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DIAGNOSIS AND TREATMENT OF IDIOPATHIC INTRACRANIAL HYPERTENSION AT A TERTIARY CARE MULTISPECIALTY HOSPITAL IN NORTHEAST INDIA

Neurology

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ABSTRACT

Idiopathic intracranial hypertension (IIH) is a condition of unknown etiology and is characterized by elevated intracranial pressure. In this case series we describe the clinical features and responses to treatment of patients presented with IIH. It is a retrospective study of four patients who were presented in our hospital between August 2020 and December 2021 and all patients ethnically belonged to northeast India. The patients were evaluated and diagnosed based on history, physical examination, imaging as well as modified Dandy criteria. Low doses of acetazolamide were recommended for all the patients. Timely diagnosis and accurate treatment of IIH is of utmost importance to avoid any irreversible visual loss.

KEYWORDS

Blindness, headache, idiopathic intracranial hypertension, intracranial pressure

INTRODUCTION

Idiopathic intracranial hypertension (IIH) is an increase in intracranial pressure (ICP) in the absence of any inflammation, structural obstruction or mass lesion [1]. It is predominantly seen in obese women of childbearing age but can also occur in persons of different age groups [2]. Diagnosis is usually made using the modified Dandy criteria that includes symptoms such as headache, nausea, pulsatile tinnitus, transient visual obscuration (pappilloedema), vomiting, absence of any localizing signs except for abducens nerve palsy, normal cerebrospinal fluid (CSF) composition, and normal brain parenchyma [3, 4] As it is a diagnosis of exclusion, therefore other causes of increased ICP must be sought with history, physical evaluation, imaging tests, and cerebrospinal fluid examination before the diagnosis can be made [5]. More awareness is needed about this condition and protocols for diagnosis and management needs to be set up to achieve good outcome. We report here four cases of IIH between August 2020 and December 2021 that were diagnosed and treated at our hospital. The main aim of this study was to document the clinical features and responses to treatment of the IIH patients from Northeast India attended in our hospital.

Case 1

A 14-year-old girl was presented to our neurology outpatient unit with complaints of headache and blurred vision for two weeks. She had a history of hypothyroidism and was on thyroxine therapy. On evaluation her blood pressure was 110/80 mm Hg, visual acuity was perception of light (PL) positive, pupils were mid dilated in both eyes and had sluggish reaction. Optic disc examination revealed marked pappilloedema and splinter hemorrhage around the disc (Fig 1 A). Magnetic resonance imaging (MRI) brain showed bilateral optic nerves that appear mildly edematous with prominent CSF spaces. High opening pressure CSF therapeutic spinal tap was done and 15 ml of CSF was drained out. She was commenced on 250 mg acetazolamide and 40 mg Lasix thrice daily. Her headache subsided gradually and her vision improved from the eleventh day. Her one month follow up revealed no headache and vision was 6/6.

Case 2

A 15-year-old boy was brought to our neurology outpatient unit with complaints of headache and double vision (diplopia). On evaluation his blood pressure was 110/80 mm Hg and visual acuity was 4/6. MRI brain revealed enlarged perioptic CSF sheaths surrounding the optic nerves on either side with cupping of the optic discs (Fig. 1B). Patchy increased signal on T2 weighted MRI images in the retrobulbar portions of the optic nerves was observed. The optic nerves and brain parenchyma were normal. He was managed with 250 mg acetazolamide and 40 mg Lasix twice daily. His headache subsided on fourth day, diplopia resolved, and acuity of vision was 6/6.

Case 3

A 28-year-old housewife experienced dimness of vision in both eyes and gradually experienced loss of vision. She had a history of hypertension and was on irregular antihypertensives. She used oral contraceptive pill as a measure of birth control. On evaluation her blood pressure was 150/90 mm and visual acuity was PL +ve bilaterally. MRI brain revealed enlargement of subarachnoid space around the optic nerves with tortuosity and mild intraocular protrusion of optic nerve head on both sides (Fig. 1C). She was managed with 250 mg acetazolamide and 40 mg Lasix thrice daily and anti hypertensives. Her vision started improving and visual acuity became 6/6 from fourth day onwards.

Case 4

A 63-year-old man was admitted to our hospital with complaints of headache, right sided weakness, difficulty in speaking, and confusion. Clinical features were suggestive of stroke infarct involving left middle cerebral artery (MCA) territory. On evaluation his blood pressure was 150/80, GCS E4V3M5, and pupils were 3 + b/l. MRI brain revealed prominence of the perioptic CSF spaces on sides, optic disc cupping, and prominent empty sella that were suggestive of benign idiopathic intracranial hypertension. There was associated diffuse irregular leptomeningeal and multiple focal parenchymal enhancements in bilateral frontoparietal region that suggested chronic meningitis/ chronic granulomatous lesion. MRI brain showed prominence of subarachnoid spaces in perioptic and supra-territorial compartment with pappilledema and empty sella (Fig. 1D). CSF therapeutic and diagnostic tapping was done. MRI after one month showed significant resolution of the parenchymal lesion in the bilateral frontoparietal region. The patient was treated with ceftriaxone for two

weeks and steroid 4 mg thrice daily. His symptoms improved from fifth day onwards.

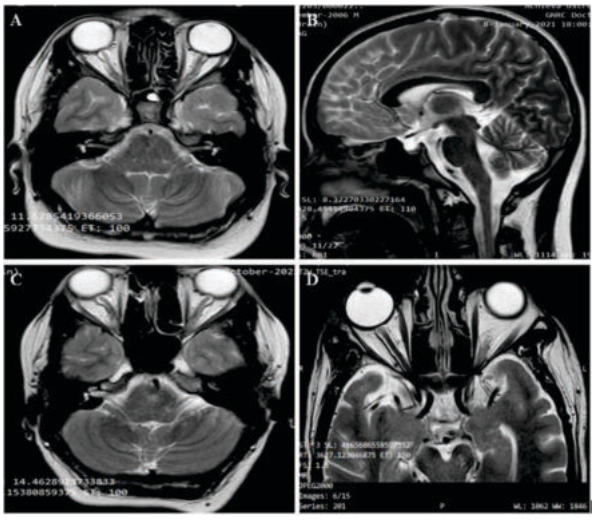

Fig 1: Magnetic resonance imaging (MRI) of the brain.

A) Mild vertical tortuosity of the bilateral optic nerves noted with prominent subarachnoid space around them and flattening of posterior sella of bilateral orbits (pappilloedema).
B) Prominent empty sella was observed.
C) Fortification with prominent sub arachnoid space around bilateral optic nerves. Bilateral pappilloedema was observed.
D) Fortification with prominent sub arachnoid space around bilateral optic nerves with bilateral pappilloedema.

DISCUSSION

IIH is characterized by elevated ICP [6]. The underlying pathogenesis of IIH is uncertain however raised ICP is the uniform characteristic feature. It appears that changes in the volume of blood, CSF and brain tissue influence intracranial pressure. IIH therefore likely represents a disorder of CSF regulation, potentially through CSF over secretion or impaired drainage. Factors such as medications (tetracycline and its derivatives, cyclosporine, lithium, nalidixic acid, nitrofurantoin, oral contraceptives, levoergosterol and tamoxifen), endocrine abnor malities, polycystic ovarian syndrome, anaemia and obstructive sleep apnoea have been found in some studies to be associated, precipitate or worsen IIH. Studies have shown that IIH is more common in women and obese individuals. Despite a high predilection for obese young women, IIH can occur in children, older adults, and in non-obese persons of either sex. We reported two males and two females with IIH in our study.

Our study used the modified Dandy criteria for diagnosis of IIH. Blood pressure needs to be clearly monitored to exclude malignant hypertension. All four patients in our study experienced headache. This is in accordance with some other studies that reported headache as the common presenting symptom [4,7] Headache of IIH is typically pulsatile, global, worsens after walking, and exacerbated by measures that increase ICP. Features consistent with nausea and photophobia have also been reported. Back, neck, and radicular pain also occurs frequently.Neck pain was reported in one of our cases. Pappilloedema was the most common symptom observed in our study. Pappilloedema commonly results in transient visual obscuration and can present as brief episodes of monocular or binocular vision loss.

Transient visual obscurations were experienced by three patients in this study. Untreated pappilloedema causes progressive to irreversible vision loss and secondary optic atrophy. Optic nerve function must be assessed by testing the visual acuity, pupillary responses, and colour vision. The level of pappilloedema should be documented as this helps in follow-up.

Brain imaging tests such MRI or Computed Tomography (CT) scan is important for excluding space-occupying lesions and other causes of raised ICP such as hydrocephalous and cerebral venous thrombosis to make the diagnosis of IIH [8]. Imaging abnormalities such as empty sella or flattening of the pituitary, tight arachnoid spaces, flattening of the posterior globe, protrusion of the optic nerve head, enhancement of the pre laminar portion and vertical tortuosity of the optic nerve sheath are associated with raised ICP in IIH patients. MRI was performed in our IIH patients.

One patient was on oral contraceptive pill at the time of presentation and this was discontinued immediately. Acetazolamide, a potent enzyme inhibitor of carbonic anhydrase is currently considered the main stay of pharmacological management of IIH [9]. It works by impending the activity at the choroid plexus reducing CSF secretion. In a recent systemic review identified two randomized control trials for the use of acetazolamide in IIH. Dosage is usually started at 1-2 mg/day. Most patients cannot tolerate high doses due to side effects such as lethargy, nausea, altered sense of taste, and paraesthesia. All four patients were started on acetazolamide. The dosage range was 250 to 500 mg/day. Doses had to be altered to suit the patient's toleratibilty to acetazolamide. In some patients lasix was added along with acetazolamide. Topiramate has been reported to be efficacious in relieving headache caused by IIH. However, none of our patients was put on it.

Surgical procedures are usually required in patients with a fulminant onset or when other treatments have failed to prevent vision loss [9]. Options include optic nerve sheath fenestration or a CSF diversion surgery such as ventriculo/lumbo-peritoneal shunting. It causes a rapid reduction in ICP often with a subsequent improvement in signs and symptoms particularly headaches. Surgery was also not indicated in any of our patients.

In conclusion, IIH can no longer be described as a rare disease in this part of the country. Clinicians must be prepared to see more cases of IIH. It is important to have a high index of suspicion and those at the tertiary health care needs to be able to diagnose and refer promptly for treatment to prevent permanent vision loss.

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