CASE REPORT

Pituitary Apoplexy in the Emergency Room: A Prior History of Pituitary Adenoma May not be Necessarily Present!

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Abstract

Pituitary apoplexy is a rare but life-threatening medical emergency that results from either a sudden hemorrhage or infarction in a pituitary tumor. Unrecognized pituitary apoplexy can lead to serious hormonal and neuro-ophthalmic complications and even death. A major challenge in the diagnosis of pituitary apoplexy is that majority of such patients don't have a prior diagnosis of pituitary adenoma. Emergency magnetic resonance imaging of the pituitary gland is the diagnostic tool of choice for this condition. This may not be readily available in many emergency rooms. A previously healthy patient who presented with sudden severe headache and confusion and was found to have pituitary apoplexy is described and a focused review of the condition is given with emphasis on early recognition and complications of this condition

Key words: Pituitary apoplexy, Headache, Hypoadrenalism, Emergency,

Introduction

Pituitary apoplexy is a rare but life-threatening medical emergency that results from either a sudden hemorrhage or infarction in a pituitary tumor. Prompt diagnosis and early therapeutic interventions are of paramount importance to avoid the serious hormonal and neurogenic complications of this condition. One of the challenges facing physicians, particularly those in the frontline (emergency room), when dealing with such patients is the absence of a prior diagnosis of pituitary adenoma (1). Furthermore, pituitary apoplexy can mimic many neurologic and non-neurologic disorders. Hence, high index of suspicion, prompt recognition and initiation of appropriate therapy offer the best opportunity for a good outcome in this medical emergency. In the absence of consensus on the management of pituitary apoplexy and in order to increase awareness of this serious medical emergency among health practitioners, recent UK-based practice guidelines have been developed (1). In this

Table 1. Results of the baseline hormonal investigation on presentation and the reference ranges

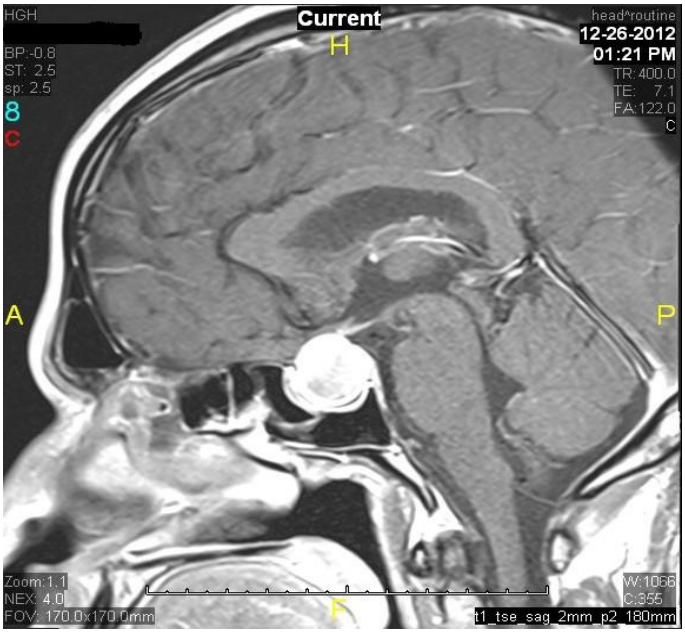
Circulating hormone levels (units)	Patients results	Reference Range
Serum prolactin (mIU/l)	72	73-407
Serum lutenizing hormone (LH) (IU/L)	< 0.5	1-9
Serum follicular stimulating hormone (FSH) (IU/L)	< 2	4-9
Serum testosterone (nmol/l)	<0.10	10.4-35
Serum free thyroxine (FT4) (pmol/l)	7.6	9-20
Serum thyroid stimulating hormone (TSH) (MIU/L)	0.64	0.45-4.5
Serum growth hormone (GH) (ug/L)	<0.01	0-10
Serum somatomedin C (IGF1) (ng/ml)	95	97-228
Serum adrenocorticotrophic hormone (ACTH) (pg/L)*	2	5-60
Serum cortisol (nmol/l)*	16	138-580

* *morning values*

paper, a previously healthy patient who presented with sudden severe headache and confusion and was found to have pituitary apoplexy is described and a focused review of the condition is given with emphasis on early recognition and complications of this condition.

Case report

A 36 year-old man presented to the emergency room at Hamad General Hospital with one-day history of confusion and 10-day headache. There was no history of associated fever, photophobia or limb weakness. He was previously healthy and had no preceding upper respiratory tract infection. He is married with one child and worked as a supervisor in a store. He never smoked or drank alcohol. Physical examination revealed a confused and agitated young man with a normal volume pulse of 100 beats per minute, blood pressure of 110/60 mmHg and a temperature of 36.8° Celsius. Fundoscopic examination revealed bilateral papilledema without ophthalmoplegia. Examination of visual field by confrontation test was not possible at presentation as the patient was confused and agitated. However, repeat examination after improvement in his mental state was unremarkable. Examination of other cranial nerves was normal. There were no signs of meningeal irritation and motor system examination was normal apart from exaggerated deep tendon reflexes. Rest of systemic examination was unremarkable. Blood chemistry showed sodium of 117 mmol/l (normal 135-145), potassium 3.7 mmol/l (3.5-5.2), corrected calcium 2.10 mmol/l (2.2-2.6), albumin 45 g/l (35-50), glucose 4.85 mmol/l, chloride 80 mmol/l (96-106), creatinine 58 umol/l (61-124) and bicarbonate 24 mmol/l (22-28). Liver function tests, complete blood count and differential cell count were all within normal range. Emergency non-contrast computerized tomography (CT scan) of the brain was non-conclusive. Magnetic resonance imaging (MRI) of the brain revealed marked enlargement of the pituitary gland most likely due to adenoma which appeared extremely bright on T1 sequence and hyperintense on T2 suggesting pituitary apoplexy (hemorrhagic type). There was also compression of the residual pituitary tissue forming a smooth strip around the tumor with mild compression of the optic chiasm (Figures 1 and 2). Laboratory results revealed low serum levels of almost all pituitary hormones (Table 1). In addition, serum procalcitonin was normal and blood and urine cultures did not grow any organism. The patient was managed initially with intravenous dexamethasone 4 mg every 6 hours that was replaced by intravenous hydrocortisone 100 mg intravenously every 6 hours along with intravenous fluid infusion with a good recovery in his general and mental status and improvement of serum electrolytes. He was discharged home in a good condition on oral hydrocortisone 30 mg daily and L-thyroxine 50 ug daily for further follow up with endocrinology and ophthalmology. A full report was given to the patient upon discharge.



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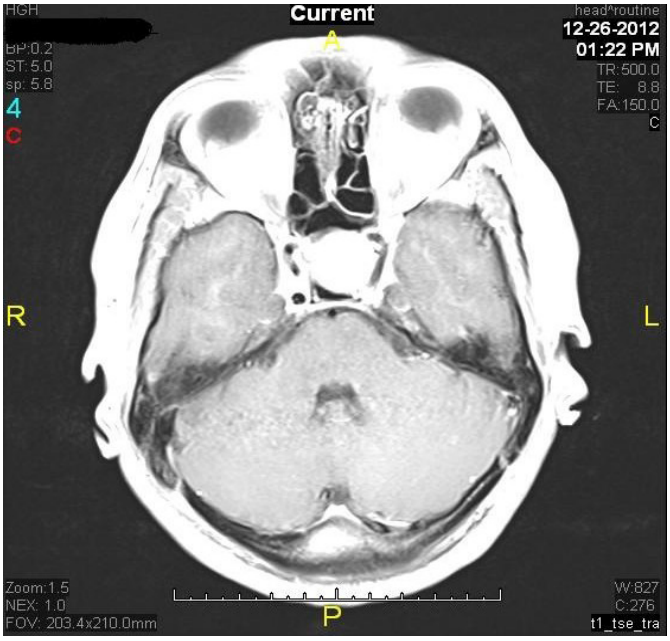

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Figures 1 and 2: Pituitary MRI scans showing markedly enlarged pituitary gland harboring an adenoma with hyperintense signal suggestive of hemorrhagic type of apoplexy**.**

Discussion

Pituitary apoplexy is a rare but life-threatening medical emergency that results from either a sudden hemorrhage or infarction in a pituitary tumor. The incidence of pituitary apoplexy varies from 1.9% to 6.8% (2-3). Recent UK guidelines for the management of pituitary apoplexy published in 2011 (1) emphasize that a diagnosis of pituitary apoplexy should be considered in all patients presenting with acute severe headache with or without neuro-ophthalmic signs. Among pituitary tumors, non-functioning pituitary macroadenomas are the most commonly involved in apoplexy. Only a minority of patients with pituitary apoplexy will have obvious precipitating factors such as coronary artery surgery (or other major surgery), pregnancy, gamma knife irradiation, endocrine stimulation tests, anticoagulant therapy or coagulopathy secondary to liver failure (4,5). The exact pathogenesis of pituitary apoplexy is not well understood, however, the sudden expansion in the sellar contents thought to compress surrounding structures and portal vessels in the hypophyseal stalk, resulting in sudden, severe headache, visual disturbances, and impairment in pituitary function (6). The majority of patients who present with pituitary apoplexy didn't receive a diagnosis of pituitary adenoma prior to their presentation suggesting that pituitary apoplexy may be the initial manifestation of undiagnosed pituitary tumors (1). The most common presenting symptom of pituitary apoplexy is headache which probably results from stretching and irritation of the dura mater in the walls of the sella supplied by the trigeminal-nerve meningeal branches (7). Nausea and vomiting are also frequent symptoms. The most fearful complications of pituitary apoplexy are the ophthalmic and hormonal ones. Compression of the optic chiasm and optic nerve may lead to decreased visual acuity, bitemporal hemianopia and sometimes complete blindness. Compression of the adjacent cavernous sinus and its nerve content (located on both sides of the sella) may result in ophthalmoplegia (particularly the third cranial nerve because of its outer location) (8). Deficiency of the anterior pituitary hormones is a frequent finding and may lead to catastrophic outcome. ACTH deficiency is the most commonly encountered and the most serious hormonal abnormality in pituitary apoplexy which may result in adrenal crisis and hypotension. Other hormones like TSH, LH, FSH, GH could also be affected. Hyponatremia may result from adrenocorical insufficiency or inappropriate ADH secretion from the posterior pituitary gland. Mild elevation of serum prolactin is the norm when hypophyseal stalk is compressed by enlarging pituitary adenoma (loss of inhibitory effect of hypothalamus on prolactin secreting

cells of the pituitary gland). Low serum prolactin level in patients with apoplexy or adenomas suggests severe destruction of the gland. Such patients are the least likely to recover from hypopituitarism after decompressive surgery (6,7,9). Pituitary MRI is the diagnostic tool of choice for pituitary apoplexy. In addition of being very accurate in diagnosing apoplexy, MRI can also detect the time when hemorrhage happens. In the first two days, the hemorrhage appears hyperintense on T1 and hypointense on T2 weighted MRI imaging. After 2 days, it appears hyperintense on both imaging modalities. It has also been found that the features of infarction, hemorrhagic infarction or hemorrhage on MRI correlate well with the histopathological reports and operative findings. In other words, histopathological features can be relatively accurately predicted from the MRI images. CT scan is less accurate in this regard as it can't differentiate cystic or degenerative changes from previous hemorrhage (1,10). Pituitary apoplexy is a medical emergency due to the risk of adrenal crisis and neuro-ophthamic complications and initial management should include fluid resuscitation, replacement of corticosteroid with intravenous hydrocortisone and support of the hemodynamic status of the patient. There is no solid evidence with regard to the choice of conservative versus surgical decompression. The 2011 UK guidelines advised that patients with pituitary apoplexy should first be stabilized medically with steroid replacement if needed and that the decision to manage conservatively or with surgical intervention should be made carefully by a multidisciplinary team, including experts in neurosurgery, endocrinology and ophthalmology. The guidelines advised that patients with severely reduced visual acuity, severe, persistent or deteriorating visual field defects, deteriorating level of consciousness or with ocular paresis should be considered for surgery (1). In our patient, pituitary apoplexy was the initial manifestation of a large pituitary tumor. The patient after recovery denied any symptoms prior to this illness. Such presentation is the commonest for pituitary apoplexy. His hormonal assay reveled deficiency of all anterior pituitary hormones including prolactin that suggests severe destruction of the gland.

In conclusion, it is strongly suggested that physicians, particularly those working in acute care settings should develop high index of suspicion for diagnosing pituitary apoplexy in any patient with new onset headache, ophthalmic or neurologic symptoms. Prompt treatment of pituitary apoplexy-related complications (particularly hormonal and neuro-ophthlamic) can prevent catastrophic outcomes.

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