Acromegaly with Pituitary Tumor Apoplexy: A Case Report

Yu-Hsin Cheng¹, Yi Liu¹, Ann-Shung Lieu²

Of functioning pituitary macroadenomas with apoplexy, prolactinoma is predominant. Acromegaly due to pituitary macroadenoma associated with apoplexy is a rare phenomenon. Herein, we report a case. The 19-year-old male was admitted with a chief complaint of headache with nausea and vomiting after jogging. His gross appearance was with typical feature of acromegaly. Brain computed tomography (CT) and magnetic resonance imaging (MRI) showed an intrasellar tumor with hemorrhage. Transsphenoidal removal of pituitary tumor was done and intratumoral hematoma was found during operation. Microscopic histology showed pituitary adenoma. Two months after surgery, his serum levels of growth hormone and insulin-like growth factor-1 were normal. The clinical manifestation of this patient must be differentiated from a ruptured aneurysm, which can be identified by brain CT scan in early approach.

Key words: acromegaly, apoplexy, pituitary adenoma

Introduction

Pituitary tumor with apoplexy is an uncommon complication, usually caused by hemorrhage or infarction within a pre-existing pituitary adenoma. Pituitary apoplexy occurs in 0.6-10.5% of all cases of pituitary adenoma. Most cases of apoplexy occur in nonfunctioning pituitary macroadenomas or prolactinomas. Apoplexy in pituitary macroadenoma with acromegaly is rare. Herein, we present a case of growth hormone-secreting pituitary adenoma with hemorrhage, whose clinical presentations resemble those of spontaneous subarachnoid hemorrhage from ruptured aneurysm.

Case Report

This 19-year-old male was well-being before and had no history of any systemic disease. He visited our emergency room with a chief complaint of blurred vision of left eye. One day ago, he suffered from sudden onset of headache with nausea and vomiting after jogging. On physical examination, a wide nasal bridge, thick lips and ears, coarse facial contour, and a larger size of hands, fingers and feet were noted. Acromegaly was suspected. On neurological examination, the patient was alert and fully oriented. Extraocular muscle movements were intact but bitemporal visual field defects were present. Computed tomography (CT) scan of the brain without enhancement showed an intrasellar tumor with hemorrhage (Fig. 1). Non contrast T1-weighted magnetic resonance imaging (MRI) revealed a heterogeneous mass in the sellar with focal high intensity area, suggesting hemorrhage (Fig. 2). The patient’s growth hormone...
Fig. 1 Noncontrast CT scans including axial view (A) coronary view (B) and sagittal view (C) showed intrasellar tumor with intratumoral hemorrhage (arrow)

Fig. 2 Noncontrast MRI scans (A) T1 weighted sagittal section showed intrasellar mass with heterogeneity (B) T1 weighted coronary section also showed intrasellar mass with heterogeneity. T2 weighted axial (C) and coronary (D) views revealed intratumoral hypointensity suggesting acute hemorrhage (arrow)
(GH) level was 297.8 μIU/ml (normal range <5 μIU/ml), and his insulin-like growth factor-1 (IGF-1) level was 869 ng/ml (Normal range: 140-366 ng/ml, in this age). The serum level of cortisol was 1.38 μg/dl (normal range: 5-25 μg). Serum levels of prolactin, thyroid stimulating hormone, and electrolytes were within the normal range. Cortisone supplements were administered first. Transsphenoidal removal of pituitary tumor was performed two days later. During operation, intratumoral hematoma was found. Microscopic examination revealed an acidophilic adenoma with area of necrosis. Serum levels of GH and IGF-1 checked two months after surgery were 1.04 μIU/ml and 170.9 ng/ml respectively. The patient continues to be followed up at neurosurgical outpatient clinic.

**Discussion**

Pituitary apoplexy is a rare complication of pituitary tumor. It is characterized by abrupt onset of headache accompanied by nausea or vomiting, visual deterioration, ophthalmoplegia, oculomotor paresis, and partial or complete pituitary function insufficiency with or without altered consciousness\(^{(2,3,6)}\). Most cases of pituitary apoplexy occur in nonfunctioning pituitary macroadenomas\(^{(2,3)}\). However, of the functioning pituitary macroadenomas with apoplexy, prolactinomas are predominant\(^{(3,5)}\). Acromegaly associated with pituitary apoplexy is rare, with the reported incidence about 3.5-4%\(^{(4,7)}\).

The pathophysiological mechanism of pituitary apoplexy is not clear. It often occurs in macroadenoma, but the exact cause for this phenomenon is still unknown. In physiological condition, pituitary gland receives most of blood supply through the hypophyseal portal system which is a capillary network. In addition, it also receives direct arterial blood supply in a lesser extend. However, the arterial supply is more dominant than portal system in pituitary adenomas. Therefore, the result of pressure difference from the special vascularity of the sellar contents may contribute to the apoplexy\(^{(1,8,9)}\). Furthermore, the vessels of pituitary adenomas are found to have incomplete maturation, poor fenestration, and often with ruptured and fragmented basal membranes. Thesees reported structural abnormalities may contribute to the susceptibility of pituitary apoplexy\(^{(1,10)}\).

Several factors precipitating apoplexy have been reported, such as head trauma, changes in arterial pressure or cerebrospinal fluid pressure, radiotherapy, anticoagulant therapy, dynamic test of pituitary function, and treatment with dopamine agonists\(^{(1,10-12)}\). However, these factors are not present in our patient.

Clinical presentations of patients with aneurysmal subarachnoid hemorrhage closely resemble those with pituitary apoplexy. Both diseases present with sudden, severe headache that can be followed by altered consciousness level, visual deterioration and possible meningismus. In our patient, the possibility ruptured anterior communicating artery aneurysm cannot be ruled out. CT scan is helpful in demonstrating intrasellar tumor and hemorrhage, while CT angiography may be necessary to demonstrate aneurysmal bleeding.

During the initial treatment of pituitary apoplexy, close monitoring of fluid and electrolyte balance, and correction of pituitary hormones deficiencies are extremely important because an unawareness of any of these conditions may put the patients at risk during and after surgery. The timing of surgery remains a debate. Currently, it is widely adopted that patients with pituitary apoplexy without severe visual loss or consciousness change which is not related to electrolyte or pituitary hormone insufficiency may be managed expectantly\(^{(2,3,10)}\).

In conclusion, in spite of its rarity, pituitary
apoplexy should be considered in patients with an acute onset of headache and progressive visual disturbance. Such condition should be differentiated from an aneurysmal subarachnoid hemorrhage, and brain CT scan is helpful to differentiate these two disease entities in initial approach.

References

肢端肥大症併腦垂體瘤中風：病例報告

鄭玉欣¹  刘  忡¹  劉安祥²

在功能性腦垂體腺瘤合併中風的患者中，以泌乳激素瘤較為常見，肢端肥大症併腦垂體瘤中風的患者較為少見，故在此提出一病例報告。一位十九歲男性患者，其入院主訴為慢跑後發生劇烈頭痛並有嘔吐眩暈之現象，患者外型表現典型肢端肥大症之特徵，腦部電腦斷層及核磁共振檢查顯示蝶鞍內腫瘤併出血，以經蝶竇方式將腫瘤切除，在術中發現腫瘤內有血腫的情形，病理報告為腦垂體腺瘤，術後二個月追蹤血清中生長激素及類胰島素生長因子皆恢復至正常。此患者在臨床上以劇烈頭痛表現，須與顱內動脈瘤破裂導致的出血做一鑑別診斷，腦部電腦斷層可做為一初步及快速排除動脈瘤破裂之檢查工具。

關鍵詞：肢端肥大症，腦中風，腦垂體腺瘤