

Benign Intracranial Hypertension Secondary to Prolonged Steroid Usage: A Case Report

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Abstract

Benign intracranial hypertension (BIH), also known as idiopathic intracranial hypertension or Pseudotumor cerebri, is a syndrome of signs and symptoms of increased intracranial pressure without causative mass or hydrocephalus identified. We report a case of 21-year-old male who was a known asthmatic since 11 years, on oral steroids without any medical supervision presented with diplopia and blurring of vision of 10 days duration with recent onset of squint and cushingoid features. Case was diagnosed as BIH based on clinical and radiological criteria. Treatment was initiated with Tab. Acetazolamide 250 mg twice daily for 2 weeks along with gradual tapering of steroids. On the follow-up after 4 weeks, a dramatic improvement in visual symptoms with resolution of papilledema was observed. BIH is commonly associated with sudden withdrawal of steroids. We report this case as it was resulted from prolonged steroid usage and which improved after gradual tapering of the steroid along with acetazolamide therapy.

Keywords: Benign intracranial hypertension, Diplopia, Papilledema, Steroids

INTRODUCTION

Benign intracranial hypertension (BIH) is a disorder defined by clinical criteria that include symptoms and signs isolated to those produced by increased intracranial pressure (ICP) (e.g. headache, papilledema, vision loss), elevated ICP with normal cerebrospinal fluid composition, and no other cause of intracranial hypertension evident on neuroimaging or other evaluations.¹ While once called BIH, to distinguish it from secondary intracranial hypertension produced by a neoplastic malignancy, it is not a benign disorder. Many patients suffer from intractable, disabling headaches, and there is a risk of severe, permanent vision loss. The presence or absence of findings on magnetic resonance imaging (MRI) does not appear to predict visual outcomes.² Recurrence of symptoms may occur

in 8-38% of patients after recovery from an episode of BIH or after a prolonged period of stability.³ A subset of individuals with idiopathic intracranial hypertension (IIH) have a more malignant or fulminant course with rapid development of vision loss within a few weeks of symptom onset which is referred to as fulminant IIH.⁴ To limit the severity of permanent vision loss, more aggressive surgical treatment measures are considered at the outset, often with temporizing measures (e.g. serial lumbar punctures, lumbar drain, and/or corticosteroids) employed until surgery can be performed. BIH is a disorder that primarily affects women of childbearing age who are overweight.⁵ Review of literature shows that BIH is commonly associated with withdrawal of steroids. Development of BIH, while on maintenance steroid therapy or increase in its dose is an uncommon phenomenon in adults.

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CASE REPORT

A 21-year-old male patient admitted with a complaint of headache and diplopia of 10 days duration. He was born to a non-consanguineous parents and developmental history was also normal. There was no family history of neurological

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disorders. He reported that he was a known case of asthma for the past 11 years, and taking oral prednisolone therapy since then without any medical supervision till the date of presentation. He had weight gain with retarded growth for the past 5-6 years. He had intermittent blurring of vision with new onset squint of 10 days duration.

On physical examination, he was noticed to have retarded growth with central fat distribution, Buffalo hump. He had delayed development of secondary sexual characters with absent axillary and pubic hair with a hypoplastic penis.

The neurological examination was noteworthy for diplopia with right-sided 6th nerve paresis. Laboratory evaluations including a complete blood count, blood electrolytes, urea, creatinine, and liver enzymes gave results at normal range.

Ophthalmologic evaluation revealed acute bilateral papilledema and diminished visual acuity. Cerebrospinal fluid pressure (CSF) pressure manometry records an increased CSF pressure of 50 mm of water. CSF analysis was normal.

CT scan cranial MRI was normal. MR venogram shows multiple collaterals and reformation of distal venous sinuses seen at sigmoid sinus and dominant right jugular vein. Since the plain study does not suggest any thrombosis, this appearance most likely represents BIH.

Following the diagnosis of BIH with cushing's syndrome, acetazolamide 250 mg twice daily was prescribed for 2 weeks along with gradual tapering of the steroids. His sixth nerve palsy resolved gradually over 1 month. On the review, 2 months later his optic disc had returned to normal and he had remained asymptomatic. After tapering the drug, patient's visual improvement and the resolution of the papilledema were very dramatic. Such a causal relationship strongly suggests prolonged steroid to be the cause of the BIH [Figures 1-5].

DISCUSSION

The first report of BIH was by the German physician Heinrich Quincke, who described it in 1893 under the name serous meningitis.⁶ The term "pseudotumor cerebri" was introduced in 1904 by his compatriot Max Nonne.

BIH or pseudotumor cerebri is a syndrome that is defined by increased ICP, absence of ventriculomegaly, no evidence of intracranial extensive lesion and normal CSF composition.

The main symptoms are headache, nausea, and vomiting, as well as pulsatile tinnitus, double vision and other visual

symptoms. If untreated, it may lead to swelling of the optic disc in the eye, which can progress to vision loss. The increased pressure leads to compression and traction of the cranial nerves that arises from the brainstem. Most commonly, the abducens nerve (sixth nerve) is involved.

Modified Dandy criteria⁷

Symptoms of raised ICP (headache, nausea, vomiting, transient visual obscurations, or papilledema)
 No localizing signs with the exception of abducens (sixth) nerve palsy
 The patient is awake and alert
 Normal CT/MRI findings without evidence of thrombosis
 LP opening pressure of >25 cm H₂O and normal biochemical and cytological composition of CSF
 No other explanation for the raised ICP

CT: Computed tomography, MRI: Magnetic resonance imaging, CSF: Cerebrospinal fluid pressure, ICP: Intracranial pressure

Our case satisfies all the six modified Dandy criteria, and hence labeled as BHI.

The strongest evidences for the association with BIH exists for young age, female sex, obesity or weight gain, prolonged use of tetracyclines or vitamin A. Steroid withdrawal and Addison's disease are clearly associated with BIH.⁸

BIH is most commonly associated with abrupt withdrawal of steroids and it was evident from the literature that symptoms were attenuated after reintroduction of steroid therapy. However, the presentation of BIH during steroid therapy and regression of symptoms after tapering of steroids was a very rare phenomenon in adults as evident in our case.

On average, BIH occurs in about one per 100,000 people and can occur in children and adults. The median age at diagnosis is 30.⁹ BIH occurs predominantly in women, especially in the ages 20-45, who are 4-8 times more likely than men to be affected. Overweight and obesity strongly predispose a person to BIH: Women who are more than 10% over their ideal body weight are 13 times more likely to develop BIH, and this figure goes up to 19 times in women who are more than 20% over their ideal body weight. In men this relationship also exists, but the increase is only five-fold in those over 20% above their ideal body weight.

A theory for the mechanism of BIH posits that either increased blood flow to the brain or increase in the brain tissue itself may result in the raised pressure. Another suggested theory was restricted venous drainage from the brain may be impaired resulting in congestion. Many patients with BIH have narrowing of the transverse sinuses.¹⁰

The treatment goal for patients with BIH is to preserve optic nerve function while managing increased ICP. Weight control is recommended for obese patients.¹¹



Figure 1: Cushingoid features



Figure 4: Magnetic resonance imaging T1 W imaging

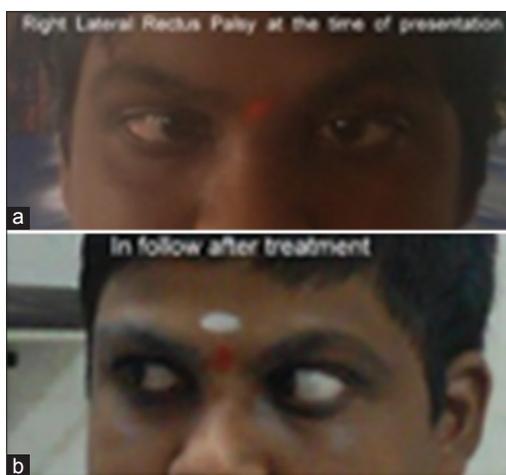


Figure 2: (a) right lateral rectus palsy (b) complete resolution after treatment



Figure 5: Magnetic resonance venogram



Figure 3: Magnetic resonance imaging T2 W imaging

As our patient has blurring of vision with decreased visual acuity he is treated with acetazolamide therapy along with gradual tapering of steroids.

Patients without visual loss most often are treated with a carbonic anhydrase inhibitor like acetazolamide,¹² furosemide to lower the ICP. The mechanism of action of acetazolamide is likely multifactorial. It has been found to reduce CSF production. Furthermore, it changes the taste of foods and sometimes causes anorexia aiding in weight loss. Furosemide has also been used to treat BIH. It has been well documented that furosemide can lower ICP. It appears to work by both diuresis and reducing sodium transport into the brain primary headache prophylaxis with amitriptyline, propranolol, or other commonly prescribed migraine prophylaxis agents, or with topiramate. Topiramate has also been used to treat BIH since it has carbonic anhydrase inhibitor activity and weight loss commonly occurs. In studies to date, it appears comparable to acetazolamide.¹³

If visual function deteriorates, surgical or other invasive interventions may be considered. Interventions include the following. Optic nerve sheath fenestration (decompression).¹⁴

Cerebrospinal fluid diversion (i.e. via a lumboperitoneal or ventriculoperitoneal shunt).¹⁵ Intracranial venous sinus stenting.¹⁶

CONCLUSION

Headache and visual symptoms after prolonged steroid usage should prompt the evaluation of BIH. The etiological factor for BIH in our case includes prolonged steroid therapy and treatment was initiated with acetazolamide along with gradual tapering of steroids. Increased awareness of this adverse effect among physicians is important, as steroids are widely used medications for varied conditions.

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