

TABLES

Table 1: Clinical, hormonal, and radiological characteristics; treatment; follow-up and results of the genetic study and family history of endocrine neoplasia characteristics of 7 pediatric patients with pituitary macroadenomas.

Patient	1	2	3	4	5	6	7
Sex	Male	Female	Male	Male	Male	Male	Male
Age at diagnosis (years)	15	15	14	8	10	14	15
Anthropometry	W: 41.5kg (-2.46SDS) L: 154.2cm (-1.5SDS) IMC: 17.45 kg/m2 (-1.29SDS)	W: 63kg (+1.36SDS) L: 152cm (-1.7SDS) IMC: 27.2 kg/m2 (-1.5SDS)	W: 40.7kg (-1.59SDS) L: 151.3cm (-1.68SDS) IMC: 17.8 kg/m2 (-0.92SDS)	W: 36.8kg (-1.63SDS) L: 152.1cm (+4.3SDS) IMC: 15.9 kg/m2 (-0.39SDS)	W: 60kg (+2.9SDS) L: 160cm (+3.3SDS) IMC: 23.4kg/m2 (+1.58SDS)	W: 70kg (+0.99SDS) L: 180cm (+1.8SDS) IMC: 21.6 kg/m2 (-0.09SDS)	W: 60kg (-0.14SDS) L: 160cm (-1.1SDS) IMC: 23.4 kg/m2 (+0.57SDS)
Clinical symptoms	Growth retardation, Galactorrhea	Secondary Amenorrhea	Growth retardation	Gigantism	Gigantism	Headache	Polyuria, polydipsia 3L/day
Tanner at diagnosis	P3G3, 12/12cc	S5P5	P3G3, 15/15cc	P1G1, 3/3	P1G1 3/3	G3P3, 15/15	G3P3, 15/15
Visual Defects	Yes	No	No	No	Yes	No	Yes
MRI	Adenoma (15mm)	Adenoma (29mm)	Adenoma (11mm)	Adenoma (16mm)	Adenoma with suprasellar extension (40mm) and cavernous sinus invasion	Adenoma with suprasellar extension (20mm)	Adenoma with suprasellar extension (33mm) and cavernous sinus invasion
Elevated pituitary hormone	Prolactin (583ng/ml)	Prolactin (>1000ng/ml)	Prolactin (70ng/ml)	IGF1 631ng/ml (+3.52 SDS)	IGF1 1115ng/ml (+5 SDS)	No	FSH 30U/l (nv: 0.16-3.33 Ui/L)
Adenoma Type	Prolactin secreting PA	Prolactin secreting PA	Prolactin secreting PA	GH-secreting PA	GH-secreting PA	Non-secreting PA	FSH-secreting PA
Other pituitary hormone defects	IGF-I: (154ng/ml (-1.1 SDS) testosterone: 34ng/dl (nv:300+/- 22ng/dl)	No	IGF-I: 181ng/ml (-0.6 SDS); testosterone: 154 ng/ml (nv:300+/- 22ng/dl)	No	No	No	testosterone: 10ng/dl (nv:300+/- 22ng/dl), cortisol: <5ug/dl (nv: 16.3 +/- 6ug/dl), TSH <0.1U/l (nv: 1.9 +/- 1.28U/l)
Transsphenoidal surgery	No	No	No	Yes	Yes	Yes	Yes
Pathological anatomy	-	-	-	Hormonal immunohistochemical expression and transcription factors compatible with sparsely granulated somatotrophic adenoma, with a prolactin expression focus.	Somatotrophic pituitary adenoma.	Adenoma with negative immunoreactivity to all pituitary hormones.	Adenoma with immunoreactivity to FSH with a 10-20% cell proliferation index and P53 negative. At reintervention same conclusion.
Radiotherapy	No	No	No	No	Yes	Yes	Yes
Medical treatment	Cabergoline 3mg/week	Cabergoline 1mg/week	Cabergoline 1mg/week	No	No	No	No
Complications	No	No	No	ACTH deficiency	Secondary panhypopituitarism	Secondary panhypopituitarism	Secondary panhypopituitarism
Follow up	3 months MRI tumor reduction (19mm) Prolactin 116ng/ml	2 years MRI tumor reduction (4mm) Prolactin 14ng/ml Cabergoline 0.5mg/week	3 months MRI tumor stabilization (7mm) Prolactin 10.9ng/ml Testosterone: 451ng/ml.	6 months IGF1 110ng/ml (-0.16DS) MRI secondary changes to surgery	10 years IGF-I: 107ng/ml(-1SDS) MRI tumor remnants persist	10 years MRI tumor remnants persist	10 years MRI tumor remnants persist
Genetics	In progress	<i>MEN1</i> Heterozygous change c.249_252delGTCT; p(Ile85Serfs*33	Negative: <i>NF1</i> , <i>AIP</i> , <i>CDKN1B</i> , <i>CDC73</i> , <i>MEN1</i> , <i>RET</i> ,	<i>AIP</i> heterozygous change c.811C>T; pArg271Trp	<i>AIP</i> heterozygous change c.811C>T; pArg271Trp	Unrealized	Unrealized

			<i>PRKARIA, BRCA1, BECA2, MLH1, MSH2, MSH6.</i>				
Secondary tumors	No	Parathyroid adenoma	No	No	No	No	No
Family history	No	Sister with parathyroid adenoma and same pathogenic variant	No	Mother has same pathogenic variant without clinical manifestations	Mother and brother have same pathogenic variant without clinical manifestations. Maternal aunt and cousin affected by pituitary adenomas with the same pathogenic variant.	No	No

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