**Hypophyseal Triplication: Case Report and Embryologic Considerations**

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CASE REPORT

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Summary: Hypophyseal triplication is a malformation that has not been described previously. We present a child with midline abnormalities who underwent epignathus excision at birth. Brain MR imaging revealed 2 paired lateral pituitary glands and an oval midline gland, each with an independent stalk, connected to a thickened third ventricle floor. Because malformations represent a failure in embryogenesis, this case may provide interesting clues on the normal development of the hypophysis.

Duplication of the pituitary gland is an extremely rare malformation; most patients do not survive beyond infancy because of the severity of associated abnormalities. Several cases present epipharyngeal teratoma, midline palatal cleft, or spinal malformations, thus revealing the failure of development of multiple midline structures. Besides the classic form with 2 complete pituitary glands located in the lateral recesses of the pituitary fossa, the spectrum of hypophyseal malformation includes incomplete duplication affecting the anterior pituitary gland or the pituitary stalk. The floor of the third ventricle usually appears thickened, and this feature has been differently interpreted as a hamartoma or as the result of the duplication of the hypothalamic nuclei, or the fusion of the mammillary bodies and tuber cinereum into a single midline mass.

We report the MR imaging findings of an hypophyseal triplication in a 7-year-old girl with concomitant craniofacial and spinal abnormalities. Because the embryogenesis of the pituitary gland is still controversial, morphologic and signal intensity characteristics of the 3 pituitary glands have been considered to gain insight into the normal development of the hypophyseal-hypothalamic axis.

Case Report

A 7-year-old first child of nonconsanguineous parents, with a normal female karyotype and a known occult spinal dysraphism, was referred to our center for worsening of scoliosis and the onset of micturition.

According to her clinical history, she was born at 36 weeks’ gestation by cesarean delivery because of a massive oropharyngeal epignathus detected by sonography examination at the fifth month of gestation. At birth, to safely manage airway obstruction, an ex utero intrapartum treatment (EXIT) procedure was performed, which consisted of maintaining placental support until the airway was secured. Fifteen days later, the protruding epignathus teratoma was removed, while a concomitant midline palate and lip cleft was repaired by serial maxillofacial surgery.

In our center, the patient underwent an MR imaging examination of the lumbosacral spine and the brain. The spinal MR imaging confirmed sacral spina bifida with butterfly vertebra at S1 and hemispndyls in the inferior part of the sacrum. Moreover, an intradural li-

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of the pituitary gland. According to the classic theory, the hypophysis is believed to arise from 2 distinct structures, namely the diencephalic neuroectoderm (posterior lobe) and the oral ectoderm (anterior lobe). At approximately 42 days of gestation, a diverticulum originating from the stomodeum fuses with the downward extending diencephalic bud, both finally housed in the pituitary fossa. In contrast to the traditional view, Gilbert (1934) found that in mammalian species, both the anterior and posterior part of the gland originate from the ventral neural ridge. In this model, resumed by Morton in 1957 in a case report of pituitary duplication, the gland is supposed to arise entirely from neuroectodermal tissue without the primary involvement of the Rathke pouch. These 2 theories primarily differ in the role of the Rathke pouch, which seems necessary for development of the adenohypophysis in the former, whereas it is not required in the latter. When a midline cleft is present, the Rathke pouch can reach the above developing gland only aside the cleft itself. In our case, both the lateral glands presented signal intensity characteristics consistent with the coexistence of neurohypophyseal and adenohypophyseal components (Figs 1A, 1C, 2A, 2C). It is interesting to note that the third gland, situated above the midline cleft, presented a homogeneous MR signal intensity, isointense to the hypothalamic floor, consistent with the presence of the neurohypophyseal component only (Figs 1B, 2B). According to Gilbert’s theory, there should be no reason for the absence of both neurohypophyseal and adenohypophyseal components in the midline gland because the Rathke pouch does not contribute to the development of any of them. Hence, the coincidence of 2 “normal” paired lateral pituitary glands with a third “incomplete” hypophysis, just above a midline cleft hampering the normal development of the Rathke pouch, strongly suggests its pivotal role in the normal development of the hypothalamic-pituitary axis.

In conclusion, we present a patient affected by multiple midline defects with triplication of the pituitary gland. This extraordinary condition seems to support the hypothesis of a Rathke pouch contribution to the genesis of the pituitary gland, thus contrasting the alternative hypothesis of an entirely neuroectodermal origin of the hypothalamic-pituitary axis.

References