Pseudotumor Cerebri As A Manifestation Of Addison's Disease

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Citation

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Abstract

We report about a 14 year old boy who presented with Pseudotumor cerebri (PTC) and was subsequently diagnosed as suffering from Primary adrenal insufficiency. The patient had no evidence of chronic adrenal failure except for hyperpigmentation of the skin. The diagnosis of PTC was made on clinical features of raised ICT, normal CSF composition and normal imaging of the brain (MRI) (1). Addison's disease was suspected as the cause of pseudotumor cerebri (PTC), which was subsequently confirmed by investigations. Replacement therapy led to complete recovery.

INTRODUCTION

Pseudotumor cerebri (Syn: idiopathic Intracranial hypertension, Benign Intracranial Hypertension) is an uncommon clinical disorder which classically presents with headache, papilledema without focal neurological signs (rarely with false localizing features), raised CSF pressure (more than 250 mm Hg) but normal CSF composition and normal imaging of brain.([2],[3]) In case of marked elevation of intracranial pressure, in the absence of mass one should distinguish pseudotumor cerebri (PTC) from occlusion of dural venous sinuses, gliomatosis cerebri, occult arteriovenous malformation, CNS infections and granulomatous meningitis ([1]).

This condition can be truly idiopathic and commonly witnessed in overweight females or can have other associations. Usual and common associations are excessive doses of tetracycline, vitamin A, administration of phenothiazine, estrogen, amiodarone and , quinolones, There are isolated instances when this condition has been described in association with hyperadrenalism, hypoadrenalism, myxoedema and hypoparathyroidism. Adrenal insufficiency is a rare association of PTC where features of raised intracranial pressure usually manifests during down titration of dose of steroid in patients of Cushing's disease who have been treated with bilateral adrenalectomy or in patients of Addison's disease.([4],[6],[7],[8]). However, PTC as presenting manifestation of Addison's disease is extremely rare and the review of literature revealed only one such case ([5]).

We present a patient who presented with classical features of pseudotumor cerebri (PTC). He was subsequently diagnosed as suffering from primary adrenal insufficiency, and responded promptly to steroid replacement.

CASE REPORT

A 14 year old male patient presented with episodic severe bursting global headache and vomiting for last one month. The frequency of headache was 2-3 per day and subsided with vomiting. Seven days before presentation the symptoms worsened and headache was almost continuous with mild blurring of vision. Three days prior to presentation patient had one episode of generalized tonic-clonic seizures (GTCS), following which he became drowsy and was referred to our hospital. There was no fever, diplopia, cough, diarrhea, focal neurological deficit, decrease urinary output, abdominal pain, alteration in bladder bowel habit, swelling of the body, seizure prior to illness, trauma, psychiatric complaints, history of incriminating drug intake, joint pain, surgery, history of stroke in past or history of chronic illness.

Examination revealed a conscious, but drowsy patient with stable vitals - Pulse 90/min regular and BP of 100/60 mmHg without postural fall. There was hyper pigmentation of body prominently over distal part of extremities and mucus membrane of oral cavity. Cardiovascular, chest and abdominal examination were unremarkable. The nervous system examination revealed bilateral extensor planter without any focal neurological deficit. Fundus examination revealed mild papilledema. .

At presentation the patient had hyponatremia (serum sodium 117 mmol/L), serum potassium 4.8 mmol/L, random blood sugar 70 mg/dl and other serum biochemistry were within normal limit. On imaging, MRI brain was normal there was no evidence of hydrocephalus or thrombosis of cerebral venous system. On lumbar puncture CSF opening pressure was high (240 mm of water) but CSF examination was within normal limits. Serum cortisol and ACTH (between 8.00-9.00 AM) were 2.9 g/dl and 554 pg/dl respectively. CT scan abdomen revealed normal appearing adrenal glands. The patient was treated with 150 mg hydrocortisone intravenously over 24 hours for 2 days with remarkable improvement. Subsequently, the dose of steroid was tapered and on day four he was switched to oral predinisolone 10mg twice daily. An attempt of decrease the dose further led to recurrence of the symptoms. The patient was discharged on 10 mg prednisolone twice daily.

DISCUSSION

In the present case PTC was diagnosed due to typical clinical presentation along with raised CSF pressure in presence of normal imaging of brain (MRI) and unremarkable CSF study. The clinical features of chronic adrenal insufficiency are non specific which includes fatigue, generalized weakness, anorexia, nausea, vomiting, weight loss, general languor and debility, diarrhea, ill defined abdominal pain, acute abdomen, personality changes- excessive irritability and restlessness. Hypotension (BP in range of 80/50 or less) with or without postural fall and progressive cutaneous and mucosal hyper pigmentation, along with associated hyponatremia and hyperkalemia suggest the diagnosis, which is confirmed by demonstration of low serum cortisol between 8-9 AM associated with high ACTH level. In the present case also adrenal insufficiency was suspected because of presence of hyper pigmentation of skin and oral mucosa with a low serum sodium and relatively high serum potassium which was subsequently proved by estimation of the cortisol and ACTH levels. The prompt response to steroid replacement further proves the causal relation between the two. However, while titrating the replacement dose of steroid the tapering should be done very gradually because rapid reduction in dose may lead to recurrence of the symptoms.

There are very few case reports and isolated instances when PTC has been described in association with hypoadrenalism. In the present case PTC was the presenting feature of Addison's disease, and unless there is a strong suspicion there may be undue delay in the diagnosis as it happened in the present case.

LEARNING POINTS

- Pseudotumor cerebri can be a presenting manifestation of Addison's disease
- Common causes of PTC should be excluded before labelling this syndrome truely idiopathic
- Patient presenting with Pseudotumor Cerebri with hyponatremia and relatively normal or high potassium level should be investigated for adrenocortical insufficiency.

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