BILATERAL PAPILLOEDEMA ASSOCIATED WITH PSEUDOTUMOUR CEREBRI IN ENUGU: A Case Report and Literature Review

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SUMMARY
A 42-year old female presented with a chronic headache associated with occasional blurring of vision. She is a myope and her aided visual acuity was 6/12 in each eye. This improved to 6/6, following a review of her refraction. She was moderately obese with a body mass index of 31.63.

The main finding was papilloedema. There were no focal central nervous system deficits. An intracranial space occupying lesion (SOL) was suspected but a computerised tomography (CT) scan of her brain showed only very small ventricles. No intracranial lesion was seen.

A diagnosis of pseudotumour cerebri (PTC) was suggested. Symptoms cleared and the papilloedema resolved after a course of treatment with acetazolamide.

The aim of this case report is to rekindle awareness of PTC and maintain a high index of suspicion towards early diagnosis. Early diagnosis prevents blindness.

Key words: pseudotumour cerebri, headache, papilloedema, and blindness

INTRODUCTION

Pseudotumour cerebri (PTC) is synonymous with idiopathic intracranial hypertension, elevated intracranial pressure, and benign intracranial hypertension. It is a disorder of unknown aetiology, and affects predominantly obese women of childbearing age. The primary problem is chronically elevated intracranial pressure (ICP), and the most important neurological manifestation is papilloedema, which may lead to progressive optic atrophy and blindness.[1] It is not associated with any clinical, laboratory or radiological evidence of an intracranial space-occupying lesion, meningeal inflammation or venous outflow obstruction.[2]

Studies in the United States of America showed an annual incidence of 0.9 cases per 100,000 population with a female-to-male ratio of 8:1 for a mean weight 38% over the ideal weight for height.[3] Children and adults have been found to be affected between the age range of 12 – 61 years.[4]

Although the condition is rare, it is critical to recognize the warning signs and symptoms to prevent devastating ophthalmic complications.[5]

CASE REPORT

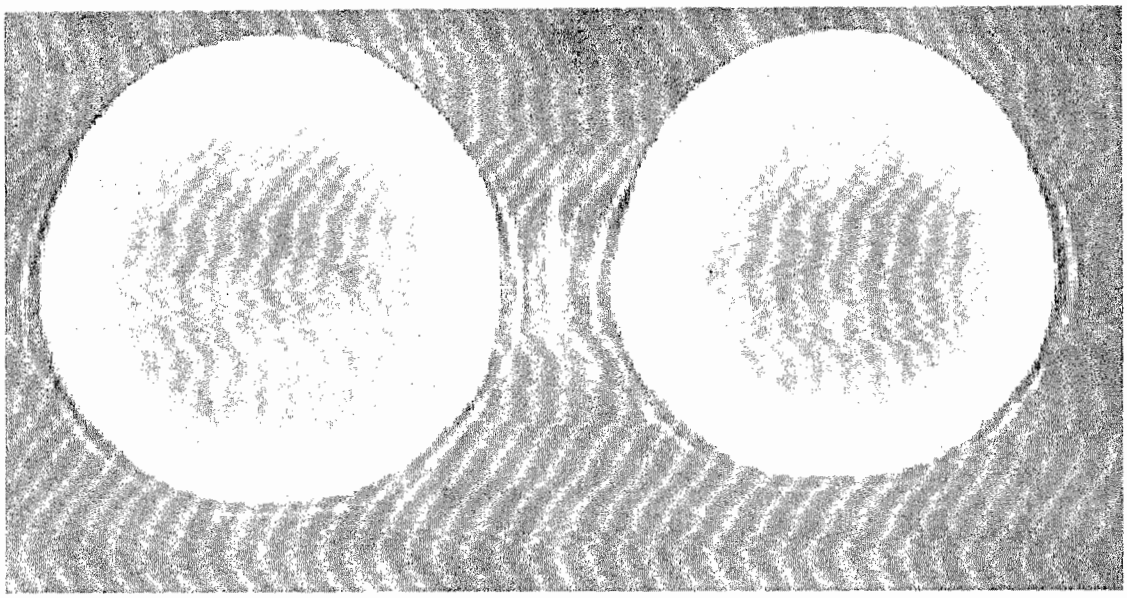
A 42-year old female was referred to the eye clinic, University of Nigeria Teaching Hospital (UNTH), Enugu for ophthalmological evaluation. She complained of intermittent headaches, associated with occasional blurring of vision over a period of one year. She is short-sighted and has worn glasses for over 25 years. Her general health was satisfactory.

On examination her visual acuity was 6/12 in each eye with glasses. This was improved after a refractive review to 6/6 in each eye. Apart from refractive error, the main finding was papilloedema. Her visual field by confrontation was normal and there were no localising central nervous system deficits. She was moderately over weight with a body mass index (BMI) of 31.63. (Her weight was 83kg, and height was 1.62m).

An intracranial space occupying lesion (SOL) was suspected. She was referred immediately to Memfys Hospital for Neurosurgery where she was thoroughly evaluated. The only abnormal finding was in the CT scan of her brain that showed very small ventricles. No intracranial SOL was seen.

The clinical and radiological picture strongly suggested pseudotumour cerebri. Figure 1 shows the very small lateral ventricles, the third ventricle was also very small but it did not show well in the photograph. The intracranial pressure was not measured.

Full blood count (FBC), electrolytes, urea and creatinine (EsU & Cr), and liver function test (LFT) were carried out. All the results were normal and the patient was started on acetazolamide tablets (500mg twice daily) and slow release potassium chloride (Slow-K) tablets (600mg twice daily).



Two weeks later the symptoms cleared. She was asked to continue her treatment for another two weeks when she developed severe paraesthesia and her treatment was stopped.

A repeat FBC, EsU & Cr showed normal results. She was followed up for one year and was found free of symptoms. She could not afford to pay for a repeat C T scan and she failed to keep further appointments.

DISCUSSION

Headache is the most frequent symptom in PTC, occurring in more than 90% of the patients.[6, 7] The headache presents in various patterns; in the case reported, headache was intermittent. Cluster-like headaches[8] and chronic daily migraine headaches[9] have been described in patients with PTC. The headache is almost always chronic and there are accompanying symptoms of increased intracranial pressure (ICP), such as pulsatile tinnitus, transient visual obscuration and radicular neck pain.[6] In this particular case, the headache occurred intermittently over a period of one year and it was associated with occasional blurring. The symptoms resolved after she had a course of treatment with acetazolamide.

Symptoms and signs are basically from prolonged raised ICP. The main symptoms are mentioned in the above. In almost all cases, the earliest sign is papilloedema, however, patients with PTC without papilloedema have been reported;[10] these patients had symptoms suggestive of PTC; and lumber puncture showed an elevated ICP. They were all obese women and their symptoms resolved after treatment with a diuretic and weight reduction. Green et al.[11] reported a case with clinical features suggestive of PTC with normal intracranial pressure. This suggests that some patients may be more susceptible to lower levels of ICP.

Where PTC is diagnosed, unusual ocular motility disturbances have been reported.[12] Bilateral sixth nerve and partial right third nerve palsies, alternating skew deviation and upbeat nystagmus have been associated with PTC. The ophthalmopareses may be related directly to very elevated cerebrospinal fluid pressure (CSF), and may be secondary to altered CSF flow in the posterior fossa. The ocular movement of the patient in this case report was normal.

Acquired choroidal folds causing defective vision from induced hyperopia[13, 14] and visual loss from peripapillary and macular choroidal neovascular membrane[15, 16, 17] have been found to be the result of prolonged papilloedema in PTC.

Pseudotumour cerebri might be benign and self limiting in a majority of cases, however the only permanent morbidity is vision loss from decompensation of papilloedema with progressive optic atrophy.

CONCLUSION

Pseudotumour cerebri is an important diagnostic consideration in the differential diagnosis for a patient with chronic headache, visual disturbances with or without papilloedema.

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