Pituitary Stalk Interruption Syndrome (PSIS): Specific MRI Findings

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Clinical Image

PSIS is a rare congenital abnormality characterized by a triad of thin or interrupted pituitary stalk, hypoplastic to absent adenohypophysis, and ectopic posterior pituitary gland. It was first described in 1987 by Fujisawa et al and had a reported incidence of 0.5/100 000 births. The mean age of diagnosis is 9.4±11.6 years, with a predilection for males. Mutations in the genes involved in pituitary embryogenesis (PROP1, LHX3, HEXXS1, PROKR2 and GPR161) and perinatal asphyxia are the main influencing factors in this disease. PSIS may present with isolated or multiple anterior pituitary hormone deficiencies. Posterior pituitary function is intact. PSIS are most often associated with several congenital malformations such as septo-optic dysplasia, optic chiasm hypoplasia, holoprosencephaly, etc. MRI shows the characteristic triad of PSIS including the absent of pituitary stalk, anterior pituitary hypoplasia and ectopic posterior pituitary which is mostly located behind optic chiasma or in the hypothalamus (Figure 1).

Keywords: Pituitary; Stalk interruption; MRI

Figure 1: Unenhanced T1-weighted coronal (A) and sagittal (B) Magnetic resonance images show the ectopic location of posterior pituitary, seen as a bright focus (arrows) posterior to optic chiasma with normal intrasellar position of a small and flattened anterior pituitary (red triangle), and the non-visualization of pituitary stalk. Sagittal T1 post contrast image (C) Demonstrates the ectopic location of posterior pituitary and confirm the absence of pituitary stalk.

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