Spontaneous Remission of Acromegaly Due to Apoplexy
Akromegalinin Apopleksiyi Bağlı Spontan Remisyonu

Cevdet Duran, Canan Ersoy*, Sinem Kıyıcı*, Naile Bolca**, Erdinç ErTürk*, Ercan Tuncel*, Sazi İmamoğlu*

Konya Education and Research Hospital, Division of Endocrinology, Konya, Turkey
*Uludag University, Medical Faculty, Division of Endocrinology, Bursa, Turkey
**Uludag University, Medical Faculty, Department of Radiology, Bursa, Turkey

Abstract
Pituitary apoplexy is a rare clinical syndrome characterized by sudden onset of headache and vomiting, accompanied by visual disturbance or ocular motility impairment and some degree of pituitary insufficiency. Pituitary apoplexy presenting with these symptoms is seen in approximately 3% of patients with surgically treated pituitary adenomas. In acromegalic patients, apoplexy can be related to some provoking factors and rarely may be spontaneous. We present the case of an acromegalic patient with spontaneous remission after apoplexy. A 39-year-old male patient had transsphenoidal microsurgery for acromegaly in 1994 but remission was not achieved. After a 10-year period without follow-up, he experienced severe headache, nausea, vomiting, photophobia, and visual disturbance in July 2005 and scheduled an appointment with an ophthalmologist. His complaints ceased spontaneously within weeks. In October 2005, sellar MRI examination revealed a 3x2.5 cm mass of adenomatous tissue with necrosis. On admission to our clinic in November 2005, visual acuity, visual fields, and ocular motility, were normal; but results of laboratory tests revealed panhypopituitarism. He was thought to have apoplexy of a growth hormone secreting tumor, and hormone replacement therapy was initiated for panhypopituitarism. Because his symptoms had regressed, surgery was not considered. On rare occasions, acromegaly spontaneously remits after apoplexy, resulting in improved control of the functional adenoma without surgical intervention. Turk Jem 2008; 12: 80-2

Key words: Acromegaly, apoplexy

Özet


Anahtar kelimeler: Akromegali, apopleksi

Introduction
Pituitary apoplexy is a rare clinical syndrome characterized by sudden onset of headache and vomiting, accompanied by visual disturbance or ocular motility impairment and some degree of pituitary insufficiency [1]. The incidence of pituitary apoplexy presenting with classical symptoms is approximately 3% in the series of surgically treated pituitary adenomas [2]. Apoplexy is encountered in macro-or microadenomas and thought to develop along with hemorrhage or ischemic infarction. Rarely, however, it occurs with other adenomatous or nonadenomatous pituitary lesions and an intact hypophysis [3]. Apoplexy rarely results
in spontaneous remission of symptoms in individuals with adenomas secreting hormones, including growth hormone (GH). In the literature, apoplexy in acromegalic patients is often reported to be associated with precipitating factors (4-10). Spontaneous remission, however, is rarely reported (11-13). In this report, an acromegalic patient with spontaneous remission related to apoplexy is reported and discussed in the context of available literature.

Case Report

A 39-year-old male patient was suspected to have acromegaly due to his appearance in 1994. Blood testing revealed a GH of 34 ng/mL. Computed tomography (CT) showed a 2x2 cm mass and some destruction of the sella turcica (Fig. 1). The patient underwent transsphenoidal hypophysectomy in the same year, and the adenoma was surgically removed. The levels of GH were elevated (14.5 ng/mL and 50 ng/mL) in the following year; an oral glucose loading test showed no suppression of GH. A CT scan was performed, and bone destruction and postoperative changes were detected in 1995 (Fig. 2). Treatment with bromocriptine was been started, but the patient took it for only one month. There was no follow-up for 10 years because of patient noncompliance.

In July 2005, he experienced severe headache, vomiting, nausea, photophobia, and visual disturbance, especially in his right eye; he scheduled an evaluation with an ophthalmologist, and a sellar magnetic resonance imaging (MRI) was planned by the ophthalmologist. MRI screening could be performed in October 2005. His complaints spontaneously improved between July and October 2005, but impotence had developed. In the sellar MRI evaluation, a mass 3x2.5 cm with necrosis inside and a rim shaped enhancement around the mass by contrast agent was seen (Fig. 3). On his admission to our clinic, his physical examination revealed acral enlargement, frontal bossing, and prognathism, consistent with acromegaly. The laboratory findings were as follows: glucose level 68 mg/dL (Normal Range [NR]: 60-100), sodium 145 mEq/L (NR: 136-152), potassium 4.3 mEq/L (NR: 3.2-5.2), thyroid-stimulating hormone 0.249 mIU/mL (NR: 0.35-4.9), free T3 1.54 pg/mL (NR: 1.71-3.71), free T4 0.65 ng/dL (NR: 0.7-1.48), follicle-stimulating hormone 2.64 mIU/mL (NR: 1.37-13.58), luteinizing hormone 0.94 mIU/mL (NR: 1.26-10), total testosterone 58.5 ng/dL (NR: 270-1734), GH 0.78 ng/mL (NR: 0.06-5), insulin-like growth factor-1 (IGF-1) 96 ng/mL (NR: 109-284), prolactin 0.66 ng/mL (NR: 2.58-18.12), adrenocorticotropic hormone (ACTH) 17.9 pg/mL (NR: 0.00-46), and cortisol 6.9 μg/dL (NR: 6-19). Panhypopituitarism due to apoplexy was suspected. Rapid ACTH stimulation test was performed, and the cortisol level was 22 μg/dL in the first hour. An insulin hypoglycemia test was then performed; maximal GH and cortisol responses were found to be 0.87 ng/mL and 5.45 μg/dL, respectively, although his blood glucose level decreased to 80 mg/dL. As the basal hormonal profiles revealed typical secondary hypothyroidism and hypogonadism, no TRH and GnRH stimulation tests were performed. A diagnosis of panhypopituitarism was made; steroid, levothyroxine, and androgen replacement treatments were applied.

Discussion

Classical pituitary apoplexy is a term describing an acute clinical syndrome, characterized by sudden onset of headache, vomiting, and visual and ocular disturbance; the incidence of pituitary apoplexy presenting with classical symptoms is approximately 3% in surgically treated pituitary adenomas (1,2). Asymptomatic pituitary hemorrhage, termed subclinical apoplexy, is more common, and this situation may occur with only isolated hormone deficiency in some patients. The pathogenesis of pituitary apoplexy remains unknown; pathology findings may reveal infarct and/or hemorrhage. In the literature, hypotheses for hemorrhage and ischemic infarction include ischemia caused by rapid growth of an adenoma or compression of the pituitary stalk compromising blood flow and causing blood vessel fragility.
Remission of acromegaly following apoplexy has rarely been reported. It has been reported that apoplexy may occur after head trauma (4), gastric bleeding (5), pituitary testing or treatment with hypothalamic hormones or analogues (6) administration of contrast agents (7), thyroidecmy (8), discontinuation of subcutaneous octreotide (9), and cerebral angiography (10) in acromegalic patients. Some cases, however, have no apparent precipitating factors (11-13). The literature on the features of acromegalic patients with apoplexy was reviewed in Table 1. In our case, spontaneous apoplexy due to ischemic necrosis was diagnosed; no precipitating factors were determined.

After apoplexy, hypopituitarism and various endocrinopathies may be observed (9). It is generally accepted that the initial management of pituitary apoplexy consists of careful monitoring of fluid and electrolyte balance, coupled with immediate replacement of deficient hormones (1). Unless glucocorticoid, levothyroxine, and intravenous electrolyte balance, coupled with immediate replacement of deficient hormones (1). Unless glucocorticoid, levothyroxine, and intravenous fluids are replaced properly, apoplexy is often fatal. In our case, the patient’s survival was highly interesting in the absence of treatment.

The indication and timing of surgery for patients with pituitary apoplexy remains controversial. Many authors have advocated surgical decompression in patients with prominent visual disturbance or neurologic deficits. Surgery may also improve pituitary function (14) as well as visual and neurologic deficits (2,13). Other studies, however, report many cases showing spontaneous recovery (14) after surgical decompression for pituitary tumor (15). In our case, no surgical intervention was considered, as the complaints resolved spontaneously; examination revealed normal movement of the ocular muscles and normal visual fields. The remainder of the neurological examination was also normal.

In conclusion, acromegaly may rarely and spontaneously show remission after apoplexy which may lead to decreased symptoms from adenoma, even without surgery. This may result in ophthalmological and neurological improvement, although panhypopituitarism persists and requires treatment.

### References