



PITUITARY STALK INTERRUPTION SYNDROME: a case of an infant

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BACKGROUND

PSIS

- ✓ Interrupted pituitary stalk
- ✓ Anterior pituitary hypoplasia/aplasia
- ✓ Absent/ ectopic posterior pituitary

- ✓ Hypothalamic hypothyroidism
- ✓ Hyperprolactinemia
- ✓ Deficiency of anterior pituitary hormones

Clinical presentation varies according to age of diagnosis

CASE

COMPLAINT

Micropenis

HISTORY: He was born in term via C/S. Birth weight was 3300 gr. He had been followed up for hypoglycemia and jaundice in newborn period and had no known disorder. There was no similar disorder in his family or consanguinity between parents

PHYSICAL EXAM

Cronological Age: 5.5 months

Weight: 7740gr (25-50p) (-0.39 SD)

Height: 67.5cm (25-50p) (-0.22 SD)

SPL: 2.5 cm (<10p) Tanner stage-1

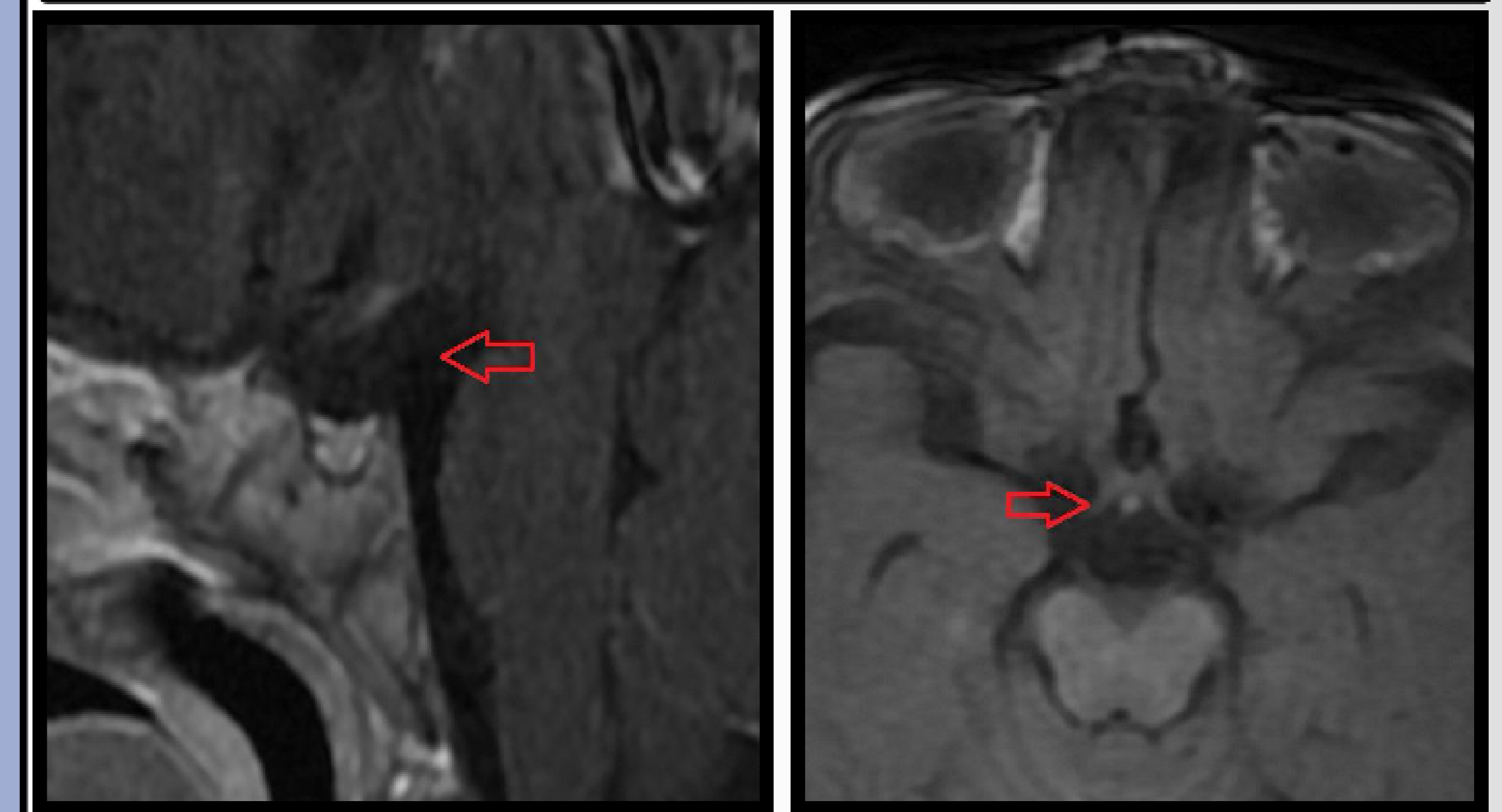
System examinations were normal

LABORATORY

- ✓ Glucose: 87 mg/dl
- ✓ Na: 139 mEq/L
- ✓ K: 5.6 mEq/L
- ✓ Urine density: 1025
- ✓ ACTH: 11.5 pg/ml
- ✓ Cortisol: 1.27 mcg/dl
- ✓ TSH: 8.96 mIU/ml
- ✓ ft4: 0.7 ng/dl
- ✓ ft3: 3.45 pg/ml
- ✓ LH: 1.5 mIU/ml
- ✓ FSH: 0.9 mIU/ml
- ✓ T. testosteron: 2.5 ng/dl
- ✓ Prolactin: 49.01 ng/ml
- ✓ IGF-1 < 25 ng/ml
- ✓ IGFBP-3: 1.13 ug/ml

Minute	CORTISOL (µg/dl)	Minute	TSH (mIU/ml)	ft4 (ng/dl)	Prolactin (ng/ml)	Minute	LH (mIU/ml)	FSH (mIU/ml)
0.	0.85	0.	4.85	0.88	41.6	0.	0.59	0.46
30.	4.88	15.	27.08	-	-	15.	2.22	0.79
		30.	29.63	-	-	30.	2.74	0.97
		45.	29.23	-	-	45.	2.85	1.21
		60.	29.75	-	-	60.	2.88	1.34
		90.	33.25	-	56.8	90.	2.63	1.44

Pituitary gland MRI



Cranial MRI: Normal

RADIOLOGY

Clinical course: Low dose ACTH stimulating test found to be concordant to central adrenal insufficiency, so as TRH test to hypothalamic deficiency. Hydrocortison, L-tyroxin and Dehydrotestosteron gel treatment started. In the last control, age: 1 year, Weight: 9.3 kg(25p)(-0.8 SD), Height: 75 cm (25-50p)(-0.5 SD), SPL 4.5 cm, Puberty Tanner Stage 1, growth rate: 7.5 cm/7 month. In laboratory, glucose, plasma electrolytes were normal, and patient was euthyroid. Patient who use to receive hydrocortison 15/mg/m2/day, L-tyroxin 2.7mcg/kg/day is planned to undergo growth hormone stimulating test when the growing slow down.

DISCUSSION

- Male/Female 2.3-6.9/1
- Mean age of diagnosis 9.4 11.6 years
- Heterogeneous clinical findings, most frequent is short stature (85.5%)
- 100% GH, 95.8% gonadotrophins, 81.8% corticotropin, 76.3% tyrotropin deficiency
- Hyperprolactinemia 36.4%
- Patogenesis ?
- Birth trauma, neonatal hypoksemia
- HESX1, LHX4, SOX3, OTX2, PROKR2, GPR161 mutations

CONCLUSION

The early diagnosis of anterior pituitary deficiency is important to avoid from possible mortality and morbidity.

