Association of pituicytoma and Cushing disease: a rare pediatric case

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6-year-old girl with growth failure and associated weight gain, premature pubarche, hypertrichosis.

24-h UFC: 897.6 μg/24 h (NV <100)
Midnight serum cortisol 24.26 μg/dl and ACTH 33.7 pg/ml
Overnight-1 mg DST: serum cortisol 3.89 μg/dl

CUSHING SYNDROME

ACTH SECRETING ADENOMA

BILATERAL SIMULTANEOUS INFERIOR PETROSAL SINUS SAMPLING:
inter-petrosal sinus gradient <1.4, suggestive of a midline lesion

PITUITARY ADENOMA (WHO grade I)

Endoscopic transsphenoidal excision of a small, soft mass.

Hydrodynamic MRI: Convex upper surface, with focal T2-weighted hyperintensity in the middle region.

PITUITARY MRI: Convex upper surface, with focal T2-weighted hyperintensity in the middle region.

Persistence of hypercortisolism

2nd transsphenoidal operation

HYSTOLOGY:
Round and elongated bipolar spindle-shaped neuroglial cells. Very low mitotic activity (0-1%). Some tumoral cells express GFAP.

PITUICYTOMA (WHO grade I)

HYSTOLOGY:
Intraglandular focal area of low signal intensity in the tardive phase, located in the upper-middle region of adenohypophysis.

Pituitary dynamic MRI: Intraglandular focal area of low signal intensity in the tardive phase, located in the upper-middle region of adenohypophysis.

Pituicytoma is a very rare low-grade glioma that originates in the neurohypophysis and infundibulum, usually causing visual defects and sometimes pituitary hormone deficiency. To date only 3 cases have been described in childhood; this is the youngest patient with this lesion. Just one case of coincidence of pituicytoma and ACTH-secreting adenoma has been reported in an adult man. This association is difficult to explain due to the different embryological origin of these two benign tumors.

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