Case 3539

Pituitary stalk interruption syndrome
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Patient: 17 years, female

Clinical History:
A 17-year-old female patient was presented to our hospital with hypopituitarism-related symptoms.

Imaging Findings:
A 17-year-old girl was referred to our hospital for the evaluation of primary amenorrhea, short stature, low body weight and prepuberal appearance. Laboratory tests showed undetectable plasmatic levels of LH and decreased levels of FSH, ACTH (18 pg/ml) and FT4 (6.9 pg/ml) with normal TSH. Blood basal GH and IGF-1 levels were low (0.2 ng/ml and 28 ng/ml, respectively), and GH response to insulin provocation test was impaired (0.3 ng/ml after 30 minutes). Pelvic ultrasound examination showed hypoplasy of uterus and ovaries, while hand X-ray examination revealed delayed bone age (13 years 6 months). Subsequently, a magnetic resonance imaging (MRI) study of the sellar region was performed to rule out anomalies of the hypothalamic-pituitary system. Sagittal (Fig. 1) and coronal (Fig. 2) SE T1-weighted images showed the presence of a small sellar fossa with anterior pituitary smaller than expected according to patient’s age. The pituitary stalk could not be recognized and a high signal spot was located in the median eminence of the infundibulum.

Discussion:
Hypopituitarism is an endocrinological disorder characterized by deficiency of one or more pituitary hormones, and is often associated with a number of structural abnormalities of the hypothalamic-pituitary system. Pituitary stalk interruption syndrome (PSIS) is a rather common cause of hypopituitarism detected on cranial imaging and is defined by absence or marked thinning of the pituitary stalk and ectopia of the posterior pituitary gland, which appears as a high signal spot in the infundibular recess of the third ventricle on MR examination due to its high phospholipid content. As a consequence of hypoplasy of the vascular component of the pituitary stalk, also the anterior pituitary gland is typically hypoplastic. Severity of endocrinological symptoms usually correlates with the degree of pituitary stalk hypoplasy. In general, patients whose pituitary stalk is present, although thin, tend to have a single hormone deficiency (usually GH), while transection of the pituitary stalk is usually associated with multiple hypopituitarism. Thus, follow-up of patients with thin pituitary stalk by cranial imaging is important to monitor pituitary stalk hypoplasy, which can evolve over time leading to worsening of hypopituitarism with onset of multiple hormone deficiency. Pathogenesis of PSIS is unclear. It has been attributed to traumatic or hypoxic-ischaemic injury of the pituitary stalk during delivery, although some authors have reported normal birth history in 54% of patients. In addition, the presence of familial forms of the disease has suggested a genetic origin for PSIS; however, genes potentially implied in hypopituitarism such as GH-N, GHRH-R and Pit-1 are not likely to be involved in the pathogenesis of PSIS. Therefore, the complexity of the embryological development of the hypothalamic-pituitary region, together with its phenotypic heterogeneity, indicates that there may be more than one genetic mutation underlying PSIS. Computed tomography can detect hypoplasy of sella turcica and pituitary gland as well as ectopic
location of posterior pituitary lobe, which appears as an enhancing spot in the infundibular region. However, MRI represents the most important tool for evaluation of patients with pituitary dysfunction, owing to its higher contrast resolution and its ability to acquire data on multiple planes. MRI typically shows a small sella turcica containing little glandular tissue fixed to skull base. On SE T1-weighted sequences the normal posterior lobe hyperintensity is absent in the posterior part of the sellar fossa, and the ectopic posterior pituitary is usually visible as an area of high signal intensity along the median eminence of the hypoplastic pituitary stalk, which is markedly thinned or undetectable. Gadolinium-enhanced, low thickness scans are reported to be even more sensitive than unenhanced sequences in visualizing a severely hypoplastic pituitary stalk, which can help differentiate pituitary stalk thinning from complete transection. Differential diagnosis must be made against surgical or traumatic pituitary stalk transection and infundibular lipoma.

**Differential Diagnosis List:** Pituitary stalk interruption syndrome.

**Final Diagnosis:** Pituitary stalk interruption syndrome.

**References:**


Description: The SE T1-weighted sagittal image showing a shrunken sella turcica and a hypoplastic anterior pituitary lobe. The pituitary stalk is interrupted and characterized by a hyperintense spot in its distal part. Origin:
Figure 2

Description: The SE T1-weighted coronal image showing hypoplasia of sella turcica and anterior pituitary lobe. Origin: