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Short Communication

Kallmann Morsier Syndrome: Anosmia as a Revealing Sign

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Abstract

We present the case of a 22-year-old patient who presented amenorrhea with congenital anosmia related to agenesis of the olfactory bulbs on the MRI defining Kallman Morsier syndrome.

Keywords: *Kallman Morsier Syndrome; Anosmia; MRI*

Observation

We report the case of a single patient aged 22-year-old, without significant medical history, admitted to the endocrinology department for exploration of primary amenorrhea associated with a notion of anosmia since childhood and in whom the clinical examination had found a thin patient (BMI 16.8 kg/m^2), pre-pubertal: stage II breasts, Tanner stage III pubic hair and sparse axillary hair. Biologically, hypogonadotropic hypogonadism was observed with an FSH level = 0.80 mIU/ml (2 - 10), LH = 0.09 mIU/ml (0.5 - 5), estradiol < 10 pg/ml.

The patient was referred to our radiology department for a hypothalamic-pituitary MRI to explore the hypothalamic-pituitary axis. MRI revealed agenesis of the olfactory bulbs (Figure 1). Exploration of the other hypothalamic-pituitary axes did not show any abnormality.

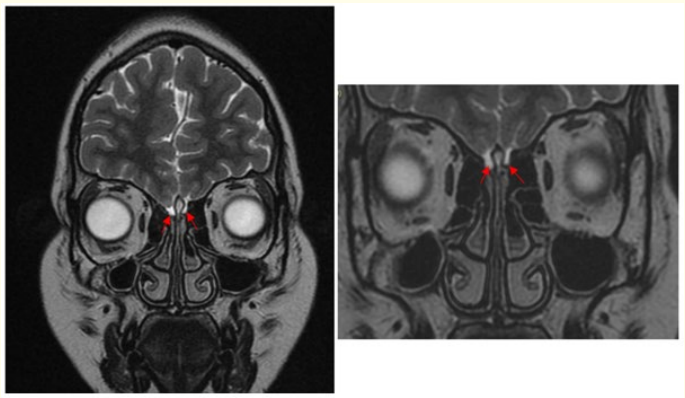

Figure 1: *Coronal section of a T2 brain MRI with magnification on the left showing agenesis of the olfactory bulbs in a 22-year-old female patient. The olfactory sulcus, rectus gyrus, and medial orbital gyrus are normal.*

Diagnostic

Kallmann Morsier Syndrome.

Comment

Kallmann syndrome is a rare genetic disorder, initially described by Maestre de San Juan in 1856. It is characterized by a combination of endocrine disorders, including hypogonadotropic hypogonadism, and abnormalities in the development of the olfactory system. This results in olfactory dysfunction, such as anosmia or hyposmia, attributable to abnormal migration of olfactory axons and neurons producing gonadotropin-releasing hormone. These symptoms constitute the main manifestations of Kallmann syndrome [1]. KS is a genetically heterogeneous disease with an incidence of 1 in 8000 to 30,000 in men and 1 in 40,000 to 120,000 in women. The estimated incidence of KS in men is 4 to 5 times higher than in women, and only 30% of cases have an identifiable genetic cause [2].

Brain MRI is the test of choice to confirm the diagnosis of Kallmann-Morsier syndrome. It is recommended to perform high-resolution coronal fast spin echo weighted T2 and T1 images with a slice thickness not exceeding 3 mm for detailed morphological analysis of the olfactory system. It often reveals an absence or hypoplasia of the furrows and olfactory bulbs. Hypoplasia of the anterior pituitary gland may also be observed [1,2]. The studies by Koenigkam-Santos., *et al*. confirmed that aplasia of the olfactory bulbs and furrows was frequently observed in patients with KS, demonstrating significant concordance with clinical findings [3].

Hormonal treatment in Kallman syndrome is primarily aimed at restoring normal pubertal development and, in some cases, restoring fertility. This involves the administration of exogenous sex steroids tailored to the patient's gender, accompanied by precise stimulation of gonadotropin-releasing hormone. although these results may vary from one individual to another [4].

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