Case Report

A Case Report of ‘Pseudo Tumor Cerebri’

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ABSTRACT

Pseudo tumor cerebri is a clinical entity of uncertain etiology characterized by intracranial hypertension. The syndrome classically manifests with headaches and visual changes in women with obesity. This case is being presented for the following points: raised ICT, bilateral CN VI palsy, bilateral papilledema (left > right), neck rigidity present without Kernig’s and Brudginski signs. Keeping the history, Neurological findings, investigative results, point to the possibility of raised intracranial pressure with CN VI involvement. The final clinical diagnosis, to the above syndrome, points out to ‘Pseudo-tumor cerebri’.

Keywords: PTC (Pseudo tumor cerebri), IIH (Idiopathic intracranial hypertension), ICT (Intracranial tension), ICSOL (Intracranial space occupying lesion), CSF (cerebro spinal fluid), HTN (Hypertension), Hypertensive encephalopathy

INTRODUCTION

Pseudo tumor cerebri is characterized by elevated intracranial pressure and normal brain ventricle size. Other terms used to describe this condition are ‘Benign intracranial hypertension’ and ‘Idiopathic intracranial hypertension’ [1]. As this condition may lead to blindness the term ‘Benign intracranial hypertension’ has fallen out of use. It presents with features of increased intracranial pressure (e.g., headache, papilledema, vision loss), with normal cerebrospinal fluid composition, and no other cause of intracranial hypertension evident on neuroimaging or other evaluations [2].

CASE HISTORY

A 25-year old female presented with severe headache and vomiting since 3 days, with no history of giddiness, chest pain, cough, shortness of breath, asthma, ingestion of outside food, and loss of appetite. She had no known allergies and she is not a tobacco user or does not take alcohol or use illicit drugs.

Headache was worse on the left side compared to the right since 3 days, severe, intermittent throbbing type, more in the morning associated with diplopia and blurring of vision and deviation of eyes with normal vision closing the other eye. There is a history of HTN noted by a local doctor 3 months ago and patient was on Amlodipine 5 mg since then. No history of diabetes/exposure to TB/chronic respiratory problem was noted.

Based on the history of 3 days of increasing headache with visual disturbance and association of vomiting, we kept the probability of increase in intracranial pressure. The diagnosis may be suspected on the basis of the history and examination. To confirm the diagnosis, as well as excluding alternative causes, several investigations are required; more investigations may be performed if the history is not typical or the patient is more likely to have an alternative problem.
Hypertensive encephalopathy was a reasonable consideration because of the presence of HTN and treatment she was on. However, the absence of progressive lethargy, confusion, seizures makes this cause unlikely.

On examination, patient was moderately built, uncomfortable, pulse 150 bpm regular, BP 150/100 mm of Hg in right upper arm in supine position, Temp 100F, respiratory rate 18/min without accessory respiratory muscle activity.

Cardiovascular examination, abdominal examination did not reveal any significant finding. The respiratory system examination was normal. On CNS examination, neck rigidity present Kernig’s negative, Brudginiski negative, higher functions normal, bilateral CN VI palsy and papilledema noted more on the left compared to the right. Motor and sensory systems as well as superficial and deep reflexes were found to be normal.

Cranial nerve VI palsy, unilateral or bilateral, is a sign of increased intracranial pressure caused by compression of nerve against petrous portion of temporal bone in its course from pons to orbit suggests optic neuropathy of any cause.

On investigating, CBP Hb 10 gm, TC 8300/cm, P-76%, L-18%, M-2%, E-4%, ESR-15 mm/h, Plt-1,50,000, serum creatinine 0.7 mg/dl, blood urea 20 mg/dl. Dengue NS1 Antigen, Dengue IgG, IgM were negative. Rapid malaria test was negative. Widal was not significant. Urine shows 2–3 pus cells, 4–6 epithelial cells in hpf, albumin traces, Sugar nil, and BS and BP were nil. Chest X-ray PA view, ECG, CT-Brain, MRI-Brain were all normal. Guarded lumbar puncture was done and the CSF study was normal.

DISCUSSION

A clinical approach to the syndrome of intracranial HTN without localizing neurological signs except CN VI palsy could be useful to consider.

We kept the possible causes of raised intracranial pressure due to medications such as high-dose vitamin A derivatives (e.g., isotretinoin for acne), long-term tetracycline antibiotics (for a variety of skin conditions) and hormonal contraceptives in view of her productive age. There was no history or clinical finding supporting the above. There are numerous other diseases, mostly rare conditions, which may lead to intracranial hypertension. If there is an underlying cause, the condition is termed ‘secondary intracranial hypertension’. Common causes of secondary intracranial hypertension include obstructive sleep apnea (a sleep-related breathing disorder), systemic lupus erythematous (SLE), chronic kidney disease and Behçet’s disease. Again, we did not find any evidence in support of these.

This case is being presented for the following points:

1. Raised ICT
2. Bilateral CN VI palsy
3. Bilateral papilledema (left > right)
4. Neck rigidity present without Kernig’s and Brudginski signs.

Keeping the history, neurological findings, investigative results, point to the possibility of raised intracranial pressure with CN VI involvement.

The final clinical diagnosis, to the above syndrome, points out to ‘Pseudo-tumor cerebri’. Patient was initially put on Mannitol 75 ml t.i.d, and then changed to oral glycerol and acetazolamide.

The patient gradually showed improvement in symptoms and coming for follow-up.

Benign intracranial hypertension is a syndrome of raised intracranial pressure occurring in the absence of an intracranial mass lesion or enlargement of cerebral ventricles due to hydrocephalus[3]. The absence of a clear identifiable aetiology for a clinical syndrome characterised by elevated ICP exists in nearly 90% of cases, and this ambiguity inevitably has led to the replacement of the misnomer ‘benign’ intracranial hypertension with IIH in light of the incidence of vision loss resulting from this condition[4]. It is most common
in young obese females. It is a condition of raised I.C.T without ICSOL, ventricular dilatation and impaired consciousness\[^5\]. Headache is the most common presentation. The ventricular system is either normal or small and generally there are no focal neurological signs. The visual field shows enlargement of blind spot\[^6\]. About 30\% of patients complain of horizontal diplopia due to sixth nerve palsy, which may be bilateral. The cause is a false localizing sign of raised ICT\[^3\]. In IIH, there is no structural obstruction to the circulation of CSF. It is believed that the problem lies in defective re-absorption of CSF either at the level of the arachnoid granulations or along cranial and spinal nerve root sheaths. There is no evidence for excessive CSF production. Early CT studies enabled clinicians to evaluate intracranial masses before lumbar puncture\[^7\]. The advent of MR imaging advanced the imaging paradigm in PTC from simply using imaging to rule out other processes (e.g., space-occupying lesions) to detecting signs thought to indicate PTC itself\[^8\]. The mainstay of therapy is weight loss with sodium reduction: acetazolamide (e.g., Diamox 250 mg p.o., QID initially, building up to 500 mg QID, or up to 4 g a day if tolerated), and discontinuation of causative medication\[^9\].

Two surgical options are cerebral decompression by CSF shunting and optic nerve sheath fenestration. Both the procedures have high degree of complications\[^10\].
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CONCLUSION

PTC is a benign self-limiting condition. It is characterized by increased intracranial pressure despite no obvious cause. There will be no focal neurological signs. C.T brain may show normal or small ventricles. It has a favourable outcome, although few cases may progress to permanent visual loss.

REFERENCES