Ectopic neurohypophysis associated with aplasia of the infundibulum

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Section: Neuroradiology
Imaging Technique: MR
Case Type: Clinical Cases
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Patient: 15 years, male

Clinical History:

The patient presented with dwarfism secondary to growth hormone deficiency.

Imaging Findings:

The patient presented with dwarfism secondary to growth hormone deficiency. Hypophyseal MRI was performed on a 1.5T MRI scanner, with SE T1-weighted, FSE T2-weighted, and post-gadolinium SE T1-weighted images in the sagittal and coronal planes. A bright spot representing ectopic neurohypophysis was detected at the undersurface of the hypothalamus-optic chiasm, associated with absence of the pituitary stalk and hypoplasia of the anterior hypophysis within the sella turcica.

Discussion:

The normal neurohypophysis demonstrates high signal intensity on T1-weighted images. T1-shortening of the neurohypophysis reflects enhanced relaxation of water protons in the vicinity of neurosecretory vesicles that function in the storage and secretion of oxytocin and vasopressin. On sagittal MR images the neurohypophyseal focus of T1-shortening lies immediately anterior to the dorsum sella and has been referred to as the posterior pituitary bright spot.

The bright spot can be seen as an ectopic zone of T1-shortening representing the functional neurohypophysis along the floor of the third ventricle; it is most frequently located at the undersurface of the hypothalamus. This bright spot fails to develop in the normal intrasellar location due to impaired formation of the pituitary infundibulum, which normally transmits carrier-bound neuropeptide hormones from the hypothalamus to the neurohypophysis. It is commonly associated with pituitary dwarfism and delayed skeletal maturation. Males are three times more commonly affected than females.

The failure of development of the pituitary infundibulum and hypoplasia of the adenohypophysis may be isolated abnormalities or part of a midline dysgenesis syndrome. The dwarf patients with ectopic neurohypophysis have a greater frequency of multiple pituitary hormone deficiency compared to the dwarf patients with normal neurohypophysis (49% vs 12%), and that of associated congenital brain anomalies (12% vs 7%).

The pathogenesis of ectopic neurohypophysis was explained originally by a traumatic transection of the pituitary stalk during delivery. A high incidence of breech delivery has been reported in these groups, but the traumatic
hypothesis could not explain the findings in the relatively high percentage of patients with normal delivery, nor account for a different feature also found in other pituitary dwarfs consisting of pituitary hypoplasia with normal posterior pituitary. A second hypothesis has then been proposed, based on dysgenesis or abnormal embryonic development of both adenohypophysis and neurohypophysis. The second theory proposes that a defective induction of mediobasal structure of the brain in the early embryo could account for both the complex morphological MRI abnormality and the clinico-endocrinological features encountered in all ectopic neurohypophysis patients. The close contiguity between the future pituitary and hypothalamus, the peculiar association with congenital midline brain anomalies, and the recent data about a possible role of Pit-1 gene, all support the hypothesis of a congenital defect.

**Differential Diagnosis List:** Ectopic neurohypophysis associated with aplasia of the infundibulum

**Final Diagnosis:** Ectopic neurohypophysis associated with aplasia of the infundibulum

**References:**


Description: Spin-echo T1-weighted sagittal image shows a bright spot along the floor of the third ventricle (arrow) associated with the absence of pituitary stalk and hypoplasia of adenohypophysis within the sella turcica. Origin:
Description: Spin-echo T1-weighted coronal image shows a bright spot along the floor of the third ventricle (arrow) associated with the absence of pituitary stalk. Origin: