

# A case of 17 year-old boy with relapsing Cushing's disease presenting vertebral compression fracture

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## INTRODUCTION

- Cushing's syndrome (CS) : The overall incidence is about 0.7–2.4 cases per million persons per year, only approximately 10% of the cases occur in children
- Cushing's disease: 75-80% of cases, hypercortisolism due to pituitary adrenocorticotrophic hormone (ACTH) secreting adenoma
- Reported cure rates following 1<sup>st</sup> line transsphenoidal surgery (TSS) in childhood Cushing's disease is ranged from 45% to 95%<sup>1</sup>.
- Herein, we report a case of 17-year-old boy with Cushing's disease who presented with obesity and compression fractures, and who showed relapsing Cushing's disease after TSS for pituitary microadenoma.

## CASE

### Brief history

A 17-year-old boy was referred to our hospital for evaluation of vertebral compression fracture and obesity. The weight gain was 23 kg but no height gain during four years. As this boy was belong to the special school due to mild intellectual disability (IQ 50-70), growth failure and obesity was neglected from previous clinicians in spite of typical cushingnoid features. Bone age as determined by the Greulich-Pyle method was delayed by 2 years, simple x-ray of whole spine showed osteopenia and compression fracture, and Z-scores of bone mineral densitometry (BMD) at the lumbar spine and femur neck were **-4.3** and **-2.3**, respectively.

V/S:100/60 mmHg-80/min-20/min-36.5°C

Height: 149.5 cm (**-4.1** SDS), weight: 63.6 kg (-0.1 SDS),

BMI: 28.5 kg/m<sup>2</sup> (**3.0** SDS)

testicle volume size 8/8cc, Pubic hair Tanner III

Selective serotonin receptor inhibitor (SSRI) was prescribed for depression



**Table 1** Levels of baseline and follow-up ACTH, and 24-hour urinary free cortisol according to clinical course

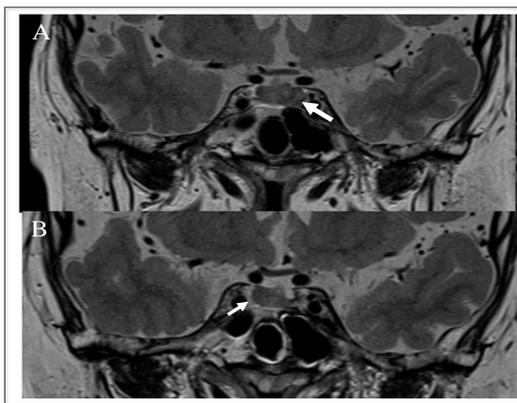
	At initial evaluation	Low dose DXM test	High dose DXM test	After 1 <sup>st</sup> operation	After 2 <sup>nd</sup> operation	The latest evaluation
AM Cortisol (ug/dL)	31.1	22.89	0.746	5.7	18.43	17.9
AM ACTH (pg/mL)	62.5	86.67	27.32	6.772	26.65	25.15
UFC (ug/day)	228.5	1363.7	5.1	302	229.7	416.0

ACTH, Adrenocorticotrophic hormone; DXM, dexamethasone; UFC, urinary free cortisol.

**Table 2.** Levels of ACTH and the ratio of central to peripheral ACTH measured during bilateral inferior petrosal sinus sampling after corticotropin-releasing hormone injection

ACTH	Peripheral (P)	Right (R)	Left (L)	R/P	L/P	R/L
Baseline	33.02 pg/ml	321.9	476.4	9.7	14.2	0.68
1min	28.77 pg/ml	863.2	1280	30	44.5	0.67
3min	63.76 pg/ml	>1500	>1500	>23.5	>23.5	1
5min	109.7 pg/ml	>1500	>1500	>13.7	>13.7	1
10min	173.1 pg/ml	>1500	>1500	>8.66	>8.66	1

IPS, inferior petrosal sinus sampling; R, right; L, left.



**Figure 1.** Brain MRI of patient with Cushing's disease showing a pituitary tumor (white arrow).(A) Coronal T2 image after ganolinium contrast at initial diagnosis showing isodense left microadenoma sized 8mm. (B) Coronal T2 image showing isodense 9mm sized right microadenoma in follow up MRI.

## CONCLUSION

- This report describes the relapsing and persistent adolescent Cushing's disease patient despite of several TSA.
- The early recognition and diagnosis of pediatric Cushing's disease will lead to recovery quickly and improve a quality of life.
- The findings emphasize the importance of monitoring growth in obese children to diagnose endocrinal disease like Cushing's disease.
- Moreover, for persistent or recurrent Cushing's disease patients, close follow-up of tumor status, severity of hypercortisolism, and patient's perspectives are the major parameters to determine the best option.
- As MRI may be negative in as many as 40% of cases of Cushing's Disease, despite the presence of a pituitary ACTH microadenoma<sup>2</sup>, not only BIPSS could be helpful for lateralization of tumor but also close monitoring of hormone status and radiologic image should be performed in surgically "cured" patients.

## REFERENCES

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