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EMPTY SELLA SYNDROME: THE GREAT IMITATOR OF VISUAL COMPLAINTS

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ABSTRACT

Introduction: Empty Sella Syndrome is a rare condition of sella turcica malformation, resulting in pituitary gland shrinkage. It may manifest as neurological symptoms, endocrine disorders, visual disturbances or even incidental findings during imaging. However, patients rarely come with typical complaints thus an extended course and review of this case is needed to prompt suspicion and aid in the diagnosis of ESS.

Case Presentation: We report a 45-year-old woman who presented with cephalgia and bitemporal hemianopia and previous transient bilateral visual loss and foggy vision. A history of ovarian cancer was noted. Physical examination showed myopic astigmatism ocular dextra sinistra (ODS) with increased intraocular pressure and bitemporal hemianopia ODS. Posterior chamber examinations were within normal limits. Optical coherence tomography showed nasal retinal nerve fiber layer ODS being the thinnest among the rest, although still within normal limits. Neurological examinations also suggested presence of central vertigo. This subsequently resulted in a suspicion towards space occupying lesion chiasmal compression through metastasis.

Discussion: MRI revealed an empty sella turcica. A laboratory examination was then ordered and showed normal endocrinology results. The diagnosis of Primary Empty Sella Syndrome with ocular hypertension was established. The patient was prescribed with timolol, codeine and dexamethasone. A month later during her follow-up, the patient did not show improvement and was suggested to do an MRI with contrast, however this patient was lost to follow up.

Conclusion: This case report highlighted the manifestation to raise suspicion and aid in the diagnosis of ESS for an optimal and improved diagnosis and treatment of ESS.

Keywords: Bitemporal Hemianopia, Empty Sella Syndrome, Primary Empty Sella Syndrome, Secondary Cephalgia

INTRODUCTION

Empty sella syndrome (ESS) is a rare condition characterized by enlargement or malformation a sella turcica causing the pituitary gland to shrink or flattened. It is caused by a herniation of subarachnoid space into the sella turcica. This syndrome is also called arachnoidocele and it is a distinct anatomical and radiographic entity. The prevalence of ESS varies, in autopsy studies 5.5-12% of ESS cases have been reported. On the other hand, an estimation of 12% cases was found on imaging by incidence.[1]

ESS has two etiologic categories, which are primary/idiopathic ESS and secondary ESS. Primary empty sella syndrome (PES) is a result of an anatomical variation in the diaphragma sellae. One of the possible mechanisms is due to the insufficiency of the sellar diaphragm and, subsequently, this will allow the arachnoid to herniate through the diaphragmatic opening.[2] The increase in Cerebrospinal Fulid (CSF) hydrostatic pressure will compress the pituitary gland against the sellar floor causing it to flatten. Another mechanism proposed is due to intracranial hypertension that may be idiopathic or can also be a result from tumors, infections, venous thrombosis or malformations. The prevalence of PES in

patients with idiopathic intracranial hypertension ranges from 70-94%.[3]

Secondary Empty Sella Syndrome (SES) can be caused by pituitary tumor shrinkage after medical treatment, radiotherapy or surgery. Other risk factors include Sheehan's syndrome, pituitary infection, pituitary apoplexy and traumatic brain injury.

PES can be an incidental finding because only a few will present with clinical symptoms related to the condition. It may be detected during imaging for neurological symptoms, endocrine disorders, or visual disturbances. Visual disturbances manifested, e.g., transient vision loss, vision blurring and/or visual field defect. Cephalgia is also a commonly reported symptom.[4] To this day, there has not been a specific guideline or recommendations for the diagnosis and management of ESS.

This case report depicts a 45 year old woman presenting with cephalgia and a variety of visual complaints with a history of ovarian cancer. A diagnosis of space occupying lesion (SOL) chiasmal compression through means of metastasis was suspected. However, on MRI revealed an empty sella turcica. This article highlighted the symptoms manifested in our empty sella syndrome patient.

CASE PRESENTATION

Our patient was a 45-year-old woman presenting to the ophthalmology clinic with a variety of visual complaints and cephalgia 3 years prior to presentation. Initially, the patient experienced transient complete loss of bilateral vision with cephalgia. 2 years ago, our patient experienced transient bilateral foggy sight which ceased spontaneously. In the same year, the patient experienced bitemporal hemianopia, with right temporal field loss worse than the left temporal field. The patient had a history of ovarian cancer.

The patient had no past interventions for these complaints beforehand. Upon examination, the patient's uncorrected visual acuity for visus oculi dextra (VOD) was 0,05 and visus oculi sinistra (VOS) was 0,1. The patient's best corrected visual acuity was VOD 0,6 with S-1,00 C-1,00 X 165 and VOS 0,7 with S-1,25 C-1,00 X10. Pinhole test was negative for both eyes, corneas were clear, anterior chambers were not shallow, the irises were regular and symmetrical with no synechiae, lenses were clear and fundoscopy were within normal limits. However, using non-contact tonometry, the patient showed increased intraocular pressure (OD = 20,7 mmhg OS = 23 mmHg). Perimetry using Humphrey's machine was performed revealing tunnel vision of both eyes. Visual field index respectively was VOD 41%, and VOS 19%.

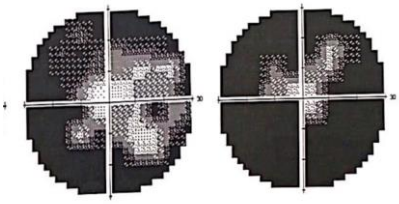

Figure 1. Perimetry examination of our patient showed visual field index of VOD 41% and VOS 19%

On funduscopic examination vitreous was clear, optic nerve disc was well demarcated with cup to disc (C:D) ratio <0,5. Macula was seen temporally to the optic disc with vessels only surrounding the periphery.

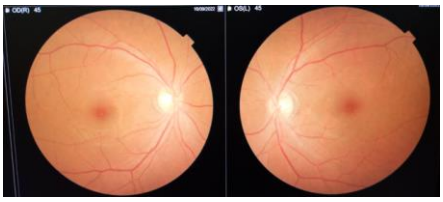

Figure 2. Funduscopic examination of our patient showed clear vitreous, well demarcated optic nerve disc with C:D ratio <0,5. Macula was seen temporally to the optic disc with vessels only surrounding the periphery.

Optical coherence tomography (OCT) examination revealed that the right and left optic nerve cup were 0,15 mm^3. Vertical C:D ratio of the right eye was 0,61 and the left eye 0,58. On retinal nerve fiber layer (RNFL) analysis, RNFL thickness map of the right and left eyes showed normal thickness appearance with superior and inferior RNFL being thicker than nasal and temporal RNFL. Temporal, superior, nasal, inferior, temporal (TSNIT) Graph also showed double-hump appearance which was correlated to a normal RNFL thickness pattern. RNFL symmetry between the right and left eye was 82% (normal range 76-95%). RNFL Quadrants of the left eye showed a thickness of 83, 133, 79, 139 on the temporal, superior, nasal and inferior quadrants respectively. RNFL Quadrants of the left eye showed a thickness of the right eye showed a thickness of 94, 135, 73, 147 on the temporal, superior, nasal and inferior quadrants respectively. Although these thicknesses were considered normal, the patient's OCT did not follow the inferior ≥ superior ≥ nasal ≥ temporal (ISNT) rule indicating that both nasal RNFLs were thinnest. However the nerve thinning pattern did not occur in an inferior, nasal, superior and temporal arrangement, glaucoma was ruled out.[5]

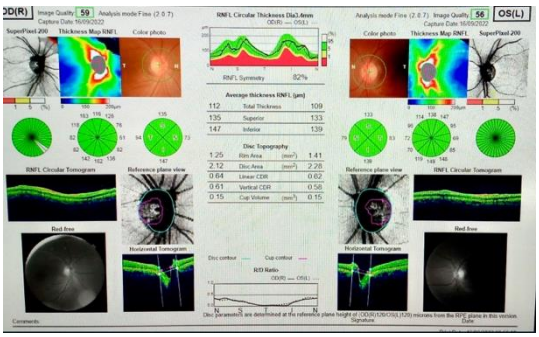

Figure 3. OCT examination of our patient showing the binasal RNFL being the thinnest among the others, although still within normal limits.

Physical neurologic examination was also done. Romberg test was abnormal both when the patients' eyes were opened and closed and coordination was poor. These findings suggested the presence of central vertigo. A preliminary diagnosis of a metastasized SOL was established. Head MRI without IV contrast was then ordered, however neither SOL, infarction nor bleeding were observed within the scans. However it revealed an empty sella turcica.

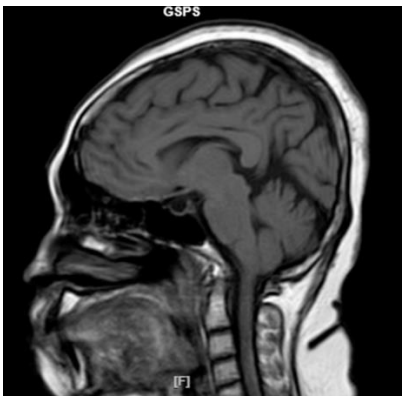

Figure 4. MRI without IV contrast of our patient for the last follow-up visit showed empty sella syndrome (white arrow) without signs of intracranial SOL, bleeding or infarction

Prolactin, Total T4 and Morning Cortisol levels were within normal limits (8,01 ng/mL, 8,20 ug/mL and 10,06 ug/mL, respectively). The patient denied history of pituitary adenoma and radiotherapy of the cephalic area. The diagnosis of PES with ocular hypertension was made. The patient was prescribed with timolol 0,5% given 2 times daily on both eyes to decrease IOP, codeine 3 x 15 mg PO to alleviate the patient's cephalgia, and dexamethasone 3 x 0,5 mg PO. The patient was also referred to Neurology and Neurosurgery departments.

After a month follow up, the patient did not show improvement. The patient's best corrected visual acuity worsened to VOD 0,6 with S-1,00 C-1,00 X165 and VOS 0,7 with S-1,25 C-1,00 X10. However IOP reduction was noted using non-contact tonometry (OD = 15, OS = 17). MRI with

contrast was suggested however the patient was lost to follow up.

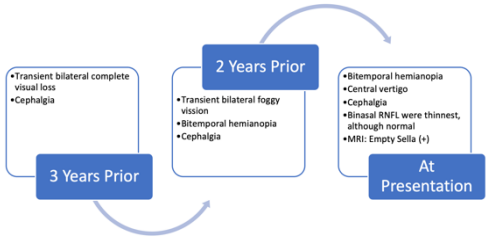

Figure 5. Our Patient's Symptoms Timeline

DISCUSSION

Primary Empty Sella Syndrome is due to congenital absence of diaphragm sellae or an increase of intracranial pressure from the CSF. This syndrome was found to be four times more common among women as with our case.[6] To this day, the etiology of this syndrome is still not clear and various hypotheses are still being studied. Patients diagnosed with PES may demonstrate symptoms that reflect the loss of endocrine dysfunctions, cephalgia, and/or visual impairments. Secondary Empty Sella Syndrome may cause progressive visual impairments as well, due to the effects of radiation therapy for pituitary adenomas that result in adhesions and fibrosis around the optic nerve, chiasm, vascular changes or radiation injury to the optic structure. However in PES, visual impairments are caused by the downward displacement of the optic nerve and chiasm as they are pushed into the pituitary fossa due to the fibrous retraction. Our patient was diagnosed with PES as a history of pituitary adenoma and radiotherapy of the cephalic area were denied.

Transient visual disturbance, nystagmus, diplopia and blurred vision were some of the commonly reported visual impairment in a retrospective analysis of PES cases.[7,8] Our patient had a variety of visual complaints and cephalgia. Although she came with bitemporal hemianopia, prior to this complaint, the patient had already experienced transient visual losses which have been reported in previous cases as well.[9]

Although transient visual losses have been reported in ESS patients, to our knowledge, transient foggy sight has never been reported as a symptom of ESS.[9,10,11,12] Other etiologies of hazy sight such as keratopathies, cataract, retinopathies and maculopathies were ruled out as physical examination showed normal results. This complaint is thought to be associated with PES.

Endocrine abnormalities in PES cases vary, ranging from normal pituitary function to prominent pituitary dysfunction. Our patient did not have a history of endocrine dysfunction such as galactorrhea, amenorrhea, irregular cycle, or infertility and further confirmed by our patient's normal hormone profile.

Radiographic findings using an MRI or computerized tomography (CT) imaging is required in order to confirm the diagnosis of empty sella and a typical presentation usually demonstrates an enlargement of the sella turcica, CSF filling the sella and compression of the pituitary gland against the sella floor.[13] Head MRI non contrast result of our patients also showed high CSF intensity in intra sella which indicates an empty sella. Other differential diagnosis such as space occupying lesion, infarct or hemorrhage were not detected in the MRI results, hence they were ruled out. PES due to an increase in intracranial pressure was also ruled out in our patient because fundoscopy revealed no presence of papilledema. Thus, the etiology of empty sella in our patient was idiopathic.

OCT was done in our patient which showed binasal RNFL being the thinnest compared to the rest, although still within normal limits. A previous case report of a patient with SES due to prolactinoma treatment which presented with bitemporal hemianopia showed presence of binasal

RNFL thinning.[12] This may indicate that RNFL thinning in ESS patients correlates with their visual field defect, and nothing specific can be found in OCT.

This case report has its limitations, one being the patient was lost to follow up. Further evaluation and treatment plans could not be initiated. Based on available literature, treatment principles of ESS was to correct hormonal imbalance and symptomatic treatment.[14] Progressive dysopsia and CSF rhinorrhea were indications of surgery.[7,15] Our patient's therapy was given to reduce cephalgia and IOP. The aim of surgery is to fill the empty sella using a fat pad, fascia, cartilage, muscles or balloon.[15] However, as our patient was lost to follow up, this treatment could not be done.

Another limitation was the inaccuracy of the patient's perimetry results. Perimetry showed presence of tunnel vision but it had a high false negative (>15% using SITA-Standard strategy). A second perimetry could not be done due to lost to follow up. Therefore, due to this inaccuracy the authors could only assume the patient's visual field loss pattern was bitemporal hemianopia and not tunnel vision as this correlated more to the patient's complaint.

CONCLUSION

Empty Sella Syndrome is a rare disease which affects mostly women. Primary Empty Sella Syndrome may manifest as cephalgia with a variety of visual impairments including bitemporal hemianopia, transient bilateral visual loss and foggy vision. One patient may experience more than one visual complaint, as manifested in our patient. Nothing specific can be found in OCT. It is therefore important to keep PES in mind, leading to early diagnosis and better treatment for the patient.

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