Thickened pituitary stalk with central diabetes insipidus: What Diagnosis?

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Introduction: Central Diabetes Insipidus (DIC) is usually the final result of lesions affecting the hypothalamic-neurohypophysial system, the known causes are: germinoma, craniopharyngioma, histiocytosis, sarcoidosis, familial cause. For the children, lymphocytic infundibuloneurohypophysitis is an uncommon cause, Germinoma is the main reason. The MRI aspect is often limited to thickness pituitary stalk with loss of hyperintensity of the neurohypophysis.

Case report: We presented the case of a 15 years old boy, referred to our clinic for late stature - weight associated with polyuria - polydipsia syndrome appeared six months earlier (estimated 5 litres/day).

Physical exam: Weight = P3, Size < P3, Tanner step I.

He shows signs of growth hormone and corticotropin deficiencies with no intracranial tumor syndrome.

Diagnostic tests:
- Hormonal evaluations demonstrated central diabetes insipidus (low urinary osmolarity and low antidiuretic hormone) with hypopituitarism, hypogonadism and growth hormone deficiency (GHD).

- Pituitary MRI: pituitary stalk enlargement of 6mm, loss of T1 hyperintensity of the posterior pituitary (Fig1, 2).

- Bone - Age = 12 years (AtlasGreuliche and Pyle)
- Assuming a secreting germinoma, rate of BHCG and αfetoprotein were normals.
- Assuming histiocytosis X, an X-ray of the skull was normal.
- Looking for sarcoidosis: normal enzyme conversion, normal thoracic CT scan and biopsy of the salivary glands showed no inflammatory granuloma...

Discussion: thickening of pituitary stalk is suggestive of germinoma, the clinical picture is dominated by a DIC(90%) of early onset, associated to hypopituitarism in 60% of cases: GHD (50 to 100%), Hypothyroidism (0 to 80%), Hypocortisolism (40 to 60%), and early puberty in nearly 50% of cases (secondary to hCG tumoral secretion). The diagnosis is relatively easy if the βhCG rate is high, or if there is a second localization or pineal localization. But in most cases, these tests are normal and the MRI does not differentiate germinoma from other tumors of the suprasellar region, or from other causes thickening pituitary stalk (Langerhans cell histiocytosis, sarcoidosis or lymphocytic hypophysitis). Its signal is homogeneous, isointensity T1, iso- or hyperintensity T2. The pituitary gland is generally normal size, but sometimes can be infiltrated by tumor process. The physiological hyperintensity of the posterior pituitary on T1 is no longer visible.

The meaning of adenohypophysal deficiency associated with Diabetes insipidus controversial.

For some authors, this deficit indicates the presence of a tumor process, for others, the occurrence of anterior pituitary hormone deficiency was noted in 49% of patients with DIC idiopathic without being frequently associated with a process tumor.

Conclusion: MRI report is the diagnosis key of isolated large pituitary stalk, especially when the diagnosis is used as part of other clinical clues; but still a long monitoring each 3-6 months, without histological evidence, can be suggested. Checking for Germinoma or histiocytosis existence, especially in child case.

There are no good imaging predictors for hypopituitarism, making clinical and hormonal evaluation of all patients with pituitary stalk lesion is crucial.

References: