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Case Report

Pituitary Apoplexy Presenting as Isolated Third Cranial Nerve Palsy with Ptosis : Two Case Reports

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Pituitary apoplexy is a clinical syndrome caused by an acute ischemic or hemorrhagic vascular accident involving a pituitary adenoma or an adjacent pituitary gland. Pituitary apoplexy may be associated with a variety of neurological and endocrinological signs and symptoms. However, isolated third cranial nerve palsy with ptosis as the presenting sign of pituitary apoplexy is very rare. We describe two cases of pituitary apoplexy presenting as sudden-onset unilateral ptosis and diplopia. In one case, brain magnetic resonance imaging (MRI) revealed a mass in the pituitary fossa with signs of hemorrhage, upward displacement of the optic chiasm, erosion of the sellar floor and invasion of the right cavernous sinus. In the other case, MRI showed a large area of insufficient enhancement in the anterior pituitary consistent with pituitary infarction or Sheehan's syndrome. We performed neurosurgical decompression via a transsphenoidal approach. Both patients showed an uneventful recovery. Both cases of isolated third cranial nerve palsy with ptosis completely resolved during the early postoperative period. We suggest that pituitary apoplexy should be included in the differential diagnosis of patients presenting with isolated third cranial nerve palsy with ptosis and that prompt neurosurgical decompression should be considered for the preservation of third cranial nerve function.

KEY WORDS : Third cranial nerve palsy · Ptosis · Pituitary apoplexy.

INTRODUCTION

Pituitary apoplexy is defined as a clinical syndrome that may include headache, visual deficits, ophthalmoplegia, or altered mental status due to an expanding mass within the sella turcica resulting from hemorrhage and/or necrosis[2,13] or infarction. This syndrome occurs most frequently in preexisting pituitary adenomas, but infarction may also occur in an apparently normal gland. Its presentation may be highly variable, acute and dramatic with rapidly developing headache, ophthalmoplegia, neurological deficits, coma, and death, or subacute with symptoms slowly evolving from days to weeks[11]. An expanding mass in the cavernous sinus can compress cranial nerves III, IV, V, and VI, thereby producing various degrees of cranial nerve palsy as the signs and symptoms[10]. However, isolated third cranial nerve palsy with ptosis as the presenting sign of pituitary apoplexy is very rare. We describe two cases of pituitary apoplexy presenting as sudden-onset isolated ptosis and diplopia.

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CASE REPORT

Case 1

A 73-year-old male presented with a five-day history of severe headaches and a three-day history of right-sided ptosis preceded by diplopia. There was no history of loss of consciousness, neck stiffness or features suggestive of seizure activity. There was a previous history of hypertension but not diabetes mellitus. Examination revealed partial ptosis of the right eye and anisocoria. In room light, the right pupil measured 4 mm, and showed no reaction to direct light. The left pupil measured 3 mm and reacted normally to light. The patient showed marked weakness when moving the eyeball on the right side. Visual acuity and visual field were unremarkable. The remaining cranial nerves were intact, and there was no evidence of sensory or motor weakness. A diagnosis of right third nerve palsy with ptosis was made. Subarachnoid hemorrhage due to a ruptured aneurysm of the posterior communicating artery was suspected. However, there was no evidence of intracerebral aneurysm or subarachnoid hemorrhage on brain computed tomographic angiography (CTA) (Fig. 1). Magnetic resonance imaging (MRI) of the head and sellar region showed a mass in the pituitary fossa with signs of hemorrhage,

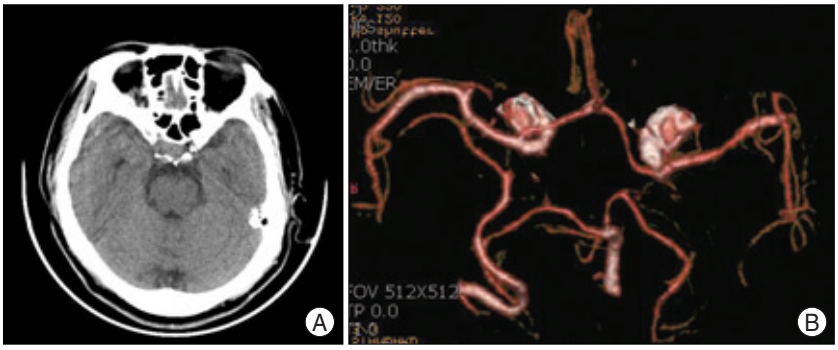

Fig. 1. A : Non-enhanced computed tomography (CT) demonstrating a noncystic homogeneous intrasellar mass without evidence of subarachnoid hemorrhage. B : CT angiography shows no evidence of intracerebral aneurysm.

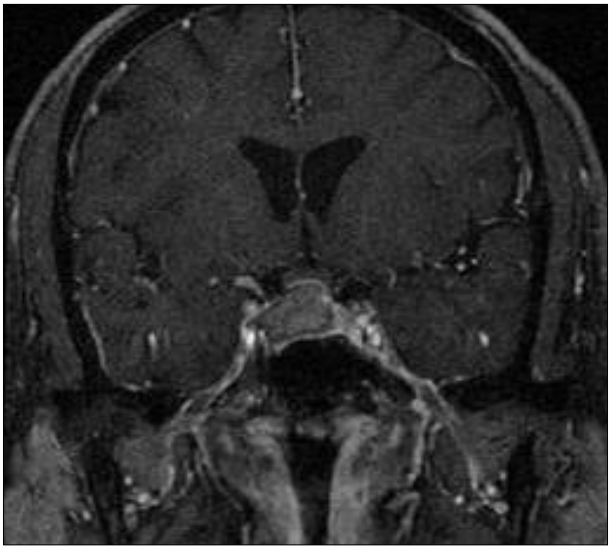

Fig. 2. Gadolinium-enhanced T1 weighted coronal magnetic resonance image of the brain showing a 1.9×1.3 cm-sized mass indicative of subacute hemorrhage in the pituitary fossa with extension toward the right cavernous sinus, upward displacement of the optic chiasm and erosion of the sellar floor.

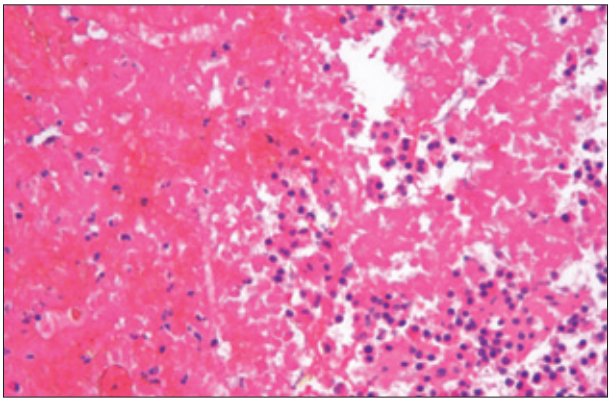

Fig. 3. Microscopic features of pituitary apoplexy. Photomicrograph shows tumor cells with coagulative necrosis and hemorrhage.

upward displacement of the optic chiasm, erosion of the sellar floor and invasion of the right cavernous sinus (Fig. 2). Under the presumptive diagnosis of pituitary apoplexy, neurosurgical decompression was performed via a transsphenoidal approach. Histology confirmed a pituitary adenoma with extensive areas of hemorrhage and necrosis consistent with apoplexy (Fig. 3). The patient was discharged on day 14, and he was completely recovered from his right third nerve palsy.

Case 2

A healthy 35-year-old woman presented to the neurosurgical unit following an episode of partial ptosis of the left eyelid and diplopia. She was five days postpartum after a normal vaginal delivery. She presented with a three-day history of diplopia and a two-day history of left-sided ptosis. Neuroophthalmic examination was notable for partial ptosis of the left eye without pupillary involvement and diplopia. There was limitation of adduction on the left side. Visual acuity and visual field were normal. There was no evidence of remaining cranial nerve palsies and sensory or motor weakness. The patient was diagnosed with left third nerve palsy with ptosis. Her vital signs and electrolytes, including sodium, were normal. Her pituitary hormone levels were normal. However, MRI showed a large area of insufficient enhancement in the anterior pituitary consistent with pituitary infarction (Fig. 4). There was no radiological evidence of intracerebral aneurysm or of invasion into the cavernous sinus. Based on these results, under the presumptive diagnosis of pituitary apoplexy caused by Sheehan's syndrome, neurosurgical decompression was performed via a transsphenoidal approach. Histological examination confirmed the diagnosis of pituitary apoplexy (Fig. 5). The patient showed an uneventful recovery, and the patient's unilateral third cranial nerve palsy with ptosis completely resolved during the early postoperative period.

DISCUSSION

Pituitary apoplexy is an acute ischemic or hemorrhagic vascular accident involving a pituitary adenoma or an adjacent pituitary gland. It is well recognized that pituitary adenomas are particularly prone to hemorrhage and necrosis. Several series have showed a 9 to 15% rate of hemorrhagic degeneration in these tumors[9]. The inherent fragility of tumor blood vessels[2] and atherosclerotic embolization[17] have been proposed as possible mechanisms for hemorrhage and infarction in pituitary adenomas. On the other hand, Sheehan's syndrome or pituitary gland necrosis, is a

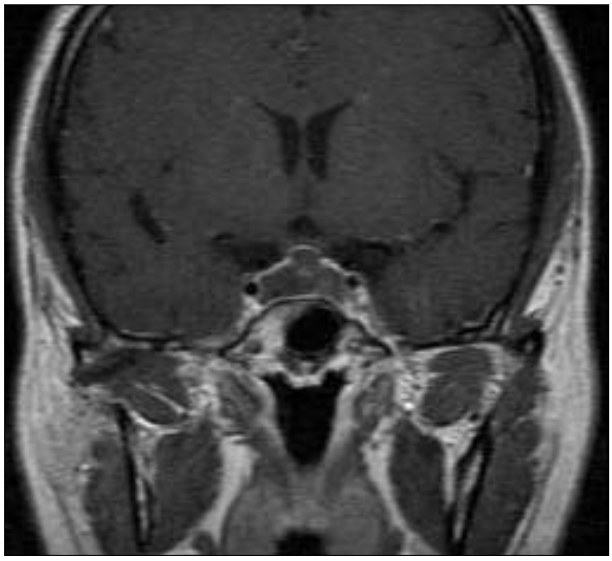

Fig. 4. Gadolinium-enhanced T1 weighted coronal magnetic resonance image showing a large area of insufficient enhancement in the anterior pituitary consistent with pituitary infarction, abutting the left cavernous sinus.

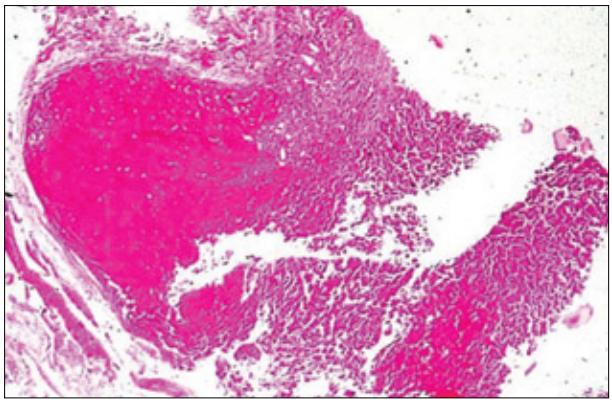

Fig. 5. In this case of Sheehan's syndrome, a photomicrograph shows an infarcted necrotic area on the left.

rare complication of postpartum hemorrhage that was initially described in 1937[16]. The pituitary gland is physiologically enlarged during pregnancy, and is therefore very sensitive to the decreased blood flow caused by massive hemorrhage and hypovolemic shock. The anterior pituitary is more susceptible to damage than the posterior pituitary. Sheehan's syndrome is rare, and it is extremely rare for ptosis to be the initial symptom of Sheehan's syndrome. These reports illustrate rare examples of a pituitary adenoma and Sheehan's syndrome with isolated third cranial nerve palsy.

The signs and symptoms of pituitary apoplexy extend along a continuum from mild to life-threatening. The most common complaint is the sudden onset of a severe headache, symptoms of meningeal irritation, such as neck pain, rigidity and photophobia, and acute visual field changes. Additional signs and symptoms include double vision (diplopia) or ophthalmoplegia caused by cranial nerve palsies, vomiting, and changes in the level of consciousness or loss of consciousness. Women with Sheehan's syndrome have varying degrees of hypopituitarism, ranging from panhypopituitarism to only selective pituitary deficiencies[3,6]. Isolated third cranial nerve palsy as the presenting sign of pituitary tumors and Sheehan's syndrome is very rare. Only a few cases of isolated third cranial nerve palsies in patients with pituitary tumors have been previously reported in the literature[4,5,7,14,15].

Previous reports have proposed several mechanisms to explain the occurrence of isolated third cranial nerve palsy in pituitary apoplexy. A large pituitary mass may compress the third cranial nerve within the lateral wall of the cavernous sinus. However, this tends to occur late in the course of tumor growth[8]. Direct invasion of the tumor through the sinus wall may also occur. Mechanical compression of the third cranial nerve against the unyielding interclinoid ligament of the cavernous sinus wall tends to bring about slow-onset nerve palsy. Sudden-onset third cranial nerve palsy has been attributed to the compromise of the vascular supply to the nerve due to compression of the vasa nervorum originating in the internal carotid artery[1,15]. These sudden symptoms have been observed in cases of pituitary apoplexy[14]. The third cranial nerve travels through the superior, lateral aspect of the cavernous sinus, at approximately the same horizontal level as the pituitary gland[12]. Due to its location, the third cranial nerve is relatively more susceptible to the laterally transmitted pressure generated by an expanding pituitary mass abutting the cavernous sinus[14]. In our cases, neuroradiological examination revealed a unilaterally expanding pituitary mass compressing the superior lateral wall of the cavernous sinus, thereby causing direct mechanical compression of the third cranial nerve or the vascular supply to the nerve, resulting in isolated third cranial nerve palsy.

Third cranial nerve palsy results from damage to the oculomotor nerve anywhere in its course from the nucleus in the dorsal mesencephalon, its fascicles in the brainstem parenchyma, the nerve root in subarachnoid space or in the cavernous sinus or posterior orbit. The most critical differential diagnoses include intracranial hemorrhage, carotid aneurysm rupture, subarachnoid hemorrhage, and bacterial meningitis. Other diagnoses to consider include cavernous sinus thrombosis and midbrain infarction[2]. The presenting sign of pituitary apoplexy with third cranial nerve palsy can mimic a number of other intracranial processes, and the diagnosis can be extremely difficult. For example, 33-56% of posterior communicating artery (PCA) aneurysms

present with ptosis. In these cases, the first suspected diagnosis was subarachnoid hemorrhage due to an aneurysm of the PCA, which can lead to further delays in definitive diagnosis. However, there was no evidence of intracerebral aneurysm or subarachnoid hemorrhage on brain CTA and MRI, and the diagnosis of pituitary apoplexy was suggested.

The treatment of patients with pituitary apoplexy includes both medical and surgical management. Intravenous steroid administration is mandatory until surgery in order to prevent acute adrenal insufficiency. Neurosurgical decompression via a transsphenoidal approach is the definitive treatment for pituitary apoplexy. Early decompression may partially or completely restore pituitary function and preserve third cranial nerve function. Early diagnosis and surgical decompression were important in the treatment of both of our patients, and both patients showed complete resolution of third cranial nerve palsy in the early postoperative period.

CONCLUSION

In conclusion, we suggest that pituitary apoplexy should be included in the differential diagnosis of patients presenting with isolated third cranial nerve palsy with ptosis and that prompt neurosurgical decompression should be considered in order to preserve third cranial nerve function in these patients.

References

1. Cahill M, Bannigan J, Eustace P : Anatomy of the extraneural blood supply to the intracranial oculomotor nerve. **Br J Ophthalmol 80** : 177-181, 1996
2. Cardoso ER, Peterson EW : Pituitary apoplexy : a review. **Neurosurgery 14** : 363-373, 1984
3. DiZerega G, Kletzky OA, Mishell DR Jr : Diagnosis of Sheehan's syndrome using a sequential pituitary stimulation test. **Am J Obstet Gynecol 132** : 348-353, 1978
4. Francois J, Neetens A : Oculomotor paralyses and tumors of the pituitary gland. **Confin Neurol 30** : 239-252, 1968
5. Kim JP, Park BJ, Kim SB, Lim YJ : Pituitary apoplexy due to pituitary adenoma infarction. **J Korean Neurosurg Soc 43** : 246-249, 2008
6. Lakhdar AA, McLaren EH, Davda NS, McKay EJ, Rubin PC : Pituitary failure from Sheehan's syndrome in the puerperium. Two case reports. **Br J Obstet Gynaecol 94** : 998-999, 1987
7. Lee CC, Cho AS, Carter WA : Emergency department presentation of pituitary apoplexy. **Am J Emerg Med 18** : 328-331, 2000
8. Lenthall RK, Jaspan T : A case of isolated third nerve palsy with pupil involvement. **Br J Radiol 73** : 569-570, 2000
9. Mohr G, Hardy J : Hemorrhage, necrosis, and apoplexy in pituitary adenomas. **Surg Neurol 18** : 181-189, 1982
10. Molitch ME : Gonadotroph-cell pituitary adenomas. **N Engl J Med 324** : 626-627, 1991
11. Onesti ST, Wisniewski T, Post KD : Clinical versus subclinical pituitary apoplexy : presentation, surgical management, and outcome in 21 patients. **Neurosurgery 26** : 980-986, 1990
12. Parkinson D : Surgical anatomy of the cavernous sinus. In : Wilkins RH, Rengachary SS (eds). **Neurosurgery**. New York : Mc Graw Hill, 1985, pp1478-1483
13. Reid RL, Quigley ME, Yen SS : Pituitary apoplexy. A review. **Arch Neurol 42** : 712-719, 1985
14. Rossitch E Jr, Carrazana EJ, Black PM : Isolated oculomotor nerve palsy following apoplexy of a pituitary adenoma. **J Neurosurg Sci 36** : 103-105, 1992
15. Saul RF, Hilliker JK : Third nerve palsy : the presenting sign of a pituitary adenoma in five patients and the only neurological sign in four patients. **J Clin Neuroophthalmol 5** : 185-193, 1985
16. Sheehan HL : Postpartum necrosis of the anterior pituitary. **J Path Bacteriol 45** : 189-214, 1937
17. Sussman EB, Porro RS : Pituitary apoplexy : the role of atheromatous emboli. **Stroke 5** : 318-323, 1974