Functional gonadotrophin axis evident as spontaneous puberty in a pediatric patient with hypopituitarism after craniopharyngioma resection: a case report

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OBJECTIVES

To report a case of hypogonadism reversal and spontaneous puberty in a craniopharyngioma patient who suffered from hypopituitarism as a result of complete surgical resection.

Figure 1. Growth chart of the child before and after surgery and rGH treatment

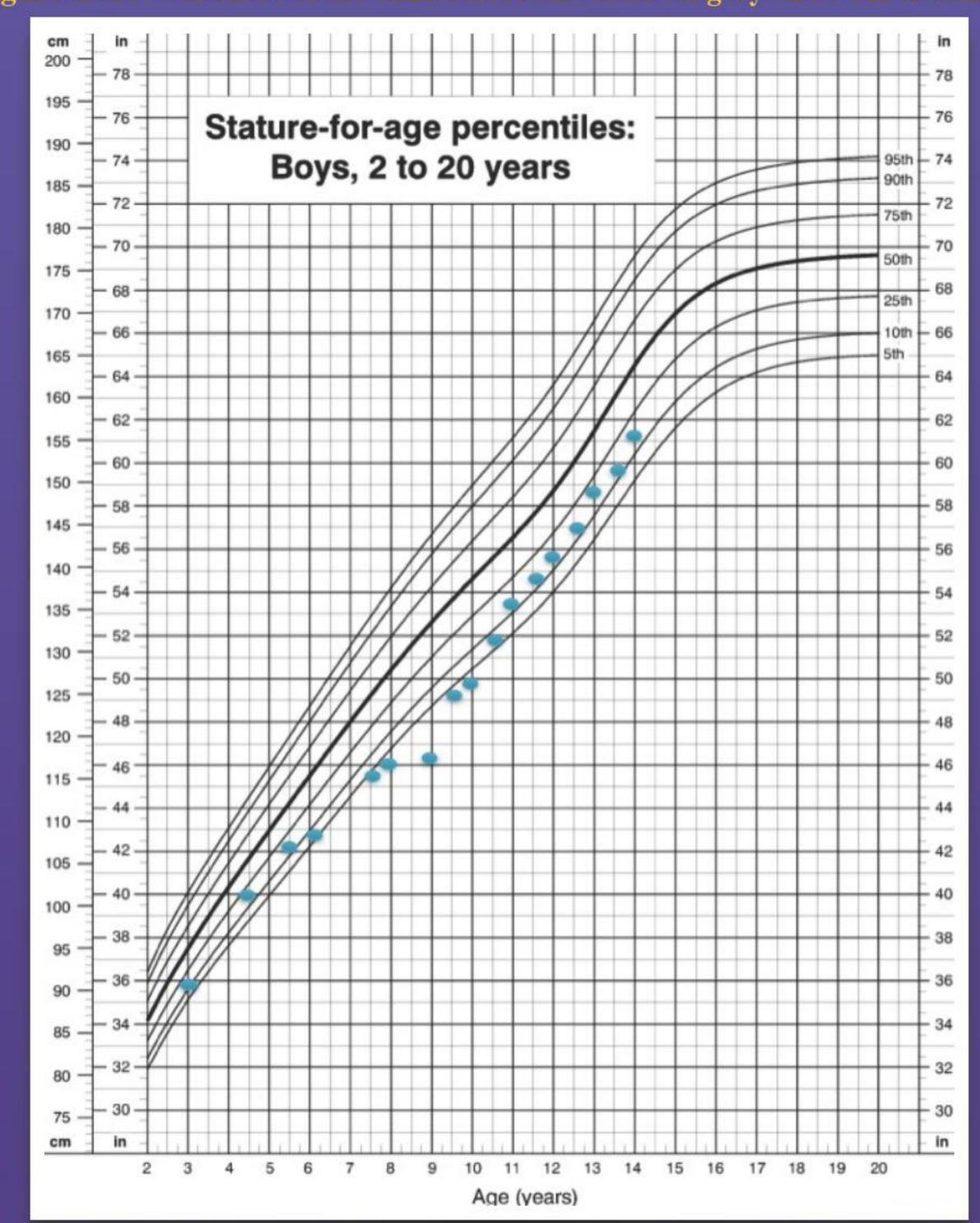


Table 1. Gonadotropins from 6 years old until puberty

	6 years	13 years	13.5 years	14 years
FSH (TU/L)	0.3		1.1	1.4
LH (IU/L)	0.04		0.9	1.8
Testosterone (ng/dl)	17		10	98
Inhibin-B (pg/ml)				216
Testicular volume (ml)		<4	10-12	

Table 2. GnRH stimulation test

GnRH stimulation test	FSH (TU/L) time 0 min	LH (TU/L) time 0 min	FSH (IU/L) time 20 min	LH (IU/L) time 20 min
13.5 years	1.1	0.9	6.3	4.2
14 years	1.4	1.8	12.1	17.2

METHODS

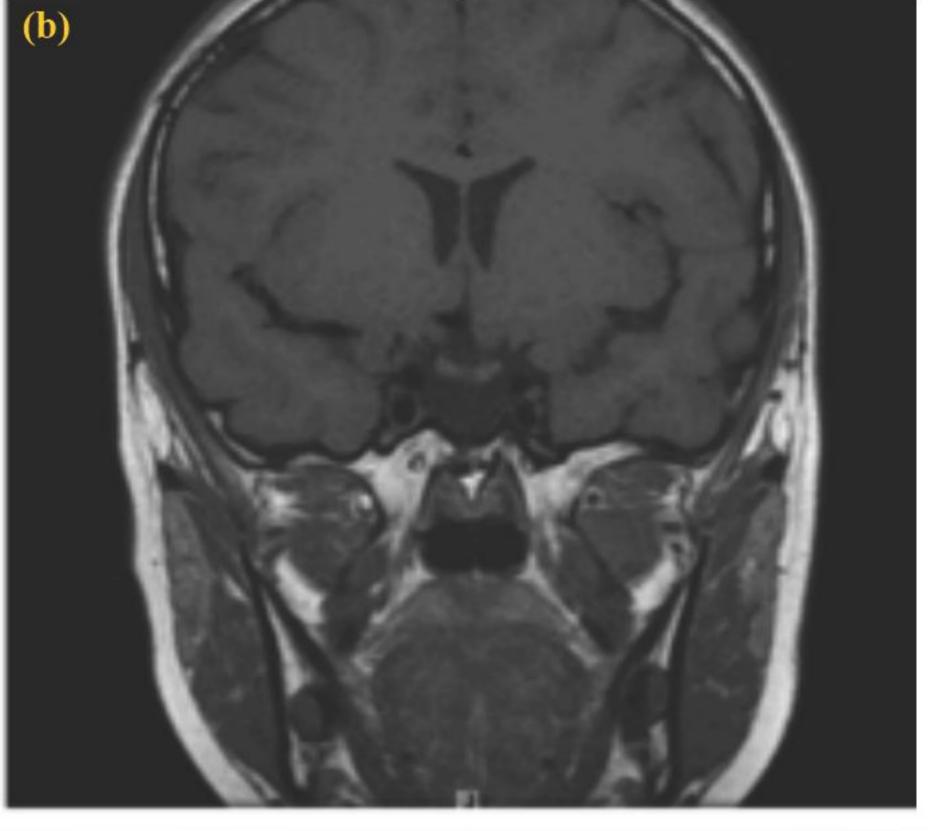
A 13-year-old boy with hypopituitarism was evaluated for right testicular sensitivity. At the age of 6 the child presented with growth arrest and a craniopharyngioma causing pituitary insufficiency was diagnosed. Treatment with hydrocortisnone and thyroxine supplementation was initiated and he was subjected to a transphenoidal surgery that resulted in complete tumor resection (Figure 2). Tumor histology indicated an adamantinous craniopharyngioma, which constitutes the predominant type occurring in children and adolescents. Postsurgical biochemical testing confirmed panhypopituitarism and he was prescribed levothyroxine, hydrocortisone and desmopressin supplementation. Recombinant growth hormone was initiated 2 years following surgery, since post surgical MRI did not depict any sign of pituitary tissue and growth had not resumed (Figure 1). Gonadotropins had been undetectable since diagnosis and the last evaluation had confirmed prepubertal status (Table 1). On clinical examination increased testicular volume was found (10-12ml) with absence of pubic hair and mild sensitivity was confirmed. The Scrotum ultrasound revealed normal appearing testes with a maximal diameter of 4.5cm left and 4.1cm right. Testosterone and adrenal androgen levels were below normal limits but a GnRH stimulation test (2.5µ/kg) provoked a fourfold increase of LH (Table 1,2). Onset of puberty was suspected.

RESULTS

Six months later, the onset of puberty was confirmed, by measurement of gonadotropins (FSH = 1,4 U/l, LH= 1,8 U/l), testosterone (98 ng/dl) and inhibin-B (216 pg/ml). The GnRH stimulation test provoked a tenfold increase in LH. The results of biochemical testing combined with the testicular volume, compared to normative values, indicated the onset of puberty in our patient (Table 1,2). The patient's treatment regarding his other hormonal deficits was continued. However, the testes enlargement together with the LH increase suggested a possible restoration of the gonadal axis, thus rendering testosterone supplementation unnecessary. Proper follow-up was scheduled to evaluate adequacy of gonadal steroid production.

Figure 2. Initial (a) and postsurgery (b) MRI images of the tumor and the pituitary area





CONCLUSIONS

Recovery of pituitary function in craniopharyngioma patients who had panhypopituitarism both prior and after complete surgical resection of the tumor has only rarely been reported. Nevertheless, this case depicts that one should consider the possibility of a functional gonadal axis prior to steroid supplementation for puberty induction to children suffering from hypopituitarism due to craniopharyngioma diagnosed and treated before the onset of puberty.

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