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Case Report

# Persisting Embryonal Infundibular Recess Masquerading as a Nasal Mass


## Abstract

Persisting embryonal infundibular recess (PEIR) is a rare anomaly of the development of the posterior pituitary wherein there is a defect in the third ventricular floor. Earlier reports have found PEIR descending only up to the sella. However, this is the first case of PEIR presenting as a pulsatile nasal mass. A 35-year-old female presented to the otorhinolaryngologist with hyposmia. Diagnostic nasal endoscopy revealed a pulsatile nasal mass. Magnetic resonance imaging revealed a cystic lesion extending from the third ventricular floor to the nasal cavity. There was no associated hydrocephalus or empty sella. The case was successfully managed by the endoscopic endonasal transsphenoidal approach.


**Keywords:** *Encephalocoele, nasal mass, persisting embryonal infundibular recess*


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## Introduction

Persisting embryonal infundibular recess (PEIR) is an anomaly of the development of the posterior pituitary. This is a rare condition with only eight cases reported in world literature.[1] In previously reported cases, the defect was demonstrated in the third ventricular floor, with the sac descending inferiorly only up to the sella. However, this is the first case of PEIR, with the sac descending into the nasal cavity as pulsatile nasal mass. It can be misdiagnosed as a nasal polyp or basal encephalocoele unless magnetic resonance imaging (MRI) identifies the communication with the third ventricular floor. There are management concerns with higher chances of cerebrospinal fluid (CSF) leak unless dealt with appropriately.

## Case Report

The case we report is that of a 35-year-old female who presented to the otorhinolaryngologist with hyposmia. Diagnostic nasal endoscopy showed a pulsatile mass in the roof of the nasopharynx extending up to the posterior margin of the middle turbinate along with multiple nasal polyps [Figure 1a]. Further evaluation with CT showed a cylindrical bony structure in the sphenoid sinus with a central hypodense area extending from the third ventricular floor to the nasopharynx [Figure 1b]. A 3 Tesla MRI showed a cystic lesion extending from with the floor of the third ventricle, to the sphenoid sinus and down into the superior aspect of the nasopharynx submucosally. There was herniation of anterior pituitary gland along the posterior margin of the defect with a shallow sella turcica. The prechiasmatic optic nerve was thinned out and stretched with a v-shaped configuration of the optic chiasm sinking into the superior portion of the defect anteriorly [Figure 1c]. The further hormonal evaluation revealed hypocortisolism. Her visual acuity and fields were found to be within the normal limits.

The differential diagnosis of an encephalocoele and PEIR were considered. The endoscopic transnasal transsphenoidal approach was chosen as anterior skull base repair can be performed effectively by this approach.

She underwent navigation-assisted endonasal endoscopic transsphenoidal repair and reconstruction under general anesthesia. Intraoperatively, there was a bony defect in skull base at the anterior edge of the sella, extending as a cylindrical bony canal down up to the anterior face of the sphenoid sinus, with a soft pulsatile mass extending from the intracranial space into the nasopharynx. It was covered with mucosa at the lower end. The bone was drilled and soft tissue overlying the sac was excised. The sac was freed all around. The optic chiasm was identified after



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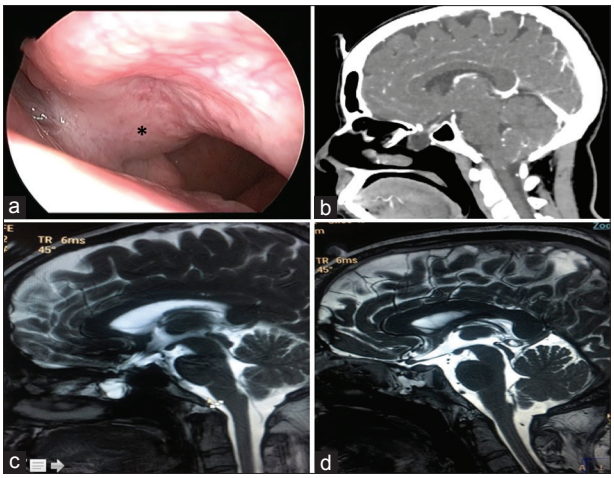


**Figure 1: (a) Diagnostic nasal endoscopy image showing persisting embryonal infundibular recess presenting as a nasal mass in the superior nasopharynx (*). (b) Contrast computed tomography brain showing a cystic lesion herniating through a bony defect anterior to shallow sella. (c) Preoperative magnetic resonance imaging constructive interference in steady state sequence showing the communication of the third ventricular floor with the cyst through a persistent embryonal infundibular recess. (d) Postoperative magnetic resonance imaging constructive interference in steady state sequence showing reduction of the sac and reconstruction of the defect**

opening the anterior dura using navigation assistance. The sac contained gliotic brain matter and CSF. The sac was arising from the floor of the third ventricle. Sac was reduced. The reconstruction of the defect was with fat, nasal septal cartilage, fascia, and right nasoseptal mucoperiosteal (Hadad) flap in that order and reinforced using fibrin glue. The lumbar drain was retained until the third postoperative day.

She underwent an immediate post-operative MRI that showed evidence of good surgical repair of the encephalocoele with the restoration of anatomy of the optic chiasm and pituitary region [Figure 1d].

Her postoperative period was uneventful, and she was discharged on the fifth postoperative day. She was fine until the last outpatient review at 6 months following surgery with complete recovery of hormonal functions. Her vision charting remained normal on follow-up.

## Discussion

The hypophysis develops from two processes of different origin joining together during the 1st month of gestation. The adenohypophysis develops from the Rathke's pouch, which is an ectodermal outpouching that grows upward from the roof of the mouth. The neurohypophysis develops from the neuroectodermal process – the infundibulum, which evaginates ventrally from the diencephalon of the developing brain. The infundibulum is initially hollow but normally obliterates by cellular proliferation during the 45 mm length stage to form the pituitary stalk except for its proximal part, which persists as the infundibular recess, at the third ventricular floor. When this normal obliteration of the infundibular recess does not take place, it persists as a communication with the third ventricular floor, PEIR.

This condition may be associated with hypo-function of the pituitary gland, as found in our case, which had hypocortisolism. Hence, complete hormonal evaluation and endocrinologist reference is essential before corrective surgery. The patient had complete recovery of pituitary function and was off hormonal replacement by the 3rd month follow-up.

In our case, the optic chiasm was V-shaped and descended into the fundus of the defect. This could be associated with visual deficits: hence, a complete visual field assessment by an ophthalmologist is essential in these cases. A proper study of the anatomy of the optic apparatus, on a good quality MRI, before corrective surgery is also essential.

The diagnosis of PEIR can be missed as a nasal polyp or basal encephalocoele, unless MRI identifies the communication with the third ventricular floor. This can be done by the close study of the third ventricular floor on a constructive interference in steady state (CISS) sequence in a 3 Tesla MRI.

The indication for surgery, in this case, was the risk of cyst rupture and CSF leak, which may lead to meningitis. Other indications are visual or hormonal deficits. Most previous reported cases, 7 out of 8, were managed by transcranial surgery.[1] One earlier case report of meningitis following Rathke's cyst repair due to PEIR was managed successfully by the endonasal endoscopic transsphenoidal approach.[2] We managed this case by dissecting and freeing the sac from the posterior septal wall, defining the edges of the defect and multilayer reconstruction of the defect using fat, nasal cartilage, fascia, and reflected mucosal Hadad flap based on the sphenopalatal artery.

## Conclusion

PEIR is a rare anomaly caused by the nonobliteration of the distal diencephalic evagination during the development of the posterior pituitary. It can occur in isolation without hydrocephalus. It could present as a nasal mass. Careful study of the floor of the third ventricle by CISS sequence on a 3 Tesla MRI is useful to identify the defect. Endonasal endoscopic transsphenoidal surgery is a good option in the management if the defect is reconstructed well.

### 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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## Conflicts of interest

There are no conflicts of interest.

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