Presumed Idiopathic Intracranial Hypertension: A Case Report and Literature Review

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SUMMARY

BACKGROUND: Idiopathic intracranial hypertension is a rare neurologic disorder which is largely self limiting and occurs as a result of increased intracranial pressure. Papilloedema is the most significant finding and may progress to visual loss.

CASE REPORT: The case of a 45 year old obese female, who presented with a 9 month old history of headache and on examination was found to have papilloedema is presented here. The patient received no medical or surgical intervention and presented 5 months later with resolution of the symptoms.

CONCLUSION: Idiopathic intracranial hypertension is a sight threatening disease in which raised intracranial pressure is associated with increased body mass index. Proper assessment and close monitoring of patients who present with these features is essential in preventing visual loss.

KEYWORDS: Headache, papilloedema, idiopathic intracranial hypertension.

INTRODUCTION

Idiopathic intracranial hypertension(IIH), previously called benign intracranial hypertension(BIH) or pseudo tumour cerebri (PTC) is a neurologic disorder characterised by increased intracranial pressure in the absence of a tumour or other diseases[1,2,3] The main symptoms include headache, nausea, vomiting, pulsatile tinnitus and visual disturbances. The visual symptoms include visual loss, double vision due to sixth cranial nerve affectation, transient visual obscurations and enlargement of the blind spot.

IIH has no known aetiology. Ninety percent of patients are obese women of child bearing age who are amennorrhaeic.[3] Drugs such as high doses of vitamin A, corticosteroids (more commonly its withdrawal),certain antibiotics particularly tetracycline, nalidixic acids. Patients with certain conditions such as hypoparathyroidism, adrenal adenomas, chronic respiratory insufficiency, iron deficiency anemia and renal disorders have also been found to have IIH.[2,4]

The diagnosis of IIH is usually based on symptoms and signs solely attributable to elevated cerebrospinal fluid pressure (CSF), normal CSF composition, normal neuroimaging studies and no other etiology of intracranial hypertension identified.

The aim of treatment is the prevention of visual loss and to alleviate symptoms. Treatment could be medical or surgical.

CASE REPORT

C.C a 45 year old negroid female presented to the outpatient clinic of the eye department with a nine month history of constant headache associated with nausea and ringing in the ears. The only significant drug history was the ingestion the drug Cordy Active(a drug containing cordyceps sinensis, zinc, vit E and carotene ,usually taken as a supplement) which the patient emphasized preceeded the onset of symptoms.

On examination the patient was found to be in good general health but obese(BMI = 34kg/mm^2). Ocular examination revealed normal visual acuity in both eyes (6/6) with mild ptosis of the left upper eyelid. The intraocular pressure was within normal limits (16mmHg in the right eye and 14mmHg in the left.)

Dilated fundoscopy revealed hyperemic slightly elevated discs in both eyes with indistinct margins superiorly, nasally and inferiorly and a few retinal flame shaped haemorraghes.

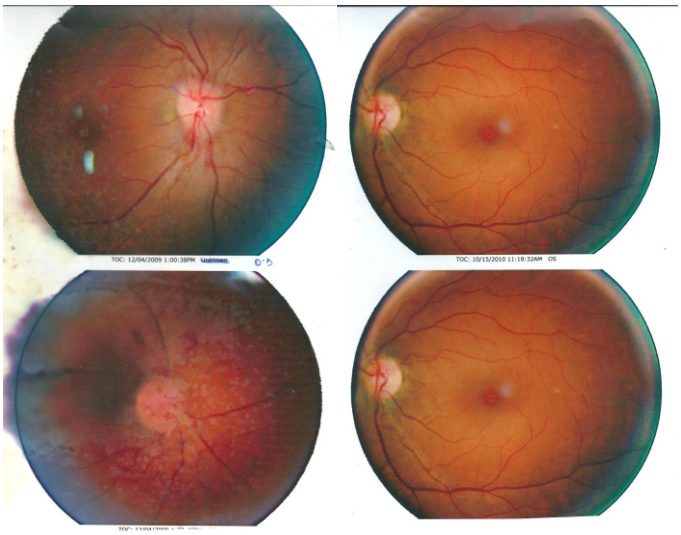

Figure 1. Fundus picture at presentation wit blurred disc margins (left) and after 5 months (right) with distinct margins.

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CT scan was essentially normal. Central visual field results revealed slight enlargement of the blind spot and small areas of relative scotoma superiorly in both eyes. The patient was not placed on any medication but asked to return for a lumbar puncture. She was however lost to follow up thereafter. Five months later patient was seen in the clinic with all the symptoms resolved and on fundoscopy disc margins were found to be distinct.

DISCUSSION

Idiopathic intracranial hypertension is largely a diagnosis of exclusion.[5,6] Patients who present with symptoms of increased intracranial pressure and papilloedema are usually considered a clinical emergency until proven otherwise by neuroimaging studies. [5] However where these imaging studies reveal no abnormality these cases are treated as cases of IIH.

The modified Dandy's criteria for diagnosis of IIH are as follows,symptoms of raised intracranial pressure (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 , lumber puncture opening pressure of >25 cmH2O and normal biochemical and cytological composition of CSF, no other explanation for the raised intracranial pressure.

Typically IIH most commonly affects women who are overweight between the ages of 15 and 45.[6] The peak incidence seems to occur in the 3rd decade with a female preponderance ranging from 2:1 in some studies to 8:1 in others.[3,6] However it can develop at any age including children.[6] The index patient falls within this age range and has a BMI of 34kg/mm^2 and is female . The condition appears to be diagnosed in many areas of the world with increasing frequency probably due to increase prevalence of obesity and increased awareness of the condition.[6] The pathogenesis of raised ICP in this condition remains unclear, however a number of factors such as CSF dynamics, obesity, drugs, prothrombotic abnormality have been suggested to play a role in its pathogenesis.[6,7] In the case of our patient even though a lumbar puncture was not done but she had symptoms of raised ICP, headache and nausea. These meet the modified Dandy's criteria for the diagnosis of IIH. Also the ingestion of the drug which is the only known factor she had recently introduced in her system may have played an aeitiological role in our case report as she gave a history of relief of symptoms on cessation of the drug. The only case of idiopathic intracranial hypertension reported in Port Harcourt by Onwuchekwa et al had drug and obesity as the likely aeitiological factors in their series.[8]

Our patient also had tinnitus which is known to exist in 8-60% of patients with this condition and is said to resolve when CSF pressure normalizes.[3,4,9]

Ophthalmologic features which are usually found in these patients include, papilloedema (the most significant finding),ocular motor nerve palses ,visual field defects which are seen in 96% of patients.[2,3,9,10] Enlargement of the blind spot is frequent and is found in virtually all patients with papilloedema.[4,9,10] Visual acuity is usually normal in early papilleodema but may deteriorate as the disease progresses. Our patient met most of this criteria apart from the CSF findings. She had normal visual acuity in both eyes, mild ptosis of the Lt upper eyelid suggestive of a partial 3rd nerve palsy, blurring of the disc margins in both eyes(fig 1) and a few retinal peripapillary haemorrhages. Her visual fields showed slight enlargement of the blind spot with initial peripheral constriction superiorly in both eyes. CT scan of the brain revealed no abnormality in this patient, as is expected in the diagnosis of IIH. Other neuroimaging studies include magnetic resonance imaging with contrast,(the study of choice) and magnetic resonance venography.[3,6]

Although medical and surgical options are available in the management of this condition, it is generally considered to be a self limiting condition. The goal of treatment is to preserve visual function and relieve symptoms. Medical therapy includes weight loss, acetazolamide and serial lumbar punctures.[3,4,5,6] Surgical treatment is instituted when visual loss progresses inspite of medical treatment. Optic nerve decompression or a cerebrospinal fluid diversion procedure is carried out.[6] The index patient in this study did not receive any form of treatment but was observed and asked to return on a follow up visit. However she defaulted and returned 5 months later with resolution of all the presenting symptoms(fig 1). This lends credence to the belief that the disease is self limiting.

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