Association of pituicytoma and Cushing disease: a rare pediatric case

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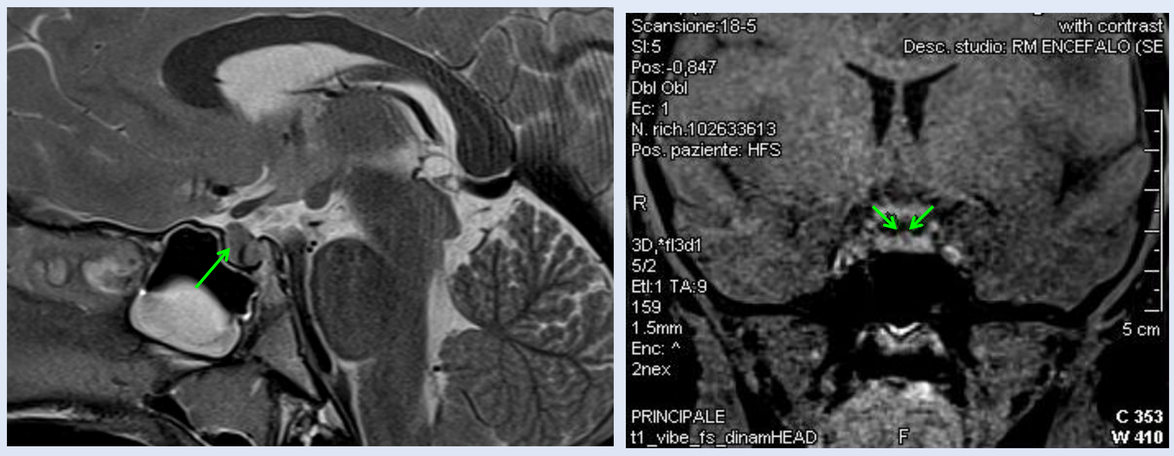
6-year-old girl with growth failure and associated weight gain, premature pubarche, hypertrichosis.

24-h UFC: **897.6 µg/24 h** *(NV <100)*
Midnight serum cortisol **24.26 µg/dl** *and ACTH* **33.7 pg/ml**
Overnigth-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**

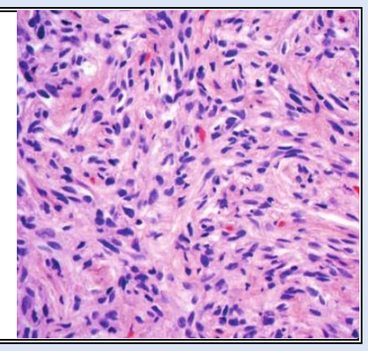


***Pituitary MRI**: Convex upper surface, with focal T2-weighted hyperintensity in the middle region.*

Endoscopic transsphenoidal excision of a small, soft mass.

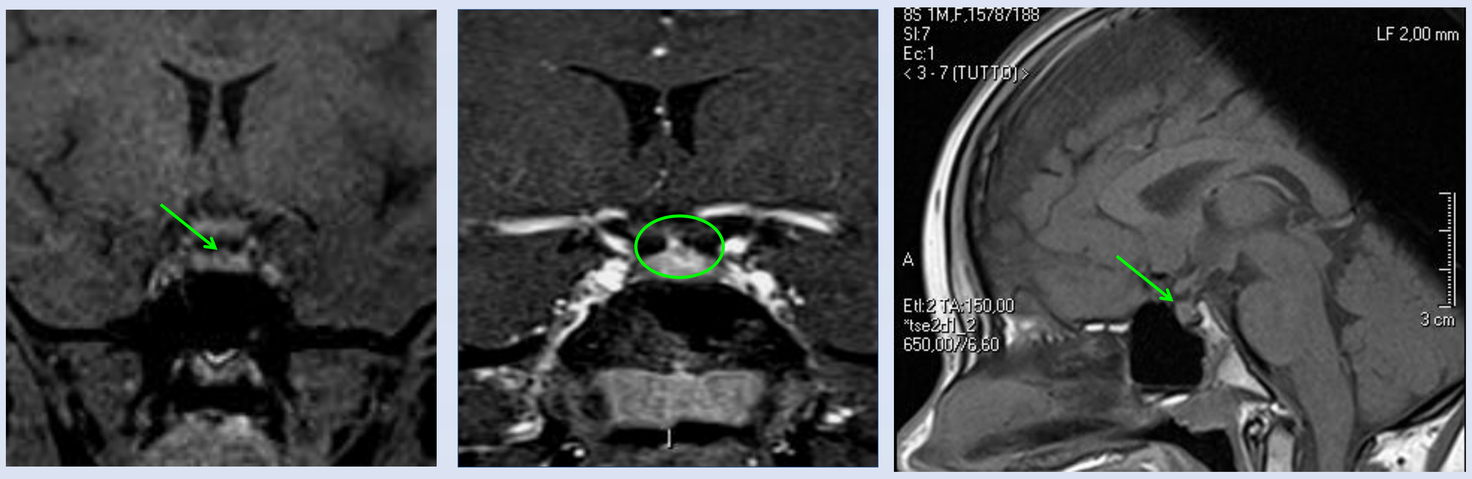
HYSTOLOGY:
Round and elongated bipolar spindle-shaped neuroglial cells. Very low mitotic activity (0-1%). Some tumoral cells express GFAP.

PITUICYTOMA *(WHO grade I)*



Persistence of hypercortisolism

2nd transsphenoidal operation



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

HYSTOLOGY:
PITUITARY ADENOMA *(WHO grade I)*

***IMMUNOPHENOTYPE*:**
ACTH+, **GH+**, LH-, FSH-, PRL-/+, TSH-
Growth fraction (MIB1) <1%

Pituicytoma is a very rare low-grade glioma that originates in the neurohypophisis 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.