### **Case 17198**



### **Duplication of the pituitary gland**

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Section: Paediatric radiology

Area of Interest: Neuroradiology brain Paediatric

Imaging Technique: CT Imaging Technique: MR Case Type: Clinical Cases

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Patient: 5 years, female

#### **Clinical History:**

A 5-year-old girl was referred to the radiology department for investigation of developmental delay and suspected focal seizures. Dysmorphic features included hypertelorism, flat nasal bridge, bifid tongue, cleft lip and palate. She had mild global developmental delay and at the age of 4, she presented with staring events with abnormal oral movements.

### **Imaging Findings:**

A Magnetic Resonance Imaging (MRI) of the brain was performed which showed a complete duplication of the pituitary gland with two posterior pituitary bright spots and paired infundibula, located in two separated infundibular recesses (Fig. 1). There was thickening of the floor of the third ventricle as a result of fusion of the mammillary bodies and the tuber cinereum (Fig. 2). The olfactory bulbs were absent and there was apparent hypertelorism. In addition, a T1- and T2- heterogeneous, tubular mass was identified in the midline frontal region, above the sphenoid bone (Fig. 3). The susceptibility-weighted imaging demonstrated predominantly chemical shift artefact within the lesion, with some presumed blooming artefact suggestive of calcification.

A Computed Tomography (CT) of the brain confirmed a predominantly fatty mass with a small focus of calcification (Fig. 4). These findings were suggestive of a teratoma. No lesion extension into the nasopharynx was observed. The CT also confirmed hypertelorism and a left-sided cleft palate (Fig. 4).

#### Discussion:

Duplication of the pituitary gland (DPG) is a very rare developmental anomaly with the most recent review reporting 42 cases worldwide [1, 2, 3]. There have been multiple theories proposed in the past to explain the occurrence of DPG. As the notochord induces formation of the pituitary plaque, splitting of the rostral notochord and pre-chordal plate during blastogenesis seems to be the most feasible explanation [1, 3, 4, 5]. The initiating factor for notochordal splitting is unknown although maternal exposure to physical and chemical teratogenic factors or ill health during the sensitive period of organogenesis are considered [5].

The pituitary duplication is almost always associated with other craniofacial developmental abnormalities. Thus, the term DPG-plus syndrome has been proposed by some authors [1, 3]. Pituitary duplication should be considered in all patients who present with midline abnormalities and MRI of the brain is essential for these patients [1, 6]. The most common of these abnormalities are facial dysmorphism such as hypertelorism or lip and palate clefting, bifid tongue, nasopharyngeal mature teratomas, agenesis of the corpus callosum, absence of the olfactory bulbs and abnormalities of the vertebrae. The tubo-mammillary fusion (hypothalamic pseudo-hamartoma) is almost always

present and is postulated to be associated with precocious or delayed puberty. Hence, clinical surveillance of

The management of patients with duplication of the pituitary gland should be multidisciplinary dependent on associated structural abnormalities, like cleft lip and palate, pharyngeal teratoma with or without intracranial extension, abnormalities of pubertal development and intellectual disabilities. Our patient was assessed by geneticists who have advised against further genetic testing as the rarity of this condition means a genetic link is not likely with a normal karyotype and microarray. The 11-month follow-up brain MRI showed a minimal interval increase in the size of the frontal midline mass, otherwise with no significant interval change in appearance.

'Written informed patient consent for publication has been obtained.'

Differential Diagnosis List: Duplication of the pituitary gland

Final Diagnosis: Duplication of the pituitary gland

#### References:

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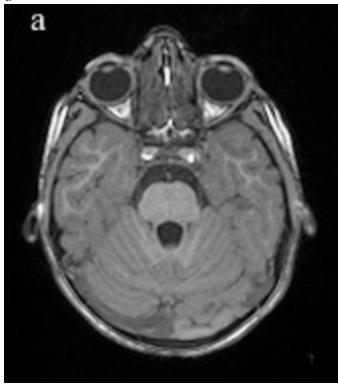
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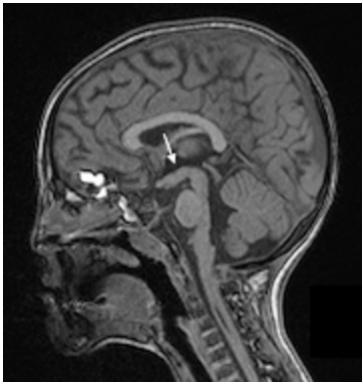


**Description:** MRI Axial and Coronal T1 images showing complete duplication of the pituitary gland with two posterior bright spots **Origin:** Department of Radiology, Children's Health Ireland at Crumlin, Dublin, Ireland, 2019

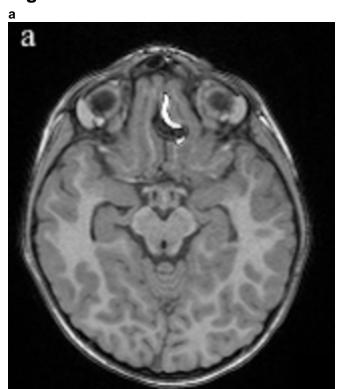


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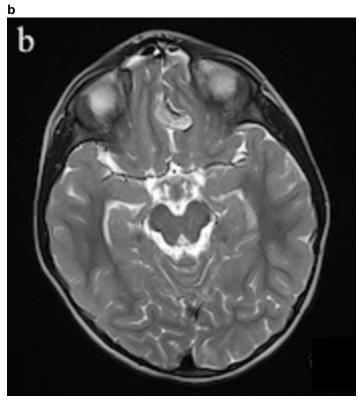
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**Description:** MRI Sagittal T1image showing thickening of the floor of the third ventricle (arrow) in keeping with fusion of the mamillary bodies and tuber cinerum **Origin:** Department of Radiology, Children's Health Ireland at Crumlin, Dublin, Ireland, 2019



**Description:** MRI. a-b Axial T1 and T2 images showing anterior frontal mass which contains hyperintense T1 fatty tissue, c Axial SWI image showing chemical shift artefact as well as possible blooming artefact suggesting additional calcification **Origin:** Department of Radiology, Children's Health Ireland at Crumlin, Dublin, Ireland, 2019



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