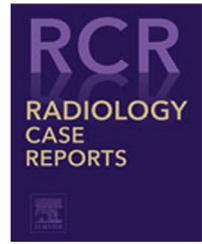


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Case Report

MRI of ectopic posterior pituitary gland with dysgenesis of pituitary stalk in a patient with hypogonadotropic hypogonadism

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ABSTRACT

The ectopic posterior pituitary is a rare condition which is characterized by the ectopic location of posterior lobe of pituitary, pituitary stalk abnormalities, and associated clinical manifestations of anterior lobe related growth hormone dysfunction or less commonly multiple anterior pituitary dysfunctions. We present a rare case of posterior ectopic pituitary and pituitary stalk hypoplasia with isolated hypogonadotropic hypogonadism in a 36-year-old female patient.

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Introduction

The classical features of ectopic posterior pituitary (EPP) (neurohypophysis) include its ventral location at the floor of the third ventricle, near the median eminence [1,2]. The associated infundibular stalk anomalies include its absence, interrupted stalk, or markedly hypoplastic stalk which is barely visible as a thin structure [1]. The clinical presentations of EPP are more commonly related to isolated growth hormone deficiency (IGHG). The condition, however, may progress to panhypopituitarism. Generally, the posterior pituitary functions remain preserved. Magnetic resonance imaging (MRI) is essential to establish the entire spectrum of pituitary anomalies, to

detect associated structural abnormalities of brain and to exclude presence of mass lesions or infiltrative disease affecting the hypothalamic–pituitary axis [3,4].

Case report

A 36-year-old female patient with a history of primary amenorrhea and suspected secondary hypogonadism had an MRI examination to exclude any pituitary gland pathology. Her growth parameters were normal since childhood. She never had any hypoglycemic symptoms to suggest either adrenal or thyroid dysfunction. There was no history of anosmia or vi-

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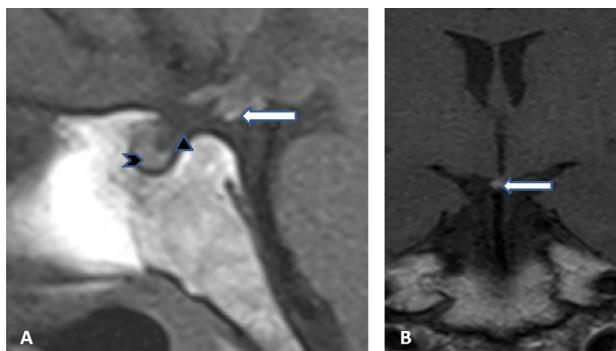


Fig. 1. – The unenhanced T1-weighted sagittal (A) and coronal (B) magnetic resonance images confirm the ectopic location of posterior pituitary, seen as a bright focus (arrows). The midline sagittal image (A) shows small sella turcica with normal intrasellar position of the anterior pituitary (chevron) and presence of thin barely visible infundibulum (triangle).

sual symptoms. Her recent endocrine investigations showed undetectable oestradiol, very low gonadotrophin levels and the short synacthen test had shown adequate adrenal reserve. Her free thyroxine level was normal. Her Insulin-like growth factor-1 was slightly below the reference range at 6.5 nmol/l (8.9-29.5). She denied ever taking any form of oestrogen treatment. MRI revealed a small sella turcica with a nonpneumatized sphenoid sinus and normal intrasellar position of the anterior pituitary gland. The posterior pituitary, however, was not visualized within the sella and was confirmed in an ectopic position near the hypothalamus, in the region of the median eminence and tuber cinerium.

The unenhanced T1-weighted magnetic resonance images did not demonstrate the characteristic bright spot of posterior pituitary within the sella which was higher in position, in the region of median eminence (Arrow, Fig. 1A and B). The pituitary stalk was present, but markedly hypoplastic (Triangle, Figs. 1 and 2). The postcontrast images showed normal enhancement of the intrasellar anterior pituitary (Chevron, Fig. 2A). The EPP was seen as an enhancing nodule near the median eminence (arrow) and the infundibulum was seen as a uniform thin nonenhancing hypoplastic structure (Triangle, Fig. 2A). The stalk measured a maximum of 0.8-1 mm in width in our case whereas the normal size range of infundibular stalk is about 3.5 mm near median eminence and 2 mm near the apex [4]. The T2-weighted sagittal image (Fig. 2C) demonstrate subtle appearance of posterior pituitary (arrow) and normal looking other midline structures. Subsequently, formal MRI examination of head and orbits was performed which did not reveal any additional structural abnormality or any mass lesions. The corpus callosum, optic nerves, and optic chiasm demonstrated a normal appearance. For comparison purpose, 2 normal looking cases are included (Fig. 3) which illustrate intrasellar position of bright focus of posterior pituitary at the T1-weighted images (arrows).

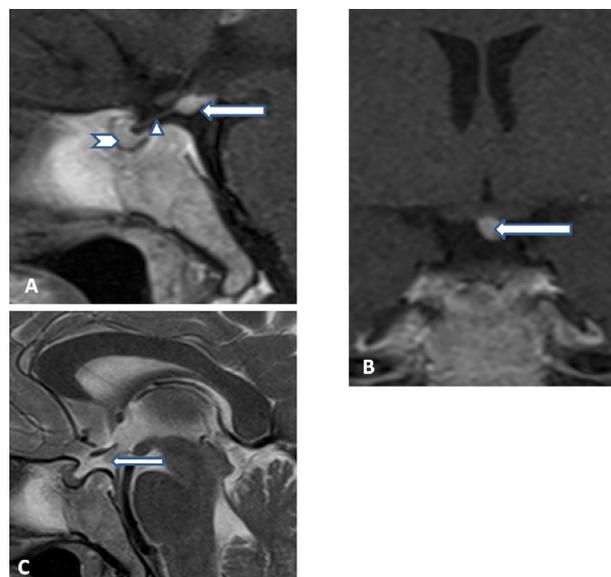


Fig. 2. – The contrast-enhanced mid-line T1-weighted sagittal (A) and coronal (B) demonstrate the ectopic posterior pituitary as an enhancing nodule (5 x 4 mm) at the median eminence (arrows). The sagittal image (a) demonstrates normally enhancing intrasellar anterior pituitary (chevron) and thin non-enhancing infundibulum (triangle). The T2-weighted sagittal image (C) shows subtle appearance of ectopic posterior pituitary (arrow) and other normal looking midline structures.

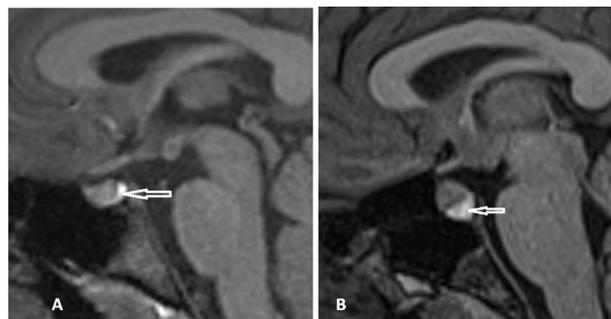


Fig. 3. – Unenhanced T1-weighted sagittal images (A and B) from two different patients, to illustrate the normal position of hyperintense focus of posterior pituitary within the sella turcica (arrows).

Discussion

The anterior pituitary is considered ectodermal in origin and develops from an evagination of the stomodeum (Rathke-cleft). The neurohypophysis or posterior pituitary is of neuroectodermal origin and develops as a downward extension of the diencephalon (infundibulum). The pituitary stalk is also referred in literature as infundibulum or infundibular stalk and is seen as a funnel-like structure connecting the median eminence of the hypothalamus to the pituitary gland. The

posterior pituitary is seen as a characteristically bright spot on unenhanced MR Imaging. Kucharezyk et al. [2] confirmed in their study that the magnetic resonance characteristics of phospholipids and not the antidiuretic hormone is responsible for the MR appearance of the posterior pituitary gland.

The median eminence is anatomically seen at the base of hypothalamus where the hypothalamic releasing or inhibiting hormones are released into portal venous capillaries which traverse the stalk and subsequently influence the adenohypophysis (anterior pituitary). Any structural disturbance in this region can adversely affect the hypothalamo-hypophyseal regulation [6,7]. The EPP can result from defective embryogenesis disturbing the neuronal migration. EPP may occur in isolation or be associated with varying degree of stalk anomalies. The specific pituitary stalk interruption syndrome include stalk hypoplasia, interruption or absence of stalk, hypoplastic anterior pituitary, and EPP [1,5,6].

Breech deliveries, neonatal hypoxia, hypoglycemia and jaundice are important predisposing factors for EPP and stalk anomalies, due to ensuing neuronal reorganization of the proximal stump of the infundibulum [1]. Several genetic factors (PITI, PROPI, HES X1, LHX 3, LHX4, SOX3) have been attributed as influencing factors in this disease entity because these genes normally contribute to the development of hypothalamic–pituitary axis [8]. The literature review also suggests association of EPP with several congenital malformations that include septo-optic dysplasia, optic chiasm hypoplasia, holoprosencephaly, periventricular nodular heterotopia and absence of internal carotid artery [9]. The genetic condition Kallman syndrome includes classical features of hyposmia or anosmia in association with hypogonadotropic hypogonadism and EPP [9]. Our case, however lacked the characteristic clinical feature of smell disorder. Furthermore, pituitary–hypothalamic axis mass lesions such as pituitary macroadenoma, craniopharyngioma, congenital hamartomas and germinomas can present with secondary hypogonadotropic hypogonadism [4,8–10]. Hypogonadotropic hypogonadism can also be caused by various infiltrative conditions that include, sarcoidosis, lymphocytic hypophysitis and histiocytosis affecting the hypothalamo-pituitary axis [11,12].

The EPP may present with IGHD or multiple anterior pituitary hormone deficiency, depending upon the severity of structural abnormality [9,10]. Panhypopituitarism with features of diabetes insipidus is however rare. Jagtap et al [1] showed in their study that MRI confirmed congenital pituitary abnormalities in 49% cases of IGHD and 94% of patients with multiple anterior pituitary hormone deficiency.

Our case showed isolated hypogonadotropic hypogonadism. The marginally low IGF-1 may reflect a degree of Growth Hormone deficiency but is of doubtful significance in view of the patient's normal growth and absence of other symptoms. This is a rarely reported congenital endocrinological manifestation of EPP with associated dysgenesis of the infundibulum and preserved anterior pituitary on MR imaging.

To conclude, clinical, biochemical and imaging correlations are essential to diagnosis and surveillance of EPP and pituitary infundibular stalk abnormalities. MRI is the imaging of choice to confirm and demonstrate the extent of congenital structural abnormalities of hypothalamo-hypophyseal axis, to establish any association with congenital brain or orbital malformations and to exclude neoplastic and infiltrative diseases which can present with secondary pituitary dysfunction.

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