CASE REPORT

Pseudotumour cerebri - a case report from Enugu, Nigeria

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ABSTRACT
Pseudotumour cerebri is an often misdiagnosed cause of headache that may lead to loss of vision. It is a mimic of an intracranial space occupying lesion with the very same signs and symptoms except for the absence of any structural abnormality. A more appropriate designation, thus, would be idiopathic intracranial hypertension. Obese females of reproductive age are especially at risk. The case reported here posed challenges as a timely neurosurgical shunt placement was not offered due to unavailability of the implantable shunt device. Serial cerebrospinal fluid letting, which is a rather obsolete treatment modality, was then resorted to until the more appropriate lumbo-peritoneal shunt was available. Further, due to the patient's late presentation, she had a persisting visual defect affecting her job. We report this case more for the purpose of illustrating the management challenges in a resource poor setting.

***Keywords**: Challenges, hypertension, idiopathic, intracranial*

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INTRODUCTION

Pseudotumour cerebri (PTC) is a neurological disorder that is characterized by increased intracranial pressure in the absence of a tumour or other identifiable diseases. Historically, the first report was by the German physician Heinrich Quincke, who described it in 1893 under the name "serous meningitis".[1] The term "pseudotumour cerebri" was introduced in 1904 by Max Nonne. The disease was renamed "benign intracranial hypertension" in 1955 and finally, revised in 1989 to "idiopathic intracranial hypertension".[2]

CASE REPORT

A 36-year old female Nigerian trader presented in January 2014 to the Neurology

Clinic of the University of Nigeria Teaching Hospital Ituku-Ozalla in Enugu, Nigeria with complaints of headache and progressive visual loss. The throbbing headache of 2 months' duration was insidious in onset, intermittent, worse in the mornings and exacerbated by reclining or bending. Over the first 2weeks the headache worsened and subsequently, stopped her from routine activities. By the 3rd week she noted that it was worsened, also, by physical exertion and caused her sleepless nights. No relief was obtained from over-the-counter medications. There was no antecedent history of fever.

She experienced vomiting which became progressively more projectile, with pulsatile tinnitus and blurring of vision more in the left eye than in the right. There was no diplopia. One month prior to presentation she lost vision in the left eye, and 3days prior to presentation her right eye vision deteriorated until she, finally, had painless loss of vision in both eyes. There was no history of diabetes mellitus, hypertension, migraine or glaucoma.

She had commenced oral contraceptive pills more than 1year earlier and regularly took a food supplement containing vitamin A 1200 micrograms daily (*RDA in females is 700mcg*).

Physical examination showed a morbidly obese young lady, preferring to sit up in bed due to painful distressing headache, with a blind stare. Her body mass index (BMI) was 39.3kg/m^2. There was no pallor, fever, digital clubbing or oedema; her pulse was 62 /min and blood pressure was 130/80mmHg. Fundoscopy of the left eye showed pale mildly oedematous disc with haemorrhages over the retinal background, while that of the right eye showed marked papilloedema and less retinal haemorrhages. The rest of the neurological and systemic examination was unremarkable.

A tentative diagnosis of idiopathic intracranial hypertension was made with a differential diagnosis of intracranial space occupying lesion.

INVESTIGATIONS

Results: *White blood cell total* of 8.7 x 10^9/L with differentials N - 70% L - 28% M - 2% E - 0% B - 0%, Hb 15.6g/dL; ESR 25mm/1st hr; Na 135mmol/L, K 4.6mmol/L, HCO^3 18mmol/L, Cl 104mmol/L, Urea 3.3 mmol/L, Cr 92 µmol/L; *Fasting blood glucose of* 78mg/dL; *Fasting lipid profile* - Cholesterol Total 4.8, HDL-C 0.9, LDL-C 2.4, VLDL 0.5, TG 1.2 (values in mmol/L); *Mantoux Test* 2mm; *HIV (1 &2) screening* Non-reactive; *Brain CT Scan was* essentially normal but for some effacement of the cortical sulci and gyri; *Brain MRI Scan* was in keeping with the earlier CT Scan; *CSF Studies showed c*lear specimen with WBC 3/mm^3, RBC Nil, AFB Negative.

She was admitted and commenced on intravenous 20% mannitol, intravenous dexamethasone and tablets prednisolone, acetazolamide, frusemide and ranitidine.

The neurosurgery and ophthalmology teams were invited to review because of the possible need for CSF shunt placement by the neurosurgeons, and assessment of visual impairment by perimetry testing by the ophthalmologists, as an objective tool to monitor recovery.

The patient had a lumbar puncture (LP) done and the CSF opening pressure was markedly raised as CSF shot across the bedside. By the second day post-LP the patient had a rapid recovery of vision in the right eye. The neurosurgeons assessed patient to have raised intracranial pressure (ICP), and to rule out an intracranial space occupying lesion, for which a brain MRI was requested.

Subsequent intra-admission CSF letting yielded improvements in visual acuity and a lessening of headaches, these were, however, short-lived as she later relapsed.
At this point the neurosurgeons concluded that she would benefit from a lumbo-peritoneal shunting but, a date for surgery could not be fixed as the shunt itself was out of stock in the hospital. The patient was then discharged home during the 5th week on medications, for review at the out-patient clinic within 1week, and possible continuation of serial CSF letting.

She presented one week later than her appointment, complaining of poor vision (hand movement only) in the right eye for 2/7 and severe headache, and so, she had a repeat therapeutic CSF letting. A few days later, she had a worsening of vision and at this point she was referred to another specialist hospital for expedited CSF shunting. She finally had a lumbo-peritoneal shunt placed with clinical improvement, and her condition remained stable, subsequently.

DISCUSSION

In Nigeria, we do not have any reliable figures for the incidence of PTC, unlike what is obtainable in the developed world. A 2014 report from Israel reported that the average annual incidence rate was 2.02 per 100 000 with an incidence of 3.17 per 100 000 for women and 0.85 per 100 000 for men.[3] The median age at diagnosis is 30years, but occurs predominantly in women. It can occur in children as in adults. Obesity strongly predisposes to PTC and was documented in 83.4% of patients in the above cited Israeli study.[3] One of the few reported cases of PTC in Nigeria made the diagnosis with the Dandy Criteria in a 14-year old girl having what was qualified as mild obesity.[4]

Cerebrospinal fluid is produced at an approximate rate of 500ml/day, and since the ventricles and subarachnoid space around the brain and spinal cord can contain only about 150ml, the rest of the CSF is drained primarily into the blood through arachnoid granulations along the superior sagittal sinus. Thus, the CSF turns over about 3.7 times in a day.[5]

The main symptoms of PTC are: headache, nausea, vomiting, pulsatile tinnitus, double vision and visual loss. And these have been identified as the reliable tool for the diagnosis of PTC such as set forth in the *Modified Dandy Criteria* (Digre and Corbett 2001), below:

1. Symptoms of raised intracranial pressure
2. No localizing signs with the exception of abducens (sixth) nerve palsy
3. The patient is awake and alert
4. Normal CT/MRI findings without evidence of thrombosis
5. LP opening pressure of >25 cmH_2O and normal biochemical and cytological composition of CSF
6. No other explanation for the raised intracranial pressure.[6]

Our index case had four of the signs and symptoms above.

CONCLUSION

A high index of suspicion remains vital in the diagnosis of PTC especially when neuroimaging is not available. We did not have a CSF manometer available but we were clinically convinced that the pressure of CSF on first spinal tap was way beyond the normal range.

The decision to perform serial CSF letting must be discussed well with the patient especially when definitive neurosurgical intervention is not immediately available for whatever reasons.

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