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Case Report

Resolution of symptomatic secondary empty sella syndrome following lumbar–peritoneal shunt

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Abstract

Background: Post-surgical empty sella is related to the removal of pituitary tumors either from the transcranial or transphenoidal route, rendering diaphragma sellae incompetent at the end of the procedure. This subsequently leads to herniation of the third ventricle and optic apparatus into the empty sella. Studies have shown that in 50% of the cases, individuals with primary and secondary empty sella syndrome have developed defects in the visual fields. Benign increased intracranial pressure, cerebrospinal rhinorrhea, papilledema, and abnormalities affecting visual acuity may also occur as a result of empty sella.

Case Description: This case report discusses a rare treatment option for the symptomatic secondary empty sella syndrome. Patient underwent lumbar drain placement and that resulted in astonishingly significant improvement in vision. Keeping in view the beneficial effect of lumbar drain, lumbar–peritoneal (LP) shunt was inserted which showed drastic improvement in vision.

Conclusion: The surgical outcome of symptomatic cases of ESS is favorable. Various common surgical options were reported in literature; however, we have discussed an unconventional surgical option with an impressive outcome.

Key Words: Empty sella turcica, optic chiasma, pituitary adenoma, visual fields

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INTRODUCTION

Post-surgical empty sella is related to the removal of pituitary tumors either from the transcranial or transphenoidal route, rendering diaphragma sellae incompetent at the end of the procedure.^[1] This subsequently leads to herniation of the third ventricle and optic apparatus into the empty sella.^[1,2] Studies have shown that in 50% of the cases, individuals with primary and secondary empty sella syndrome have developed defects in the visual fields.^[2] Benign increased intracranial pressure, cerebrospinal rhinorrhea, papilledema, and abnormalities affecting visual acuity may also occur as a

result of empty sella.^[1,2] In this report we have described the case of a patient who developed empty sella syndrome after transphenoidal surgery. She presented with

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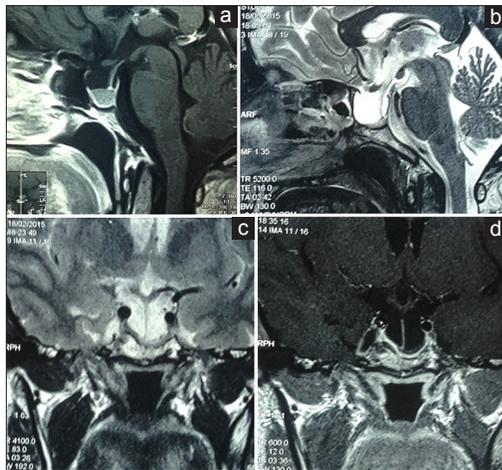


Figure 1: (a) Presurgical contrast enhanced T1 Sagittal MR image of the pituitary gland showing macroadenoma. Post surgical images (b) Sagittal T2 (c) Coronal T2 and (d) Post contrast T1 weighted Coronal images of the pituitary gland showing residual gland and evident empty sella turcica with downward herniation of optic chiasm and suprasellar structures

profound visual impairment. She was treated successfully using a lumbo-peritoneal shunt.

CASE REPORT

A 51-year-old lady was admitted in Aga Khan University Hospital, Karachi, with complaints of headache and blurring of vision. She was diagnosed as having pituitary macroadenoma [Figure 1a] 8 months back for which she underwent endoscopic pituitary adenoma resection. The patient had persistent generalized headache that ameliorated on head elevation up to 40°-45° and got worse in the morning. It was associated with phonophobia but not associated with nausea or vomiting. She had bitemporal hemianopia with decreased visual acuity in both eyes. The symptoms got worsened following surgery. She also developed increased urinary frequency post-operatively. Other than pituitary macroadenoma resection, the patient underwent partial thyroidectomy for a solitary nodule in 2000 (euthyroid), and hysterectomy in 2001 due to dysfunctional uterine bleeding. She had a medical history of hepatitis C treatment with interferon 10 years back. Currently, she is on selective serotonin reuptake inhibitor (SSRI), desmopressin nasal spray, and certain analgesics. An examination showed significant bitemporal hemianopia without papilloedema. Higher mental functions and motor and sensory examination were unremarkable. Cranial nerves were all intact. Magnetic resonance imaging (MRI) of the brain showed evident empty sella turcica with downward herniation of optic chiasm [Figure 1b-d]. The patient was planned for lumbar drain insertion to lower down intracranial pressure (ICP) which would help in ameliorating downward herniation of optic chiasm. The opening pressure of more than 35 cm of H₂O was measured.

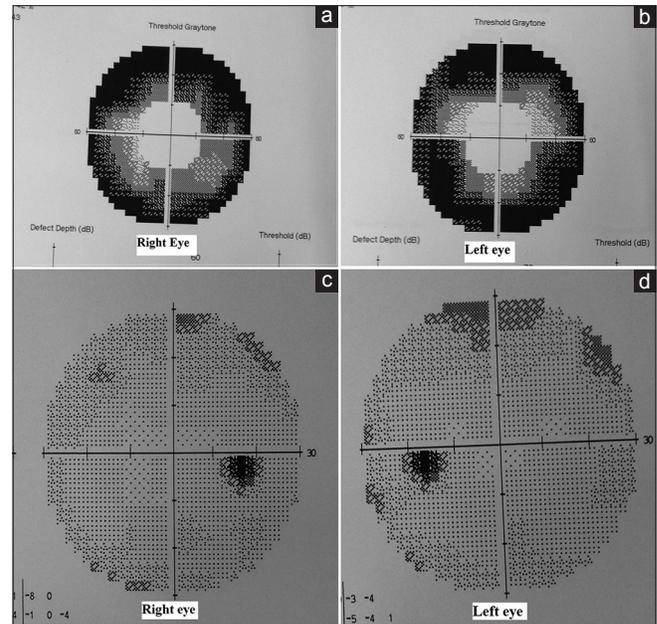


Figure 2: (a and b) Right and left eye visual field before LP shunt insertion (c and d) Right and left eye Visual field after LP shunt insertion. Visual field testing results obtained showed dramatic improvement of the preprocedural visual field defects

The outcome of the intervention was noticed soon after the surgery with transient improvement in vision. For long-term outcome, lumbar-peritoneal (LP) shunt was placed after a period of 1 month which resulted in both subjective and objective improvement in visual acuity [Figures 2 and 3]. Inclusively, associated diabetes insipidus got better.

DISCUSSION

This case report highlights a rare treatment option for the symptomatic secondary empty sella syndrome resulting from post-surgical pituitary resection. Various studies have mentioned ways to prevent downward herniation of optic chiasma that have been resulting in visual defects in subjects. Around 50% of patients show visual field defects associated with both primary and secondary empty sella syndromes.^[1] Visual alterations may be due to traction on the chiasm or involvement of chiasm blood vessels. In secondary empty sella syndrome, the incidence is much higher because of the underlying sellar pathology. Clinically, the patients complain about clouding of vision, color vision defects, photophobia, and various visual field defects (bitemporal hemi- or quadrantanopia, generalized field constriction, quadrine constriction, central scotoma, homonymous hemiachromatopsia mimicking the lesion in patients with a suprasellar pituitary tumor).^[2] Secondary empty sella has been noted to follow the below:

- Sellar or parasellar surgery
- Radiation therapy for an intrasellar expansion
- Bromocriptine therapy for a pituitary adenoma.

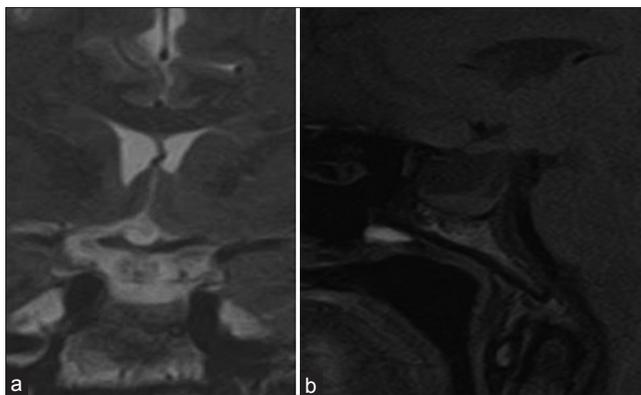


Figure 3: (a and b) T1 and T2 weight MR images from the sellar region following Lumbar-Peritoneal shunt procedure shows a reversal of chiasmatic herniation and reduction in the degree of empty sella turcica

Radiation-induced vascular changes or strangulation of the optic nerves or chiasm are thought to compromise local blood flow. Downward herniation of the optic pathways is present in the majority of cases.^[5] Several materials have been suggested for filling the sellar space and reconstruction of the sellar floor. They include bioabsorbable materials, muscle, fat, dural substitutes, cartilage, bone fragments, ceramic substances, titanium plates, and others.^[3,4] The surgical process of propping up the optic chiasm is called chiasmopexy, which includes inserting muscle, cartilage, or silicone sponge under optic chiasm, packing the sella with fat, muscle, or cartilage to elevate the pituitary gland, pituitary stalk, and chiasm and transsphenoidal placement of a detachable balloon.^[6] Lumbar drain insertion has a mechanism of reducing the cerebrospinal fluid (CSF) pressure that may build up and contribute in downward optic chiasma herniation.

The ultimate possible improvement in symptoms of a patient with secondary empty sella syndrome is placing ventriculoperitoneal shunt for long-term beneficial effects.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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