Case Reports : Visual Field Defect in Primary Empty Sella Syndrome

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ผู้ป่วย empty sella syndrome นอกจากมีความผิดปกติเกี่ยวกับระบบฮอร์โมนแล้ว ยังอาจส่งผลกระทบต่อลานสายตาของผู้ป่วยด้วย ดังนั้นนอกจากจักษุแพทย์จะสามารถให้การรักษาปัญหาเกี่ยวกับลานสายตาของผู้ป่วยแล้ว ยังอาจช่วยนำไปสู่การวินิจฉัยผู้ป่วยที่มีปัญหา empty sella syndrome ได้ตั้งแต่ในระยะต้นได้อีกด้วย

Introduction

Empty sella occurs as a result of a deficient diaphragma sellae, with subarachnoid space protruding into the cavity of sella tursica. Pituitary gland is flattened and distorted against the sella floor and walls. The sella is usually enlarged, but this feature is not always present. The term "primary empty sella" is used when anomaly occurs in patients who never had surgery radiation on pituitary or Para pituitary tumor[1]. The diagnosis of empty sella is radiographically made when the sella tursica is seen to be enlarged or deformed that is partially or completely filled with cerebrospinal fluid[2].

Most patients with primary empty sella are asymptomatic and incidentally detected. The presence of empty sella was found in 5 percent of normal subjects on autopsy studies[3].

Typically primary empty sella syndrome occurs in obese, multiparous women, ranging in age from 27 to 72 years, with a mean age of 49 years old. Empty sella Aas been associated with hypertension, pseudotumor cerebri, hypopituitarism, spontaneous cerebrospinal fluid (CSF) rhinorrhoea, visual field defects, diminished visual acuity and headache[4].

Endocrine abnormalities are not commonly found in empty sella patient. However if such abnormalities are found, they are most commonly found in the form of Hyper-prolactinaemia. Growth hormone, thyrotropin and gonadotropin deficiencies are also noted. To establish the diagnosis of hypopituitarism (partial or complete) and to determine the hypophyseal cause of dysfunction, measurement of the following hormones are required in order to obtain serum thyrotropin, T4,

corticotrophin, cortisol, LH, testosterone, basal stimulated growth hormone and prolactin level[5].

The cause of visual field changes is due to either traction on the chiasm or involvement of chiasmal blood vessels, which result in ischemic damage to the optic nerve[2].

In this report we describe two patients with primary empty sella syndrome, who had visual field defected.

Case reports

Case 1

A 33-year-old man has chronic progressive visual loss in both eyes one year prior came to the hospital.

On eye examination, his visual acuity measured to 20/20 in the right eye and 20/20 in the left eye. Anterior segment was examined and found to be within normal limit in both eyes and no RAPD. The IOP measured 12 and 10 mmHg in the right and the left eye respectively. There were normal cupping with sharp disc margin, however they both looked pale. Visual field showed a tubular field defect in both eyes. (Fig 1) The Visual fields were thought to be highly suggestive of chronic papilledema that had progressed into atrophic stage. The patient was thought to have developed pseudotumor cerebri and this was confirmed by high initial pressure in lumbar puncture.

MRI of brain revealed an empty sella. Most of the sella was filled with CSF and traction of the optic nerves and chiasm. Based on these findings, a diagnosis of primary empty sella syndrome was made.

The endocrine studies showed hypothyroidism and adrenal insufficiency, the patient was then replaced with hormone therapy. Finally, the Patient was operated through trans-sphenoidal approach which has been traditionally known as "chiasmapexy". On during the 4 months follow up, there was still no improvement on his visual fields.

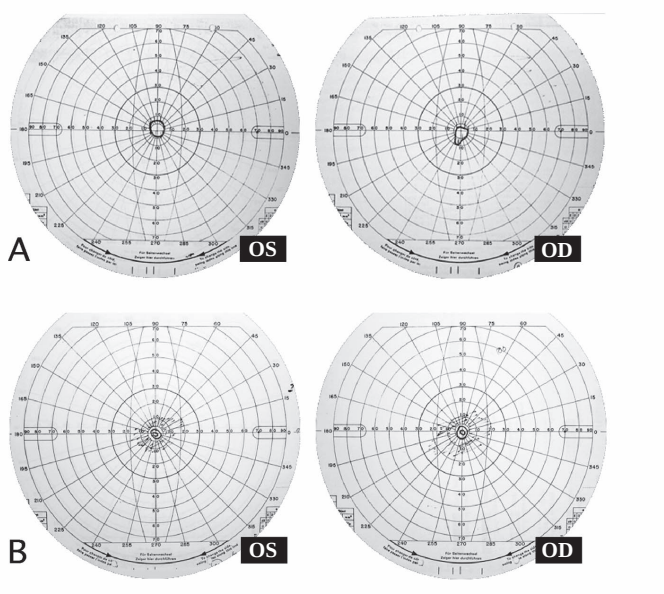

Figure 1 This 33-year-old man presented with chronic progressive visual loss. Visual acuity was 20/20 bilaterally. Fundus shows bilateral optic disc atrophy. A; Visual fields performed by Goldmann perimetry shows central island of vision on both eyes. B; 4 months after chiasmapexy, there was still no improvement on his visual fields.

Case 2

A 56-year-old multiparous woman was found to have multinodular goiter and hypertension after feeling ill about ten years prior to this

visit. She was a regular attendant of the out patient department with good control of both underlying conditions.

Her illness began 2 months ago when she first developed painless visual loss in both eyes. The visual acuity was found to be hand motion in the right eye and 10/200 in the left eye. The intraocular pressure (IOP) measured to be 15 and 16 mmHg in the right and left eye respectively. Fundoscopic examination showed a full blown bilateral papilledema with sphincter hemorrhage. Visual field showed an inferonasal and central field defects in the right eye and two inferior para-central scotomas in the left eye. (Fig 2) MRI of the brain revealed flattening of pituitary gland at floor of sella turcica with normal midline of pituitary stalk. There was no herniation of the optic chiasm or nerve into the sella tursica Neurological examination was completely normal as well as the endocrinological examination. Lumbar puncture revealed high initial pressure of 23 mmH_2O. She was diagnosed as pseudotumor cerebri with primary empty sella syndrome and acetazolamide was prescribed. The papilledema was seen to have improved in during the three months follow up, and the repeated lumbar puncture showed a normal initial pressure.

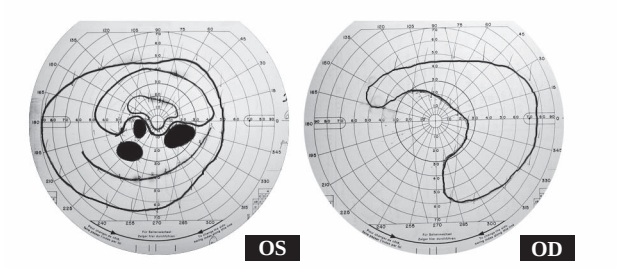

Figure 2 This 56-year-old woman presented with chronic progressive visual loss. Visual acuity was hand motion in the right eye, 10/200 in the left eye. Fundus shows bilateral papilledema with sphincter hemorrhage. Visual fields done by Goldmann perimetry shows inferonasal and central field defects in the right eye and two inferior para-central scotoma in the left eye.

Discussion

Primary empty sella syndrome is postulated as a congenital defect in diaphragm, coupled with normal and constant pulsation of cerebrospinal fluid, pushes the subarachnoid space further and further into the sella, which results in the compression of pituitary gland and the enlargement of sella[5]. This enlargement of sella turcica can be caused by pituitary tumor, suprasellar tumor, optic neuroma, tuberculum sella meningioma, intratrasellar internal carotid syndrome, or increased intracranial pressure[4].

Empty sella is a term that was first introduced by Busch in l951 to describe herniation of the suprasellar arachnoid space, and possibly the optic nerve, into the sella turcica because of a diaphragmatic defect. Busch performed an autopsy study of 788 subjects without known pituitary disease. In 38.4% he found a complete covering of pituitary gland by the diaphragm. He observed an empty sella in 5.5%. In 1926 Schaeffer may have been the first to observe various anatomical forms of the diaphragma sellae; these ranged from densely thick with a complete roof to that composed of a peripheral veil. Busch, in his study of 788 sellae in patients without a history of pituitary disorders, classified the diaphragma sellae into the following categories, which were later resorted by Kaufman in l972[3].

Primary empty sella syndrome typically occur in middle-aged, obese, multiparous woman, many of whom are also have hypertension and pseudotumor cerebri[6]. The associated disorders of an empty sella are mucopolysaccharidosis, virilization and renal tubular acidosis. Headache is the most common symptom and with no characteristic pattern or localization. In 1991, Foresti reported 500 consecutive patients, aged from 11 to 82 years, who underwent MR imaging of the brain for a variety of conditions not related to pathologic processes of the sellar or juxtasellar regions, this report showed primary totally empty sella in 28/248 males (11.3%) and in 34/252 females (13.5%). Primary partially empty sella was found in 40/248 males (16.1%) and in 38/252 females (15.1%). A progressive increase in the incidence of the signs of primary empty sella was observed with aging, the increase being more conspicuous in the 5th decade of life in females (37.5%) and in the 6th decade in males (40%). On the whole, signs of primary empty sella were detected in 140/500 cases (38%), in 9.6% of the subjects under 40 and in 39.9% of those above 40[7].

Reported mechanisms of visual loss associated with primary empty sella include prolapsed of chiasm into the sella, compression of chiasm from above by the anterior cerebral arteries, and occasionally, the involvement of optic nerves and chiasm by arachnoidal adhesion[6]. The incidence is approximately 20% in primary empty sella syndrome. In secondary empty sella syndrome, the incidence is much higher because of the underlying sellar pathology.

Pituitary function is stated to be normal in most patients. Only 31% of these were referred due to endocrinological problems. There is a general agreement in the literature that anterior pituitary dysfunction necessitating hormonal replacement therapy is rare in primary empty sella syndrome. Pituitary hyper-secretion strongly points to the presence of a pituitary adenoma that is partly necrotized. The involvement of the posterior pituitary gland rarely occurs. To establish the diagnoses of hypopituitarism, the measurement of the following hormones are required; serum thyrotropin, T4, corticotrophin, cortisol, LH, testosterone, basal and stimulated growth hormone and prolactin levels. Children with empty sella and GH deficiencies may be prone to develop rhGH-

induced pseudotumor cerebri. The resumption of rhGH therapy after 3 months at a lower dose lead to an excellent catch-up growth without the recurrence of pseudotumor cerebri[8].

Visual field defects were bitemporal hemianopia, binasal hemianopia, quadrantanopia, generalized field constriction, central scotoma, homonymous hemiachromatopsia[1]. Pollock and Bromberg[2] suggested that stable visual field defects, particularly nasal defects or generalized constriction in primary empty sella syndrome might be secondary to previous, under diagnosed pseudotumor cerebri[3]. Harri Rouhiainen et al[1] suggested that primary empty sella syndrome was found in patients with glaucomatous optic disc and visual field changes, so follow up visual field is essential. Only when no progression is observed primary empty sella syndrome be blamed for defect and unnecessary to start the anti-glaucomatous therapy[6]. Although primary empty sella syndrome may cause visual field changes through mechanical traction or ischemic effects on chiasm, visual field defects are not always secondary to the syndrome. Therefore if there are some degrees of glaucomatous field and disc changes are detected, further investigation for glaucoma should be performed[2].

Visual disturbances may be absent or minimal in primary empty sellae and secondary empty sellae with herniation of the supra sella visual system. Progression of the symptoms--visual field defects, optic atrophy, and loss of vision is not inevitable. There was no correlation between the severity of visual symptoms and the degree of herniation of the SVS in either the primary or secondary sellae[9].

Patients with primary empty sella are asymptomatic and do not require any management except if visual deficits are developed or cerebrospinal fluid leakage occurs. Surgical treatment would then be considered to be necessary. Most of the empty sella cases that need surgical treatment are operated on through a transsphenoidal approach. This procedure has been traditionally known as "chiasmapexy"[10].

From previous discussion, it can be said that it is necessary to properly perform both neurological, endocrinological assessment with ophthalmologic examination on patients who have visual field defects and thought to be associated with pseudotumor cerebri with primary empty sella. Particular attention should be paid to the IOP measurement, optic disc and visual field patterns on follow up visits.

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