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Complete Surgical Resolution of Bilateral Total Opthalmoplegia without Visual Field Defect in an Acromegalic Patient Presented with Pituitary Apoplexy

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Abstract. Pituitary apoplexy (PA), which is one of the most serious life-threatening complications of pituitary adenoma, is characterized by abrupt onset of headache, nausea, vomiting, visual disturbances and oculomotor paresis. Combination of oculomotor cranial nerve paralysis with normal visual fields is very rare in PA. We report a 60-year-old acromegalic man presented with panhypopituitarism and bilateral total opthalmoplegia without a visual field defect. At initial evaluation his clinical findings were compatible with adrenal crisis and eye examination revealed total opthalmoplegia, bilateral ptosis and normal vision. MRI showed a large heterogeneous mass in the pituitary fossa. Although clinical findings due to adrenal crisis improved after glucocorticoid therapy there was no improvement in opthalmoplegia and ptosis. The patient underwent transsphenoidal excision of the pituitary mass. Histological examination revealed an adenoma with large areas of hemorrhagic infarction and most of the cells were positive for GH in immunohistochemical analysis. Although opthalmoplegia was severe at presentation, total recovery was achieved 3 months after transsphenoidal surgery. Therefore the presented case clearly demonstrates that opthalmoplegia without a visual field defect due to PA has a good prognosis and early diagnosis and treatment including surgical decompression are crucially important.

Key words: Pituitary apoplexy, Acromegaly, Opthalmoplegia, Hypopituitarism

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PITUITARY apoplexy (PA), which is one of the most serious life-threatening complications of pituitary adenoma, is characterized by abrupt onset of headache, nausea, vomiting, visual disturbances, oculomotor paresis, drowsiness, confusion and/or coma. PA is a rare event and its incidence in the large series of pituitary adenomas has been reported as 1.6% [1]. Recently, three clinical series including the largest groups in the literature from different centers have been published [1–3]. Among 77 patients analyzed in these 3 series, only 3 acromegalic patients with PA have been reported. When apoplexy arises in functioning adenomas, it may result in spontaneous remission of the symptoms caused by excess hormone. Spontaneous remission of acromegaly after PA has been previously reported in the literature as case reports [4–10].

Abrupt onset oculomotor nerve paralysis and decreased visual acuity and/or visual field defects may be the presenting features of PA [11]. However, oculomotor nerve paralysis without visual field defect is very rare in patients with PA and has been previously reported in a few cases as the presenting feature [11–14].

Here, we report a unique patient with acromegaly presented with panhypopituitarism and bilateral total opthalmoplegia without a visual field defect.

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Table 1. Basal hormone levels on admission and 3 months* after surgery

	On admission[1]	At 3rd month	Normal reference ranges
fT_3 (pg/ml)	1.18	2.44	2.2–4.7
fT_4 (pg/ml)	4.16	15.90	8–20
TSH (mIU/ml)	0.07	0.05	0.2–4.5
PRL (ng/ml)	0.32	0.54	2–18
Cortisol (μg/dl)	2.39	23.87	9–23
FSH (mIU/ml)	1.47	1.12	1.4–18
LH (mIU/ml)	0.32	0.36	1.5–9.3
IGF-I (ng/ml)	5.7	10.75	78–258 *for ages 50–70 yr*
total testosterone (ng/dl)	Undetectable	Undetectable	134–625

* The patient was on corticosteroid and thyroid hormone replacement.
[1] Three days after the onset of the symptoms

Case Report

The patient was a 60-year-old man who was admitted to the hospital because of suddenly developed headache, bilateral ptosis, malaise, nausea and vomiting. His headache was present for 3 days before the day of hospital admission. On the day of admission his headache increased and vomiting started, and during transport to the hospital bilateral ptosis abruptly onset. He had been diabetic for 5 years. On physical examination, he has most of the physical features of acromegaly; thickening of facial skin and typical acromegalic face, excessive sweating, increased hand, finger and foot size. Approximately 15 years ago, the patient was diagnosed as acromegaly and he was recommended surgical treatment for his pituitary macroadenoma. But the patient rejected any kind of treatment.

Based on his initial clinical findings including hypotension (80/50 mmHg), nausea and vomiting, adrenal failure was suspected. On neurological examination, the patient was alert and fully oriented. Eye examination revealed bilateral ptosis, total opthalmoplegia (no eye movements and non-reactive pupils), and normal vision/ visual fields. Except hyperglycemia (185 mg/dl), biochemical values including electrolytes were normal on admission (data not shown). On the day of admission (3 days after the onset) his initial hormone profile was compatible with panhypopituitarism (Table 1).

Pituitary magnetic resonance imaging (MRI) showed a large heterogeneous mass in the pituitary fossa with linear areas of intermediate and high intensity, suggesting hemorrhage. The mass extended into both cavernous sinuses and sphenoid sinus. But there was no compression on optic chiasm. The mass had a heterogeneous pattern of enhancement after gadolinium administration (Fig. 1a, b, c, d).

Based on the clinical, hormonal and radiological findings he was diagnosed as PA with panhypopituitarism and bilateral total opthalmoplegia. Glucorticoid replacement was started on the day of hospital admission (with initial dose of 50 mg intravenous prednisolone daily and tapered slowly to the dose of 7.5 mg/ day po) and complaints due to adrenal failure including nausea, vomiting and hypotension improved. However there was no improvement in opthalmoplegia and ptosis. Thyroxine therapy was also commenced. When his hormonal and clinical status became suitable for surgery (2 months after the hospital admission), he had transsphenoidal surgery for the pituitary mass. Histological examination revealed an adenoma with large areas of hemorrhagic infarction. On immunohistochemistry, most of the cells were positive for GH.

The patient was re-evaluated 3 months after the surgery (Table 1). He had been on glucocorticoid and levothyroxine replacement without any complaint. Ptosis and ocular movements were completely recovered 3 months after the surgery.

Discussion

Pituitary apoplexy, an acute massive infarction of the pituitary gland, is a rare but life-threatening disorder that requires prompt diagnosis and rapid medical intervention. This clinical syndrome which is characterized by acute onset of headache, nausea, vomiting may be accompanied by loss of vision or ocular move-

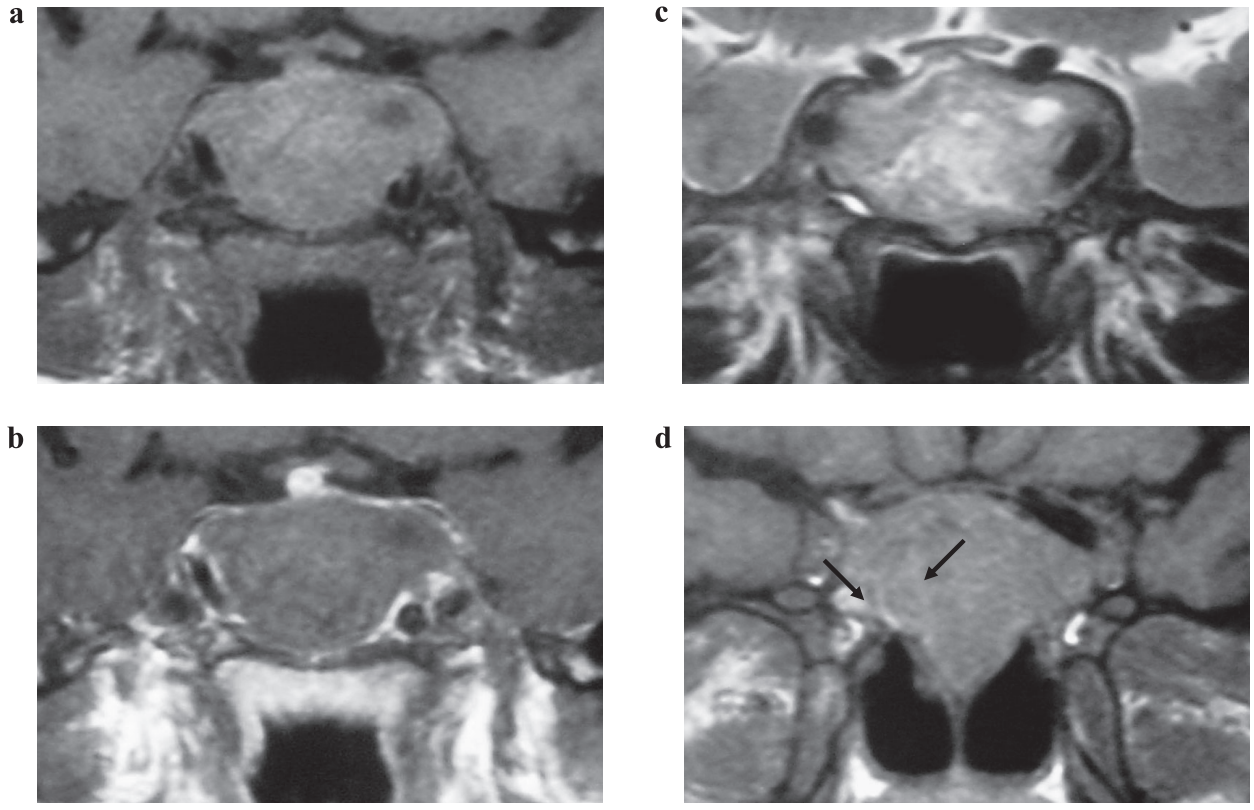


Fig. 1. Pituitary MRI images revealing pituitary apoplexy.
a: T1-weighted MRI image showed a large heterogeneous mass in the pituitary fossa.
b: The enhancement effect after gadolinium administration in the pituitary gland surrounding the mass and bilateral cavernous sinuses.
c: T-2 weighted MRI image showed a large heterogeneous mass in the pituitary fossa.
d: T1-weighted MRI images showed a large heterogeneous mass in the pituitary fossa with linear areas of intermediate and high intensity (pointed by arrows), suggesting hemorrhage.

ments and/or pituitary insufficiency [1, 2].

In addition to the visual and ocular problems, PA may lead to numerous endocrinopathies which can be transient or permanent. When apoplexy arises in functioning adenomas, it may result in spontaneous remission of symptoms and sometimes may lead to hypopituitarism. Similar to the present case, spontaneous remission of acromegaly after PA has been reported previously [1, 2]. Although the patient had previous diagnosis and typical clinical features of acromegaly, basal hormone levels clearly showed deficiency of GH besides other pituitary hormones (Table 1). The secondary adrenal failure recovered dramatically after glucocorticoid replacement. Acute adrenal crisis due to loss of adrenocorticotropic hormone is the most life-threatening endocrine disorder which is detected in two thirds of the patients with PA [15]. Thus, glucocoticoids should be given empirically in all cases of suspected PA.

The cranial nerves III, IV, V, and VI can be compressed due to PA. As a result, certain degree of ocular palsy is present in 40% of patients which may be unilateral or bilateral [11, 12]. Isolated opthalmoplegia without loss of vision as seen in this case is very rarely the presenting sign of a pituitary adenoma or PA. The main reason of the isolated opthalmoplegia was the growth pattern of the pituitary adenoma that the mass extended into both cavernous sinuses but there was no compression on optic chiasm. It has been previously demonstrated that most of the pituitary adenomas in acromegaly tend to have lateral or infrasellar invasion [16]. The possible cause of this invasion pattern could be explained by the somatotroph cells are located at the lateral wings of the pituitary gland and thereby GH se-

creting adenomas generally originate from the lateral wings of the adenohypophysis [17]. Opthalmoplegia may occur slowly as a result of mechanical compression or rapidly due to PA as in the present case. Cranial nerves III, IV, and VI may be entrapped in the cavernous sinus by an expanding hematoma, thus may cause various degrees of opthalmoplegia, ptosis, and pupillary defects [11, 12]. If diagnosed correctly and treated either medically or surgically, opthalmoplegia secondary to PA appears to be reversible. It has been reported that opthalmoplegia without a visual field defect has a better prognosis than oculomotor nerve palsy following visual loss [18]. In a study evaluating management and outcomes of 45 cases it has been demonstrated that patients who lack or exhibit mild and non-progressive neuro-ophthalmological signs, can be managed conservatively [3]. However in the present case, there was no improvement of opthalmoplegia after glucocoticoid therapy and the patient underwent a transsphenoidal excision of the pituitary mass. Three months after the surgery his eye movements and ptosis completely recovered.

In summary, we report a patient with acromegaly case presented with panhypopituitarism and bilateral total opthalmoplegia without a visual field defect. Although opthalmoplegia was severe at presentation, total recovery was achieved 3 months after the transsphenoidal surgery. Therefore the present case clearly demonstrates that opthalmoplegia without a visual field defect due to PA has a good prognosis when early diagnosis and appropriate treatment are provided.

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