VISUAL VIGNETTES-PERSISTENT HEADACHES WITH APPARENT EMPTY SELLA SYNDROME

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Running title: Persistent Headaches With ESS

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**Case Presentation**

A 37 year old lady with a background history of hypertension presented with chronic headache and visual blurring. On examination, she was overweight (BMI: 27 kg/m²), her BP was 170/120 mmHg and she had no neurological deficit. Her visual acuity was diminished with enlarged blind spot bilaterally. Funduscopic examination, Figure 1, revealed mildly engorged retinal veins, hyperemic-swollen discs and cotton wool exudates consistent with hypertensive retinopathy. CT brain was normal. A diagnosis of hypertensive emergency was made. Persistent headache despite controlled BP prompted a MRI brain. After the MRI, she was referred to the endocrinologist for an apparent empty sella. Figure 2a and 2b showed an enlarged sella with a rim of normal pituitary tissue seen at the base of the sella, without any mass lesion, dural sinus thrombosis or enlarged ventricles. Reassessment indicated she had regular menses, normal complete blood count and serum electrolytes including serum corrected calcium: 9.0 mg/dL (normal, 8.8-10.4), with an early morning cortisol: 14.8 µg/dL (normal, 5.2-22.3), fT4: 1.2 ng/dL (normal, 0.8-1.8), TSH: 0.9 mIU/L (normal, 0.5-4.8), IGF-1: 175 ng/mL (normal, 109-284), estradiol: 53.9 pg/mL (normal, 19-144), LH: 1.7 IU/L (normal, 0.5-16.9), FSH: 2.6 IU/L (normal, 1.5-9.1) and prolactin: 15.9 ng/mL (normal, 2.8-29.5). Lumbar puncture demonstrated an elevated opening pressure of 47 cm H₂O (normal, < 25 cm H₂O for overweight/obese) with normal cerebrospinal fluid (CSF) composition. What is the diagnosis?

**Answer**

A diagnosis of idiopathic intracranial hypertension (IIH) was finally made and she was started on acetazolamide 500 mg twice daily resulting in improvement of headache and vision. IIH is classically seen in middle aged overweight females with normal endocrine function, as in our patient. Empty sella syndrome from IIH is often overlooked despite being reported in up to 94% of cases. (1) Chronic elevation of intracranial pressure in IIH causes intrasellar herniation of CSF through a rudimentary diaphragmatic sellae, expanding the sella turcica. (2) As the sella is filled with CSF, the pituitary tissue is flattened to the base of the sella with the infundibulum extending down to the floor of the sella, giving rise to an apparent empty sella. The empty sella has been shown to be reversible with treatment of IIH. (3) In patients with intact pituitary function and chronic headache or visual deterioration, a funduscopic evaluation with measurement of CSF opening pressure should be considered to avoid missing the diagnosis of IIH and unnecessary investigations for an apparent empty sella seen on MRI.
References:


Figure Captions

Figure 1: Fundoscopic images showing mildly engorged retinal veins, hyperemic and swollen discs with blurred disc margins (right superior) without the physiologic cups of optic discs being ablated and cotton wool exudates (right posterior pole/ left nasal)
Figure 2: Sagittal (a) and coronal (b) MRI of the brain showing expanded sella turcica, with flattened pituitary tissue at the sella floor (indicated by arrows)