

Gigantism Secondary to Growth Hormone Secreting Pituitary Microadenoma

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Background

Pituitary gigantism is an extremely rare disorder characterised by growth hormone (GH) excess that occurs before fusion of the epiphyseal growth plates. It is usually caused by a pituitary adenoma, with an estimated incidence of 3 cases per million. About 4-5% of pituitary adenomas occur in a familial setting.

Presenting History

A 13 years 6 month old Chinese boy presented to the paediatric endocrine outpatient clinic with tall stature. He was noted by his parents to have a continued growth spurt since 9 years old, and he required new shoes and clothes every school term. He had no headaches or visual disturbances. There is no family history of tall stature (Figure 1)

On examination, his height measured 191.3 cm (> 2 SD above the 97th percentile) and his weight measured 66.0 kg (90th-97th percentile). He had no gynaecomastia or features of Marfan syndrome. There was mild prognathism. He had Tanner Stage 4 for pubic hair and his testicular volumes measured 15 mls bilaterally. His bone age was 14 years, with a predicted height of 206.0 cm (well above the upper limit of his target range which was 174.0-189.0 cm).



Fugure 1

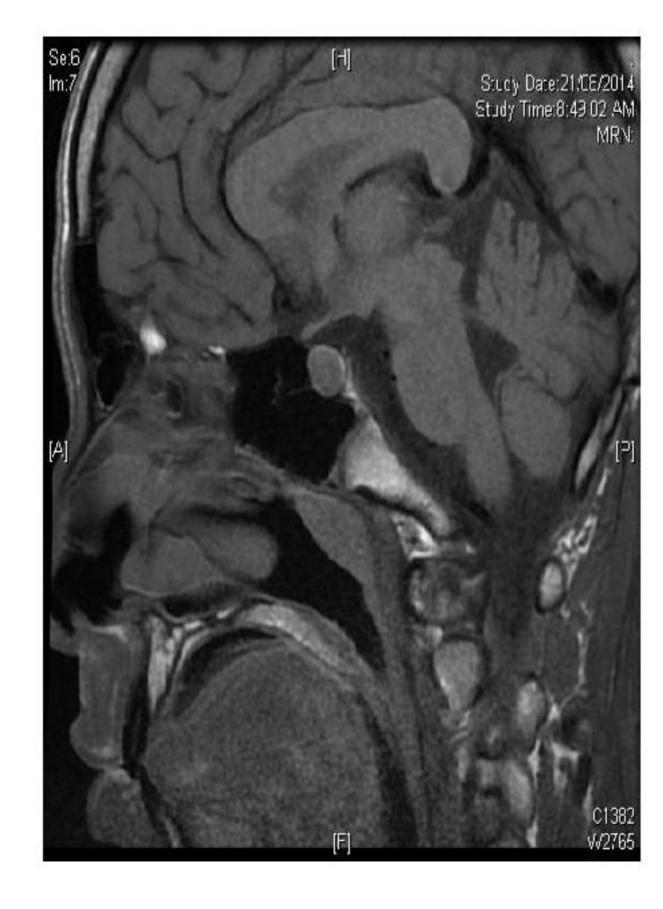


Figure 2: MRI pituitary

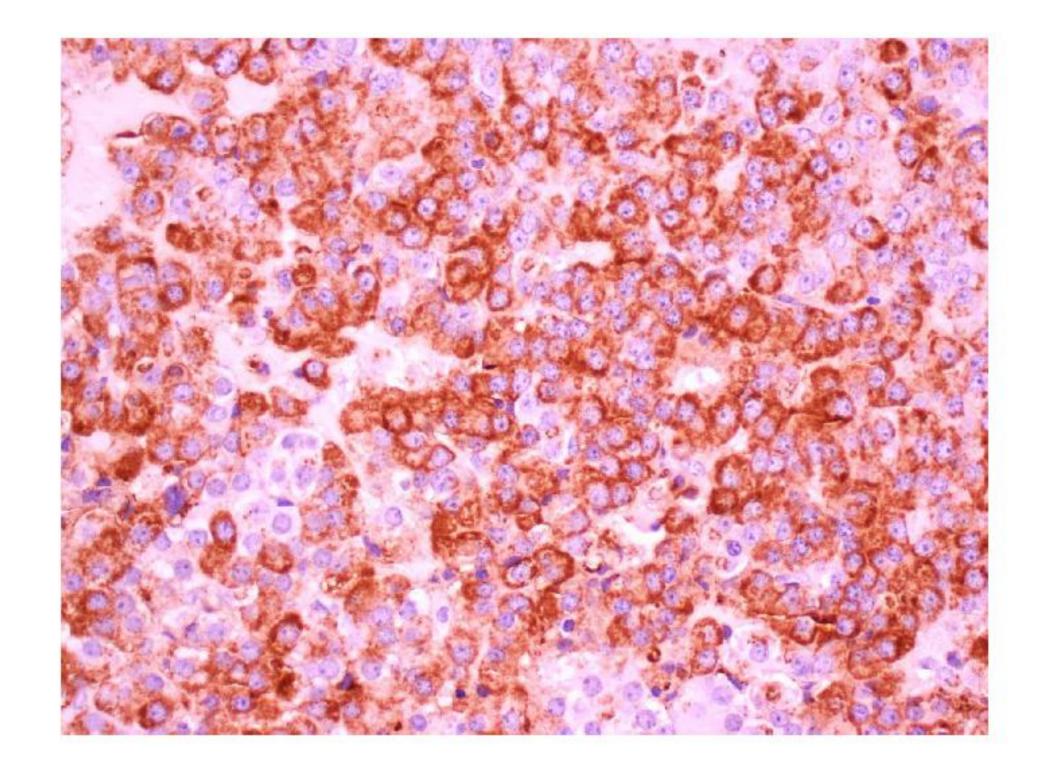


Figure 2: The tumour cells are strongly and diffusely positive for growth hormone on immunohistochemistry.

Clinical Evaluation

An oral glucose suppression test (OGTT) showed failure of GH suppression with a paradoxical rise after 60 minutes to a peak GH of 25.0 ug/L and an elevated IGF-1 of 877.0 ug/L (183-850). (Table 1). Serum prolactin was normal at 198 mIU/L (72.0-320.0).

A pituitary MRI (Figure 2) showed a pituitary macroadenoma measuring 12 x 9 x 7 mm. Perimetry was normal. He underwent endonasal transsphenoidal hypophysectomy and he developed central diabetes insipidus post operatively. Histology confirmed a pituitary adenoma which was positive for growth hormone. (Figure 3)

A repeat OGTT (Table 2) 3 months after surgery showed adequate suppression of GH levels to 0.73 ug/L with normalization of IGF-1 to 228 ug/L (183-850) and a pituitary MRI showed no tumour recurrence. He had no evidence of hypopituitarism on follow up.

Time (min)	0	30	60	90	120	150	180
Glucose (mmol/L)	4.0	5.1	4.9	5.1	5.0	4.8	4.0
GH (μg/L)	12.2	11.4	18.8	17.2	19.8	20.9	25.0

Table 1

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Time (min)	0	30	60	90	120	150	180
Glucose (mmol/L)	4.8	5.6	5.6	5.4	5.6	5.0	4.6
GH (μg/L)	1.75	0.94	0.42	0.41	0.93	0.73	2.83

Table 2

Discussion

Hypersecretion of GH in childhood causes gigantism with potential clinical symptoms including accelerated growth velocity with tall stature and enlargement of the hands and feet. Most cases are caused by benign pituitary adenomas. The median age at diagnosis has been reported to be at 12 years.

The gold standard for making the diagnosis of GH excess is failure to suppress serum GH level to less than 1 ug/L after For well-circumscribed p pituitary OGTT. adenomas, transsphenoidal surgery is the recommended treatment and it may be curative. Adjunctive therapy includes medical treatment with somatostatin analogs or radiotherapy.

Our patient achieved successful surgical and biochemical cure with transsphenoidal surgery. He will be followed up to monitor for tumour recurrence and pituitary function.

References:

- 1. Melmed S, Ho K, Klibanski A, Reichlin S, Thorner M. Clinical review 75: recent advances in pathogenesis, diagnosis, and management of acromegaly. J Clin Endocrinol Metab 1995;80:3395-
- 2. Abe T, Tara LA, Ludecke DK. Growth hormone-secreting pituitary adenomas in childhood and adolescence: features and results of transnasal surgery. Neurosurgery 1999;45:1-10.
- 3. Kane LA, Leinung MC, Scheithauer BW, Bergstralh EJ, Laws ER Jr, Groover RV, et al. Pituitary adenomas in childhood and adolescence. J Clin Endocrinol Metab 1994;79:1135-40.
- 4. Katznelson L1, Laws ER Jr, Melmed S, Molitch ME, Murad MH, Utz A, Wass JA; Endocrine Society. Acromegaly: an endocrine society clinical practice guideline. J Clin Endocrinol Metab. 2014 Nov;99 (11):3933-51. doi: 10.1210/jc.2014-2700. Epub 2014 Oct 30.







