Pituitary Apoplexy without Optic Disc Pallor : A Case Report

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รายงานผู้ป่วยโรคพิทูอิทารี่ อะโปเพล็กซี่โดยที่ขั้วประสาทตาไม่ซีด

ยศอนันต์ ยศไพบูลย์* พ.บ., จุฬาภรณ์ แสงวิจิตร* พ.บ., ไชยวิทย์ ธนะไพศาล** พ.บ.

**ภาควิชาจักษุวิทยา และ **ภาควิชาศัลยศาสตร์ คณะแพทยศาสตร์ มหาวิทยาลัยขอนแก่น*

โรคพิทูอิทารี่ อะโปเพล็กซี่ เกิดจากภาวะเลือดออกอย่างเฉียบพลันภายในเนื้องอกของต่อมพิทูอิทารี่ ทำให้ก้อนเนื้องอกโตขึ้นอย่างรวดเร็ว และไปกดทับอวัยวะข้างเคียง ถ้าก้อนเนื้องอกที่โตขึ้นไปกดทับเส้นประสาทตา จะทำให้ผู้ป่วยมีสายตามัวลงอย่างเฉียบพลัน และลานสายตาผิดปกติด้วย ผู้ป่วยโรคพิทูอิทารี่ อะโปเพล็กซี่นี้มักจะมีขั้วประสาทตาฝ่อและซีด ซึ่งเป็นผลจากการถูกกดทับ ผู้เขียนได้รายงานผู้ป่วย 1 ราย ที่เป็นโรคพิทูทารี่ อะโปเพล็กซี่ โดยที่ขั้วประสาทตาไม่ฝ่อซีด ได้วิจารณ์ถึงวิธีการวินิจฉัย ลักษณะการดำเนินของโรค และผลหลังผ่าตัดไว้อย่างละเอียด. *(ศรีนครินทร์เวชสาร 2529; 2 : 155 - 61)*

Pituitary apoplexy, characterized by acute infarction and hemorrhage within pituitary tumor, caused rapid expansion of the tumor and compressed on adjacent structures. Compression of the optic nerve and optic chiasm caused acute visual loss and visual field defects. These patients almost always had optic disc atrophy due to such compression. A 24-year-old patient of pituitary apoplexy without optic disc atrophy was reported. Diagnostic approaches, clinical course, and surgical outcome were discussed in details.

INTRODUCTION

Pituitary apoplexy, a rare but life threatening disorder, was first reported by Bleibtreu in 1905.[1] It was characterized by an acute massive infarction with secondary hemorrhage and edema within the tumorous pituitary gland.[2,3] This caused rapid expansion of the lesion and acute compression of adjacent structures, including the optic nerve, optic chiasm and the ocular motor nerves in the cavernous sinus.[1,4] Sometimes the patients may loss conscious, coma and succumb to death, due to acute distortion of the third ventricle by the expanding tumor.[5]

Compression of the optic nerve and chiasm caused acute visual loss and/or visual field abnormalities, which brought the patients to see ophthalmologist.[1,4-7] Most of these

patients usually had optic disc atrophy due to such compression, but some patients may occasionally present with normal optic disc, [4,8,9] even when there was large expanding tumor. We herein reported a patient with apoplexy in previous pituitary prolactinoma, whose optic discs were completely normal. Diagnostic approaches and surgical outcome in patients with apparently normal optic disc were discussed.

CASE REPORT

A 24-year-old woman complained of blurred vision in her both eyes for one week. She had history of amenorrhea for two years and was investigated for primary infertility at local provincial hospital. Hysterogram was performed 8 days ago to rule out the tubal causes of infertility, using about 50 ml. of Conray 420 as the contrast media. One day after the procedure, she developed sudden onset of bilateral deep orbital ache, radiating to the temple. At the same time, she also had progressive visual loss, which made her come to eye clinic at Srinagarind Hospital on November 20, 1985.

At first seen, her visual acuity was hand movement in the right eye and 6/60 in the left eye. Applanation tonometry revealed intraocular pressure of 17 mm.Hg in both eyes. Slit-lamp biomicroscopic findings of anterior segment were completely normal. The pupils were equal, and reacted to light well. The fundi were also normal (Fig 1). She was first diagnosed as retrobulbar neuritis of unknown cause. Prednisolone 60 mg. daily and antacid were given with no significant improvement.

Five days later, she complained that the vision in her both eyes became worse. The visual acuity ran down to only light perception in the right eye and 2/60 in the left eye. At this time, the fundi were still normal. The visual field by confrontation test demonstrated bitemporal hemianopia. Additional history taking also revealed galactorrhea. Amenorrhea - Galactorrhea Syndrome was then suspected. The skull X-ray was requested and showed a ballooned and destroyed sella turcica (Fig 2). Computerized tomographic scan revealed a large well encapsulated mass, 40 mm. in size, in the pituitary fossa (Fig 3). The patient was definitely diagnosed as pituitary apoplexy and planned for surgical decompression of the optic chiasm.

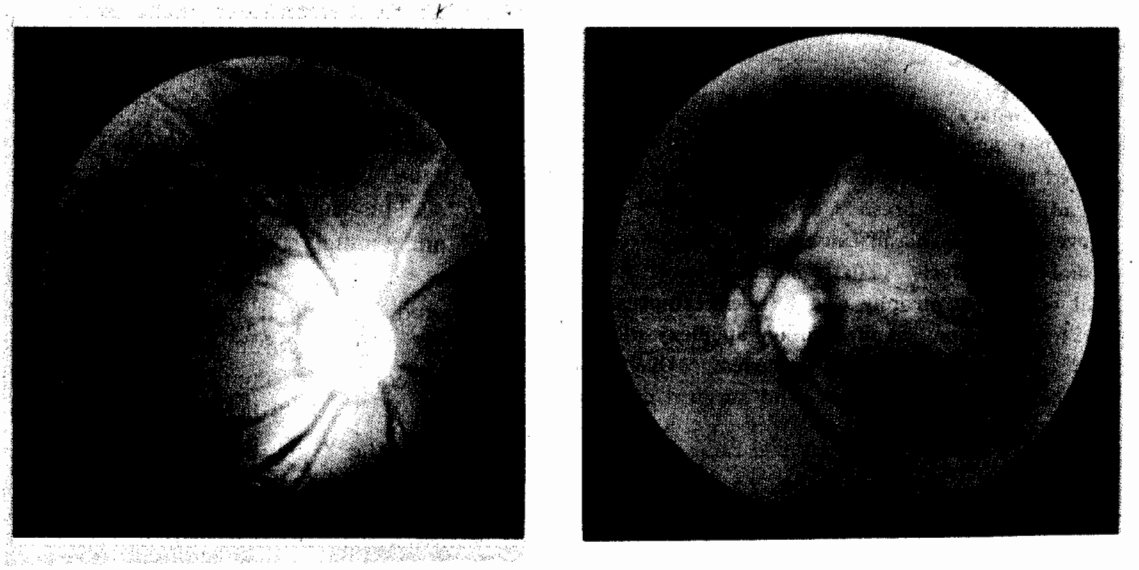

Fig 1 Fundus photograph of the patient revealed normal looking fundus in her both eyes.

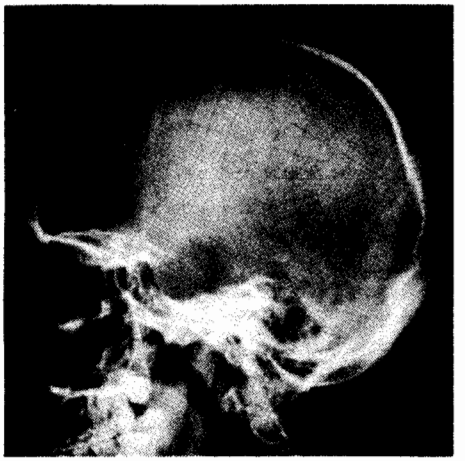

Fig 2 Skull film of the patient showed a ballooned and destroyed sella turcica.

Table 1 Pre-operative hormonal level study in the patient

Determination	(normal values)	Results
T_4	(4.5-12.5 μg/dl.)	1.8
T_3	(94-169 ng/dl.)	120
TSH	(0-3.5 μiu/ml.)	3.8
Prolactin	(0-20 ng/ml.)	> 200
Free T_4	(0.6-1.7 ng/dl.)	0.5

During the course of the next 4 days in hospital, some improvement in the visual function occurred. She was able to count finger at 1/2 ft. in her right nasal field, and had gained vision of 1/60 in her left eye. Visual field testing at this time was demonstrated in Fig 4. During this time, the hormonal level studies were performed for baseline pre-operative information. (Table 1)

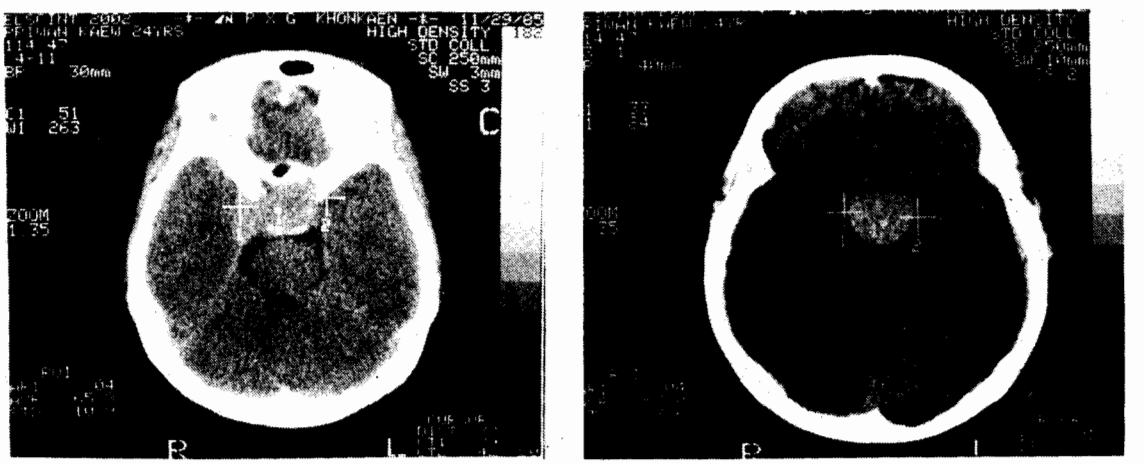

Fig 3 Computerized tomographic scan of the patient demonstrated a large well encapsulated tumor mass.

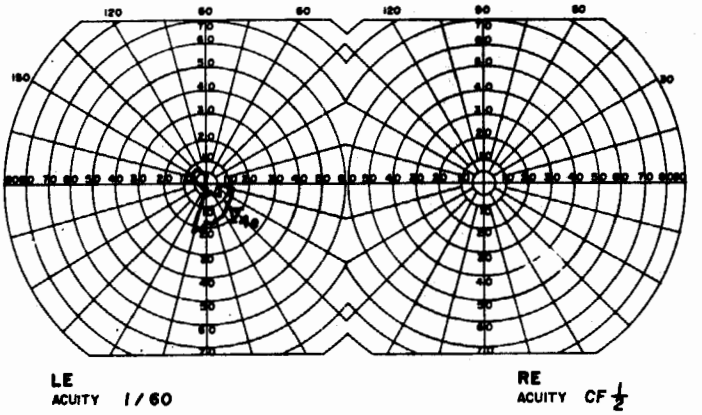

Fig 4 Visual fields of the patient examined before surgery. The right eye can not be tested because of poor vision.

One week after hospitalization, subfrontal hypophysectomy was carried out to remove the pituitary tumor. Intra-operatively, large chocolate soft tissue mass, about 30 mm. in size, was demonstrated in the pituitary fossa, pushing the optic chiasm posteriorly. Old hemolysed blood 8 ml. was aspirated from inside the mass, and residual yellowish tumorous tissue was surgically removed. The light and electron microscopy confirmed the clinical diagnosis of pituitary apoplexy in prolactinoma.

Post-operative course of the patient was uneventful. She had no serious complication except mild ecchymosis and numbness on the right-sided forehead. Bromocriptine (250 mg.) three times daily was started 5 days after surgery, to inhibit the synthesis and release of the prolactin.[10] At this time, she was able to see 6/6 with her right eye and 6/24 with here feft eye. Visual field examination revealed some improvement of the hemianopic field (Fig 5). She was discharged 10 days after the operation, and followed up weekly.

One month later, visual function tests were repeated and demonstrated visual acuity of 6/6 in the right eye and 6/12 in the left eye. Visual field test at the same time showed nearly full recovery of the peripheral field. (Fig 6)

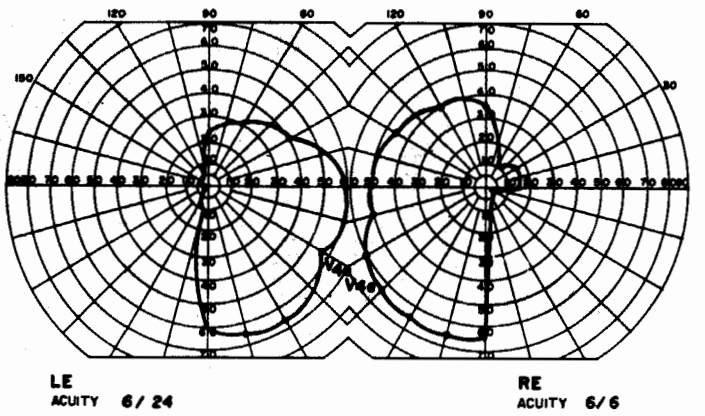

Fig 5 Visual fields of the patient examined after surgery, revealed some improvement of the hemianopic fields.

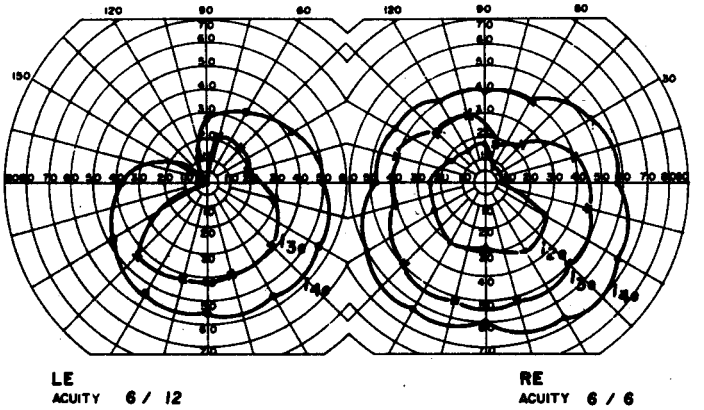

Fig 6 Visual fields of the patient examined one month after surgery, revealed nearly full recovery of the peripheral visual fields.

DISCUSSION

Most pituitary tumors grow so slowly that the signs and symptoms are unnoticed by the patients,[4,11] but some tumors grow rapidly because of apoplectic episodes or spontaneous hemorrhage within the tumors.[1-3] This causes rapid expansion of the tumor and compression on its adjacent structures. Although apoplectic episode is also found in non-tumorous pituitary gland, it is commonly associated with pre-existing pituitary tumor.[1,12] Chromophobe adenoma seems to be the most common tumor cell type found in cases of pituitary apoplexy, because of its rather higer frequency among all types of pituitary tumors.[13]

The clinical manifestation in pituitary apoplexy, including loss of pituitary function, neurological signs and symptoms, are determined by pattern of tumor growth and degree of hemorrhage within the tumor.[1,4] If expansion of the tumor, caused by infarction, edema and hemorrhage, rapidly compresses on the adjacent structures, the compromise of neurological function usually occurs. Upward expansion causes visual field defects and impaired visual acuity,[1,14,15] due to compression on the optic chiasm, as in this patient. Expansion within the sella frequently results in compression of the cavernous sinus which forms the lateral boundary of the pituitary fossa.[16] The cranial nerves III, IV, V, VI, branches of the sympathetic chain and the carotid artery, which course through the cavernous sinus are probably affected.[1,4,16] However, these cranial nerves involvement are not present in this patient.

As reported by Reid,[1] the first symptom in this patient is the sudden onset of retro-orbital ache, radiating to the temple. This symptom is thought to result from meningeal irritation or dural stretch on the lateral wall of the sella.[1]

During the pre-operative course in the hospital, some improvement in the visual function occurred in our patient. Cushing and Walker[4] had also noticed this spontaneous recovery, following by the failure of the vision later. They attributed this to be caused by change in the tension of the tumor growth due to edema or increased vascularity. In this report, visual improvement is thought to be caused by corticosteriod given during the treatment for suspicious retrobulbar neuritis. Steriod causes decrease in tissue edema and also inflammation, which make the

clinical symptom better. Later in the course, the lesion expands progressively and the visual function runs down again. However, spontaneous complete recovery of the symptom did occur,[17] re-ossification and marked reduction in size of the previously enlarged sella turcica were also reported, but in only a few instances.[17]

Concerning about treatment, pituitary apoplexy is considered a grave event, attended by high rate of mortality and morbidity.[1,5] Emergency surgical decompression of the optic chiasm is therefore recommended.[2,5,17] However, delay of a few days in this patient does not interfere with a satisfactory recovery of vision.[5] In fact, it gives time for surgeon to study the baseline hormonal level and to replace adequate corticosteriod. Subfrontral and transspheniodal approaches are two common choices of surgery.[5,17] Surgical outcome depends on pre-operative condition of the patients. Several factors are reported to be of prognostic value in predicting the visual outcome after surgery.[18-22] According to Lundstrom et al,[18] poor visual function without anatomical change of the optic disc was a favorable prognostic sign. Klauber et al[19] reported that visual improvement could be expected if pre-operative visual acuity better than 6/24, intact optic nerve and visual field defect less than two quadrants. Nelson [20] concluded from his study that tumor size and pre-operative prolactin level were important factors in predicting the surgical visual outcome. These do not concur with our patient because nearly full visual recovery occurred in this patient eventhough there were large tumor size and prolactin level more than 200 ng/ml. In this report, the condition of the optic disc seem to be the most important factor in predicting post-operative visual function. However, it is somewhat risky to predict on the basis of the disc appearance alone, because we have often seen return of vision in some patients to surprisingly good level in spite of far-advanced optic disc pallor.[12]

The most striking observation in our patient is that she had completely normal optic disc in her both eyes. Normally, compression on the optic nerve by the pituitary tumor often produces optic disc pallor of various degree.[11] The reason why the optic discs still appear normal may be attributable to time factor. Before the apoplectice episode, slow growing adenoma in this patient has no effect on the optic nerve. When sudden expansion caused by pituitary apoplexy occurs, it rapidly compresses on the optic chiasm and causes acute visual loss and visual field defect.[4-7] By Wallerian degeneration, the optic disc will become pallor in the next 2-3 weeks. This makes the disc appear normal at the time of initial examination. In fact, according to Wilson and Falconer,[6] pallor of the disc was recorded in 56% of the patients with pituitary tumors. Similarly, Chamlin et al[7] found optic disc atrophy in only 50% of his series. However, available data about optic disc pallor in pituitary apoplexy is not present. So absence of the optic disc pallor does not exclude the presence of pituitary tumor and its apoplectic episodes.[4] Careful visual field testing is therefore recommended in every patients who complain of visual loss for which no adequate causes can be detected, and it may reveal the visual field abnormalities caused by the compression. Plain skull film may also show enlarged sella turcica with or without bony destruction. However, these can not be regarded as an early sign, because they are secondary to compression and expansion. In fact, other symptoms such as headache and those of endocrine disturbances such as amenorrhea, galactorrhea and impotence may be present in the earliest stage of the disease.[10] And with the advent of neuroendocrine assay technique and development of fine matrix computerized tomography,[10,13] this lesion may be early diagnosed before there has been suprasellar extension to cause visual symptoms. Therefore, history taking about repro-

duction and sexual dysfunction, endocrine hormonal assay, history of sudden visual loss, careful visual field examination, plain skull X-ray and computerized tomography in combination may help to make early diagnosis in patients suspected of pituitary tumors and apoplexy, even though the optic discs are apparently normal.

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