Ectopic neurohypophysis in a boy: A case report and review

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Abstract

Ectopic neurohypophysis is rare anomaly which is characterized by ectopic location of posterior pituitary lobe (neurohypophysis), pituitary stalk abnormalities and association with dysfunction of anterior lobe related with growth hormone or with multiple dysfunction of the same. We present a rare case of posterior ectopic pituitary and pituitary stalk hypoplasia isolated in 2 year-old male patient.

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Introduction

The ectopic posterior pituitary (EPP) is a disorder that is characterized by ectopic location at the median eminence (floor of the third ventricle). Infundibular stalk anomalies usually include absence, interrupted stalk or hypoplastic stalk as in our case, which is barely visible [1]. Magnetic resonance imaging (MRI) is the key examination as it depicts pituitary anomalies and helps excluding differential diagnoses such as tumors and other infiltrative diseases that can affect the hypothalamo-pituitary axis [2].

Case report

A 2-years-old male patients presented to our hospital with a seizure complicating hypoglycemia. Actual symptoms are manifest in the form of hypotonia and hypoglycemia (the lowest measurement glycemia was 1.7 mmol/L). First time the crisis of hypoglycemia was at 5-months with measurement of 2.2 mmol/L after operation of anal atresia with recto-perianal fistula. Also, this patient has a low growth hormone (1.63 ng/mL) and because of that he had a lower height than the standard for the age (- SD 2.64). The values of laboratory tests were normal for FT4, FT3, and cortisol.

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Physiological examination revealed no other deviations. He had no positive familiarly history.

The unenhanced T1-weighted magnetic resonance images on coronal and sagittal plane (Figs. 1 and 2) through the median eminence revealed a small flattened anterior pituitary lobe and markedly hypoplastic pituitary stalk. The ectopic posterior pituitary appeared as a hyperintense nodule at the median eminence and the infundibulum was seen as a uniform thin hypoplastic structure.

The stalk measured a maximum of 1.9 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 [3]. The T1 Fat Sat-weighted axial images (Fig. 3) demonstrate subtle appearance of posterior pituitary and T2-weighted axial images (Fig. 4) demonstrated normal looking other midline structures and also of without additional abnormality of head and orbits. The corpus callosum, optic nerves, and optic chiasm were a normal appearance.

Due to frequent hypoglycemic crises and lower growth, has been raised suspicion on pathology of the pituitary gland, and an MRI examination is recommended.

**Discussion**

The prevalence of EFP is unknown, but to date is around 1000 cases with, and without the full triad. The pituitary gland consists of anterior lobe (adenohypophysis) and posterior lobe (neurohypophysis). The adenohypophysis is ectodermal in origin and develops from an evagination of the stomodeum (Rathke-cleft). The neurohypophysis is neuroectodermal in origin and formed by the evagination of neural tissue (downward extension of the diencephalon-infundibulum) from the floor of the third ventricle, and on the end of migration it is encapsulated together with the adenohypophysis. The pituitary stalk is a funnel-like structure that is connecting the median eminence of the hypothalamus to the 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
The neurohypophysis is seen as a bright spot on unenhanced T1-weighted imaging, owing the presence of phospholipids which contains the arginine vasopressin neurohypophysis complex which have T1-shortening effect [8]. The characteristic MRI findings of EEP are the presence of a high T1-signal tissue nodule at varying locations, most commonly along the median eminence [9].

Conclusion

MRI is the method of choice for the diagnosis of EEP, extent of congenital structural abnormalities of hypothalamo-hypophyseal axis, and association with brain or orbital malformations.

Patient consent

Patient consent has been obtained.

REFERENCES

[1] Jagtap VS, Acharya SV, Sarathi V, Lila AR, Budyal SR, Kasaliwal R. Ectopic posterior pituitary and stalk abnormality predicts severity and coexisting hormone deficiencies in patients with congenital growth hormone deficiency. Pituitary 2012;15(2):243–50.
[2] Patkar D, Patankar T, Krishnan A, Prasad S, Shah J, Limdi J. MR imaging in children with ectopic pituitary gland and anterior hypopituitarism. J Postgrad Med 1999;45(3):81–3.
[3] Satogami N, Miki Y, Koyama T, Kataoka M, Togashi K. Pituitary stalk: high-resolution MR imaging at 3T. AJNR 2010;31(2):355–9.
[4] Brickmann JM, Clements M, Tyrell R, McNay D, Woods K, Warner J, et al. Molecular effects of novel mutations in Hesx1/HESX1 associated with human pituitary disorders. development 2001;128:5189–99.
[5] Mitchell LA, Thomas PQ, Zacharin MR, Scheffer IE. Ectopic posterior pituitary lobe and periventricular heterotopia: Cerebral malformations with the same underlying mechanism? AJNR Am J Neuroradiol 2002;23:1475–81.
[6] Ullman MC, Seigel SF, Hirsch WL, Finegold DN, Foley TP, Pitmityer Jr. Stalk and ectopic hyperintense T1 signal on magnetic resonance imaging. Am J Dis Child 1993;147:647–525.
[7] Mitchell LA, Thomas PQ, Zacharin MR, Scheffer IE. Ectopic posterior pituitary lobe and periventricular heterotopia: cerebral malformations with the same underlying mechanism? AJNR Am J Neuroradiol 2002;23(9):1475–81.
[8] Kucharezyk J, Kucharezyk W, Berry I. Histochecmical characterization and functional significance of the hyperintense signal on MR images of the posterior pituitary. Am J Neuroradiol 1989;10:1079–83.
[9] BS, El Gamal T, Allison JD, et al. Frequency and variation of the posterior pituitary bright signal on MR images. AJR AM J Roentgenol 1989;153(5):1033–8.