE TO IDIOPATHIC INTRACRANIAL HYPERTENSION: A CASE REPORT

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ABSTRACT

Introduction: Papilledema is highly predicted and common to encounter in all causes of raised intracranial pressure including idiopathic intracranial hypertension; it is usually bilateral and symmetrical. Unilateral and asymmetrical papilledema is considered highly atypical and a rarity of presentation, posing significant diagnostic challenge to the poorly understood pathological phenomenon of IIH.

Case: We report a 40-year-old African female with idiopathic intracranial hypertension who fully fulfills the Modified Danddy Criteria; presenting with unilateral papilledema, with Left eye fundoscopic examination showing extensively hyperemic and swollen optic disc with an associated tortuously engorged retinal vein. Brain imaging via MRI revealed partial empty sella, bilateral prominent fluid signal projection that is more evident in the left optic nerve sheath and a prominent meckel's cave on the left side measuring (6.1 mm) in the transverse diameter. MRA concluded an attenuated most lateral side of the left transverse sinus. Lumbar puncture was attempted and revealed an opening pressure of (45 cmH2O), and otherwise normal CSF cytology and chemical composition. Significant symptomatic relief was depicted upon lumbar puncture.

Discussion: papilledema is universally encountered as a bilateral and symmetrical phenomenon; thus, presentation of IIH with unilateral papilledema has been under extensive research recently in attempts to contemplate the exact pathophysiology. Amongst many other proposed theories, we endorse the variation of the optic canal diameter as a potential mechanism for asymmetry of papilledema in IIH. As larger optic canal diameter is postulated to be associated with higher CSF pressure force transduction with subsequent optic nerve damage.

Conclusion: Unilateral and asymmetrical papilledema is considered substantially unique and anecdotal rarities of presentation of IIH; posing a significant diagnostic troublesome and dilemma. Physicians should be aware and highly vigilant of such deviations of usual presentations to avoid consequential diagnostic and management adverse outcomes and unwanted complications.

Keywords: Benign Intracranial Hypertension, Papilledema, Symptomatic Relief

1. INTRODUCTION

Idiopathic Intracranial Hypertension (IIH) is a neurological disorder of unknown etiology that affects young, obese females with not yet identifiable risk factors to yield a poorly understood syndrome; that is characterized by elevated intracranial pressure, headache, visual disturbances and papilledema Giridharan et al. (2018). The term (IIH) implies to isolated raised intracranial pressure (ICP) that is unrelated to an intracranial, meningeal or cerebral venous pathology Mollan et al. (2018). With a prevalence rate of (3 in 100,000) Biousse et al. (2012), IIH has received considerable attention by the scientific literature in regard to its risk factors, associations and clinical variations; considering the highly elusive pathophysiology of the disease Corbett et al. (1982) .

Several studies and scientific theories have been pledged for the purpose of understanding the pathophysiological basis of IIH: specifically, increased secretion, decreased absorption of cerebrospinal fluid (CSF) and cerebral venous outflow obstruction due to venous sinus stenosis. Giridharan et al. (2018), Corbett et al. (1982).

With papilledema and transient visual loss being the most common to encounter; IIH has unique and vast ocular and visual presentations that range from asymptomatic optic nerve edema to various visual field defects and progressive optic nerve atrophy with complete blindness Lee & Wall (2012)

Since papilledema is looked at as edema of the optic nerve head, secondary to raised intracranial pressure; it's highly predicted and common to encounter in all causes of raised intracranial pressure including idiopathic intracranial hypertension, cranial hemorrhages, mass lesions etc. Bono & Quattrone (2009). It's hypothesized that pathologically elevated intracranial pressure renders the optic nerve susceptible to physical pressure, ischemia and subsequent degeneration Bono & Quattrone (2009).

And while papilledema is often bilateral and only unilaterally encountered in minor etiologies of increased intracranial pressure; it is now a generalization, that papilledema in IIH is almost always bilateral Sher et al. (1983), Friedman & Jacobson (2002). Thus, cases of IIH with unilateral papilledema and are considered highly unique and anecdotal rarities of presentation posing a considerable diagnostic dilemma with potential consequences of misdiagnosis, delayed management and occurrences of complications.

2. CASE PRESENTATION

We report a 40-year-old, multiparous, previously healthy, Sudanese female who presented with headache history for one year. The patient describes headache as on-and-off, severe, unilateral, right-side distributed and of throbbing nature that is markedly worsened by straining, head manipulation and motion. As the conditioned progressed, headache started to acquire a more persistent course instead of episodic attacks and develop markedly disturbing associations of vomiting, visual disturbances, eye pain (retro-orbital), lacrimation, photophobia and phonophobia. Visual disturbances were unilateral and only confined to the left eye; initially reported as mild blurring of vision, flashes and floaters, accommodation deficits and diplopia to slowly progress into more serious ailments of blurred vision, visual fields defects and scotomas, frank aumarosis fugax-like phenomena and complete but reversible obscuration of vision in the left eye. The

patient denies any presence of other neurological deficit, nor symptoms of meningeal irritation or disturbances of higher cortical functions.

On bedside examination, the patient weighted (91 kg), with (156 cm) height and BMI of (37.39 kg1/m2), that is well orientated to time, place and person with preserved normal intellect. All vitals were within normal range, neurological examination revealed no motor or sensory deficits depicted; muscle power, reflexes and dermatomoal sensory modalities were all normal. There were also no signs of cortical impairment or meningeal irritations. Eyes examinations were significant for decreased visual acuity of the left eye, with normal both direct and consensual light reflex and preserved color vision in both eyes. Fundoscopic examination of the right eye came out normal; however, left eye examination showed extensively hyperemic and swollen optic disc with an associated tortuously engorged retinal vein.

Brain imaging via MRI Figure 1 and Figure 2 revealed partial empty sella, bilateral prominent fluid signal projection that is more evident in the left optic nerve sheath and prominent meckel's cave on the left side measuring (6.1 mm) in the transverse diameter. Subsequent MRA Figure 3 concluded an attenuated most lateral side of the left transverse sinus.

Figure 1

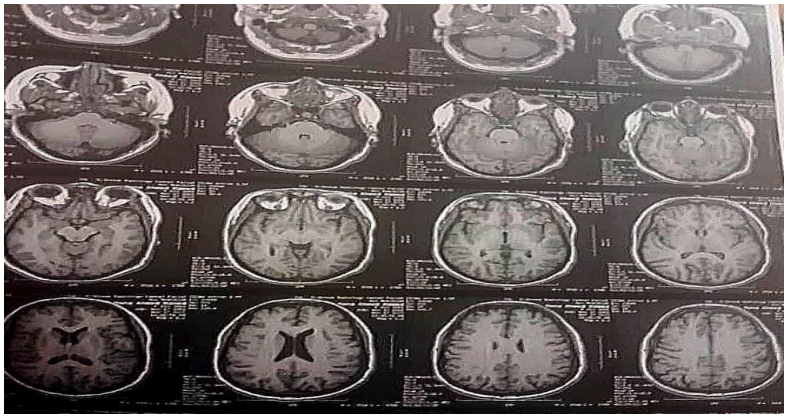

Figure 1 Brain MRI Showing Partial Empty Sella Features, with Enhanced Fluid Signal Projection of the Left Optic Nerve Sheath

Figure 2

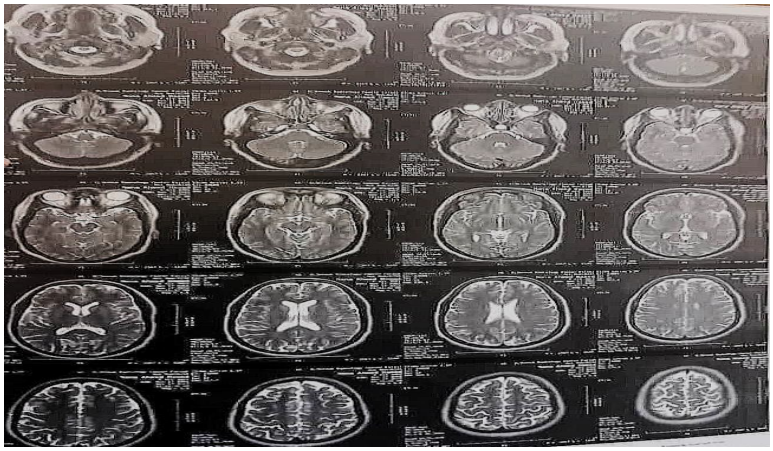

Figure 2 Showing Furthermore Features of Empty Sella Phenomenon, with more Prominently Enlarged Meckels Cave on the Left Side

Figure 3

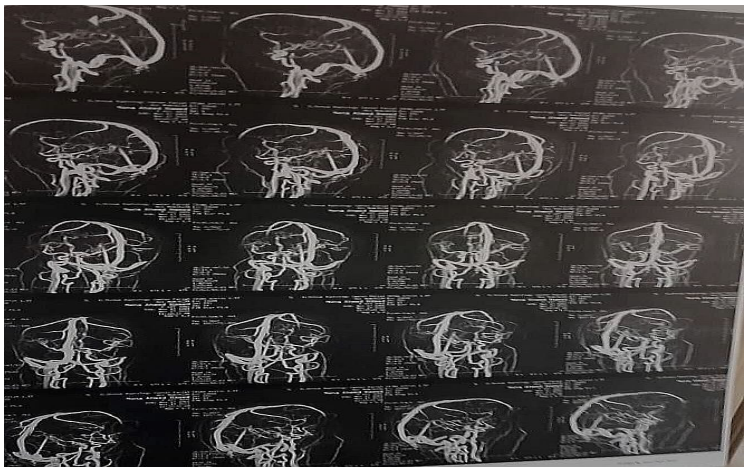

Figure 3 Brain MRV Showing an Attenuated Most Lateral Side of the Left Transverse Sinus of the Brain

Lumbar puncture was attempted and revealed an opening pressure of (45 cmH2O), and otherwise normal CSF cytology and chemical composition. Significant symptomatic relief was depicted following the lumbar puncture, IIH was confirmed, and the patient was enrolled in the neurological management protocol of IIH as per guidelines.

3. DISCUSSION

We present a unique case of a 40-year-old, African female with Idiopathic Increased Intracranial Hypertension (IIH); endorsing relentless symptoms and signs of increased intracranial pressure, preserved consciousness and cognition, intact motor and sensory modalities, significantly elevated CSF opening pressure and otherwise normal CSF analysis, and with no evidence of any other possible etiology for the increased intracranial pressure. Although our patient fully meets all the Modified Dandy Criteria Lee et al. (2020), it is the presentation with unilateral papilledema what makes such case highly unique and uncommon to encounter during clinical practice.

Papilledema is a common optic pathology, characterized by edema of the optic nerve head, secondary to raised intracranial pressure. It has been constantly linked to numerous causes of increased intracranial pressure including benign intracranial hypertension (BIH). Bono & Quattrone (2009), Lee et al. (2020).

When exceeding the physiological limits; pathologically elevated intracranial pressure is transmitted to the subarachnoid space surrounding the optic nerve rendering the ganglion cells prone to damage by physical compressive forces and subsequent micro-vascular ischemia. Bono & Quattrone (2009). The etiologies of papilledema are vast and hold considerable overlap and variation in the pathogenesis, degrees of severity, pattern of progression and clinical presentation; and it is now well observed that papilledema seems to be almost always of bilateral occurrence. Bono & Quattrone (2009)

As for any other causes of increased intracranial pressure, IIH holds papilledema as one characteristic manifestation; it is also typically encountered bilateral and symmetrical Digre (2003). As for such, the asymmetry and unilaterality of papilledema in IIH are considered highly atypical rarities of presentation making

up less than 4% of IIH cases. Brosh & Strassman (2013) The relative lack of literature regarding the pathophysiology of IIH caused a considerable spike in the number of hypotheses tackling the variations of its associated papilledema. Differences in bony optic canal diameter and optic nerve sheath anomalies to are two main postulated mechanisms that had been held responsible for unilateral papilledema in IIH in multiple studies Bidot et al. (2016), Swinkin et al. (2022) reflecting poor visual function and severe papilledema or optic atrophy associated with a larger optic canal. Potential mechanisms include alteration of local CSF flow or bony remodeling at the optic canals Bidot et al. (2016). It's seems that the diameter of the optic canal is directly proportional to the degree of elevated pressure transmission and subsequent optic nerve damage. Conversely, a relatively higher protection against papilledema is provided by the smaller sized optic canal via reduced transduction of CSF pressure from the suprasellar cistern to the perioptic subarachnoid space Swinkin et al. (2022). Optic nerve sheath anomalies rendering the optic nerve vulnerable to damage by the pressure forces is a less supported hypothesis for the development of unilateral papilledema in IIH Brosh & Strassman (2013), as some studies concluded that patients of IIH with asymmetrical papilledema had no gross differences between the two optic sheaths. Huna-Baron et al. (2001) However, other reports proposed microscopic nerve sheath anomalies and disturbed trabecular meshwork as potential contributors to differences in CSF pressure transduction; such anomalies can reflect no gross changes in the optic nerve sheaths when visualized via MRI and CT imaging modalities. Bidot et al. (2016), Killer et al. (2003).

Finally, the well documented changes in the lamina cerebrosa are one strong potential explanation for the development of such rarity of presentation. Age-related, trauma, surgery and repetitive inflammatory changes cause significant loss of compliance of the lamina cerebrosa; greatly potentiate the CSF pressure transduction and subsequent damage due to axoplasmic flow stasis Lepore et al. (1992). This is strongly supported by the observation of higher IIH frequency among older patient's groups. Lepore et al. (1992), Sapkota et al. (2018).

CONFLICT OF INTERESTS

None.

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