

CHILD WITH GROWTH HORMONE DEFICIENCY DUE TO PERSISTENT CRANIOPHARYNGEAL DUCT

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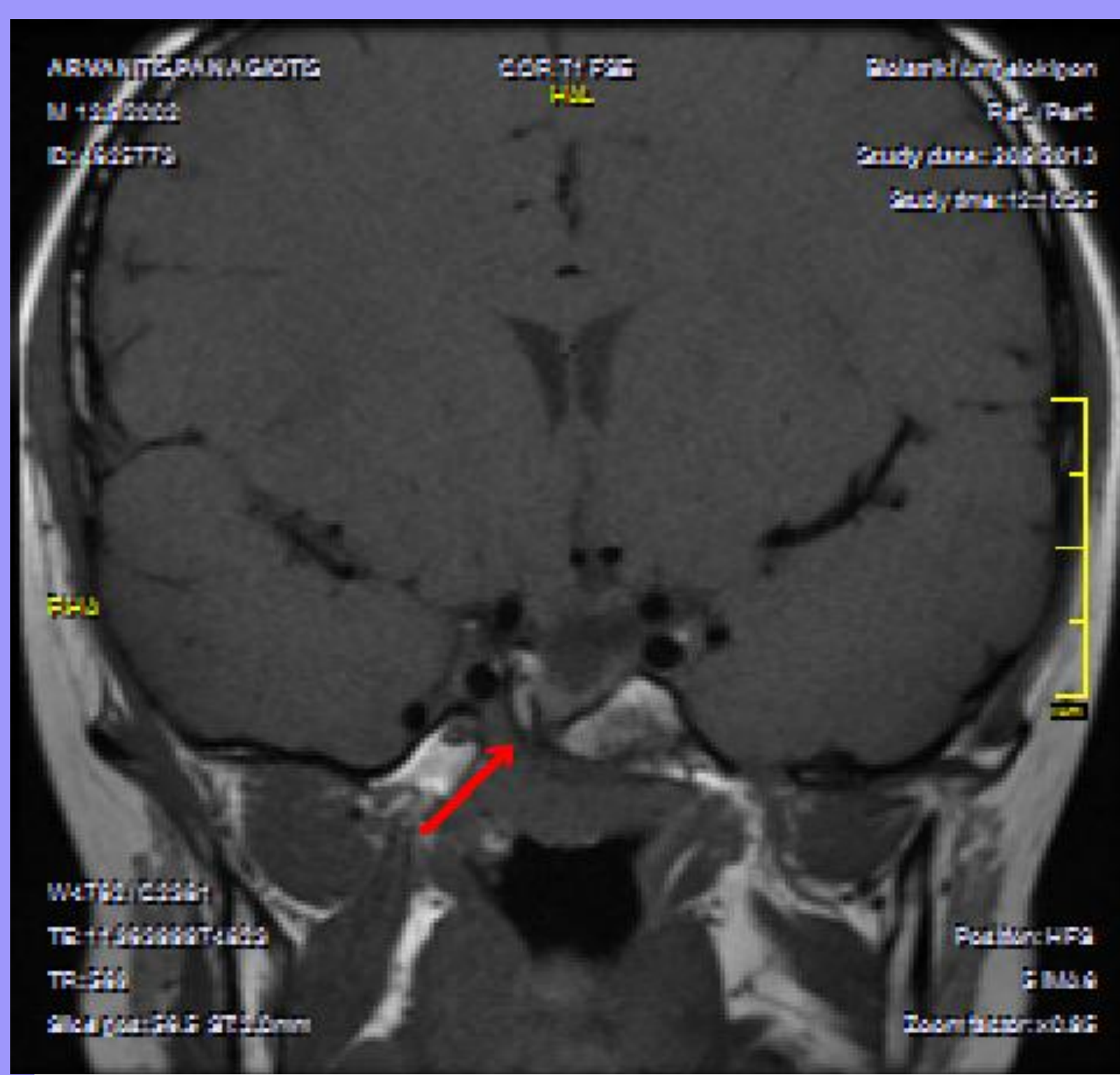
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Introduction

A child with slow growth rate (< -2 SDS) polyuria and polydipsia warrants urgent investigation for hypothalamic-pituitary tumors. The differential diagnosis in this case includes Craniopharyngioma, Histiocytosis, Dysgerminoma, Hypophysitis. We describe the case of a 11 year old boy with growth rate deceleration, polyuria and polydipsia because of persistent craniopharyngeal duct.

Case Report

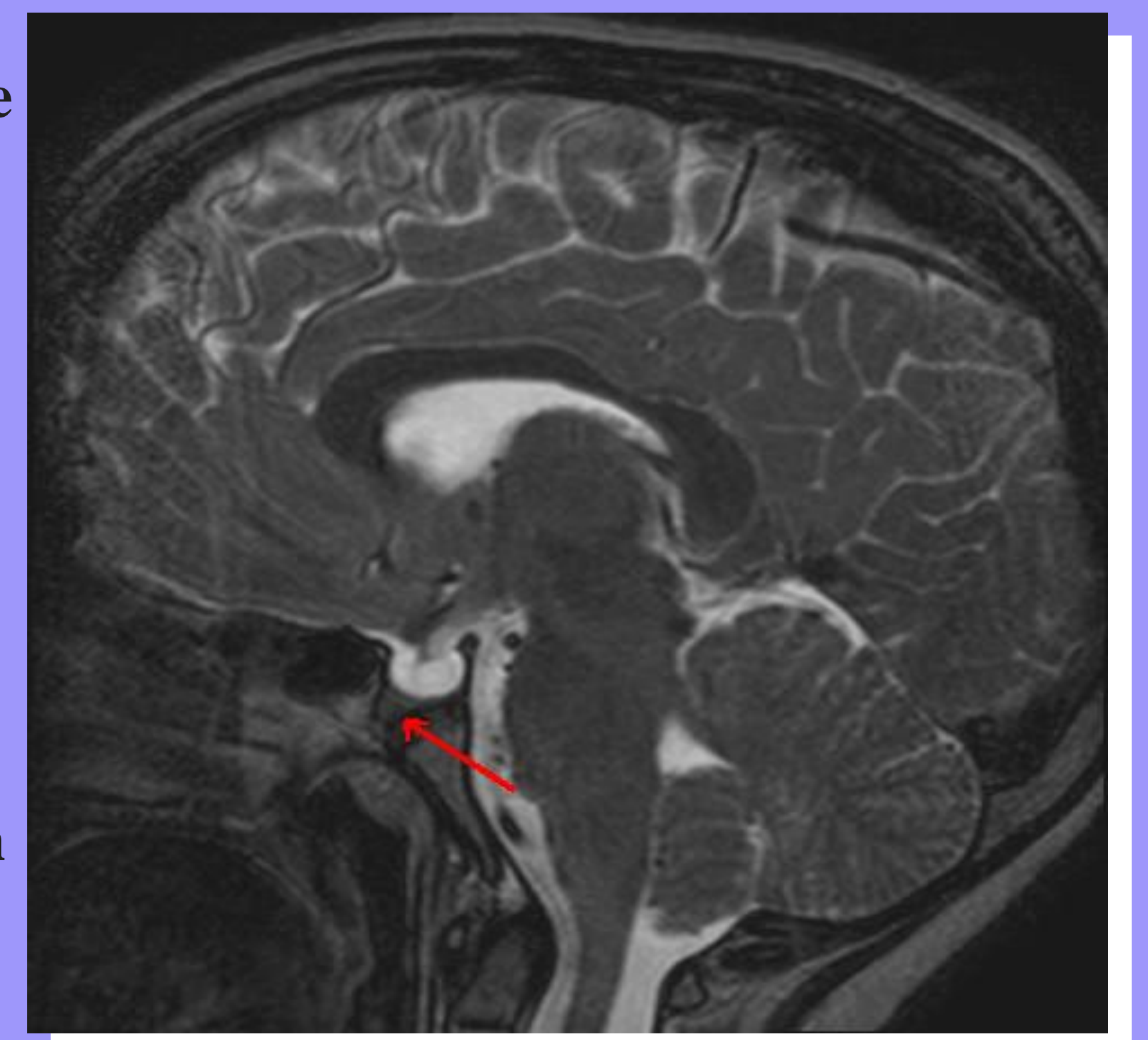
A 11-year-old boy was referred to our department for evaluation of short stature and growth rate retardation with polyuria, polydipsia and nocturia without headache the last 2 years. He mentioned also fatigue and right ear ache three months before admission. He was born by vaginal delivery at term to a non- consanguineous couple with normal perinatal period. His past medical history was unremarkable. *On physical examination*, his height was 132.2cm (3rd %ile), his weight 35.3kg (50th %ile). The parental target height was 181.5cm (50-75 %ile). The boy had no dysmorphic features. His sexual development was at Tanner Stage I. Detailed systemic clinical examinations was within normal range. *Laboratory investigations* showed a normal CBC, liver function test, renal function test. The IGF-I was 115ng/ml , TSH: 4.28 μ IU/ml and FT4:0.72 ng/dl. Bone age was 8.5 years, which was 2.5 years delayed. Functional and imaging investigation of the pituitary, was performed. Growth hormone stimulation tests after clonidine and glucagen were low (hGHmax 1.6ng/ml and 2.2ng/ml respectively) so as the response of cortisol was blunt whereas morning cortisol value was within normal values.



The Magnetic Resonance Imaging of Hypothalamus-Hypophysis showed a bone gap within the sphenoid bone width 2.8mm and length 4mm approximately, corresponding to the plane of sella turcica and immediately to the right of the midline.

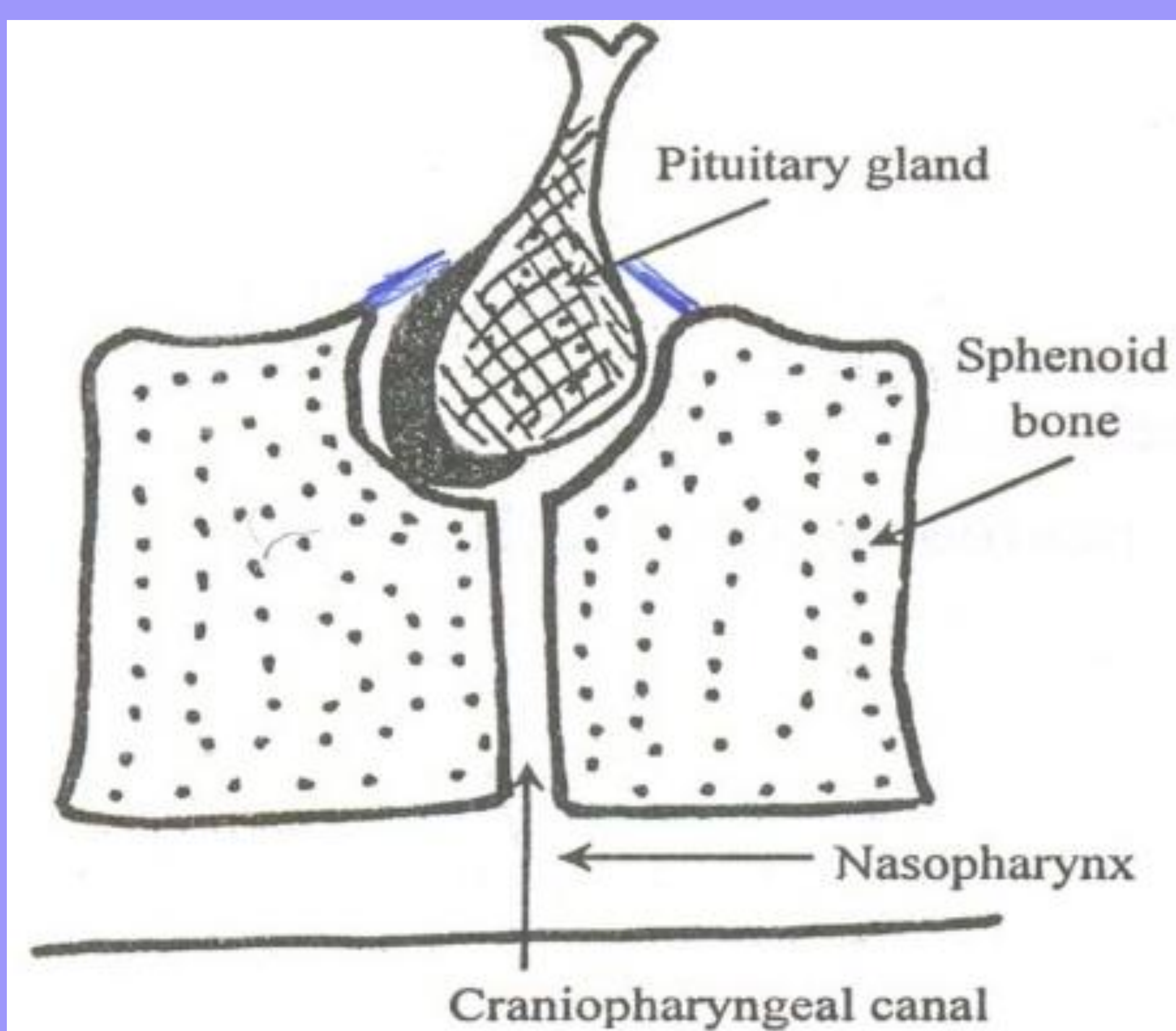
The whole picture advocates for persistent craniopharyngeal duct.

There was denoted a prolapse of the anterior pituitary through the craniopharyngeal duct to the nasopharynx and projection of anterior pituitary of at least the upper limit of the nasopharynx while asymmetric visualization of soft tissue of the nasopharynx with a right ingrowth. The posterior pituitary was not recognised in normal or ectopic position.



The patient was placed on thyroxine and growth hormone substitution therapy.

Discussion



Persistent Craniopharyngeal Duct is a rare basal skull congenital defect of unknown etiology and has a diameter less than 1mm. Craniopharyngeal duct represents a remnant of the stem of Rathke's pouch that goes through the sphenoidal synchondrosis, extending from the floor of the sella turcica to the undersurface of this bone and connecting the pituitary fossa with the nasopharynx cavity.

Canals (ducts) of larger diameter (>1 mm) are even rarer and are referred to as broad craniopharyngeal duct or transphenoidal canal. This may coexist with arachnoid cyst, cystic pituitary adenoma, Rathke's cleft cyst, anencephaly, craniopharyngioma and craniocoele.

It may be complicated by hypopituitarism, cerebrospinal fluid rhinorrhea, meningitis, sinusitis, hydrocephalus, upper airway obstruction in infants, surgical resection of pituitary undetected as nasal polyps.

There are limited references of persistent craniopharyngeal duct in the literature. To our knowledge, there is no reference of persistent craniopharyngeal duct and associated hypopituitarism.

Literature

1. Kizilkilic, et al. Hypothalamic Hamartoma Associated with a Craniopharyngeal Canal AJNR Am J Neuroradiol 2005 January; 26:65-67
2. Gazanfer Ekinci, et al. Transsphenoidal (Large Craniopharyngeal) Canal Associated with a Normally Functioning Pituitary Gland and Nasopharyngeal Extension, Hyperprolactinemia, and Hypothalamic Hamartoma AJR, 2003; 180:76-77
3. Hughes, et al. Persistent hypophyseal (craniopharyngeal) canal The British Journal of Radiology 1999; 72:204-206
4. Currarino G, et al. Transsphenoidal canal (large craniopharyngeal canal) and its pathologic implications Am J Neuroradiol 1985; 6:39-43.