

A 5-year-old patient with Cushing's disease

Ushijima Kikumi, Sasaki Takako, Kitamura Miyuki, Yatsuga Shuichi, Koga Yasutoshi
Department of Pediatrics and Child Health, Kurume University School of Medicine, Japan

Nothing to declare

【Backgrounds】

The incidence of Cushing's syndrome in children is 0.2 to 0.5 / 1,000,000 / year¹. Cushing's disease (CD) is more infrequent especially under 7-year-old children². The typical symptoms are not often observed in childhood. The early diagnosis is difficult, if pituitary tumor is not detected by pituitary MRI.

【Case Report】

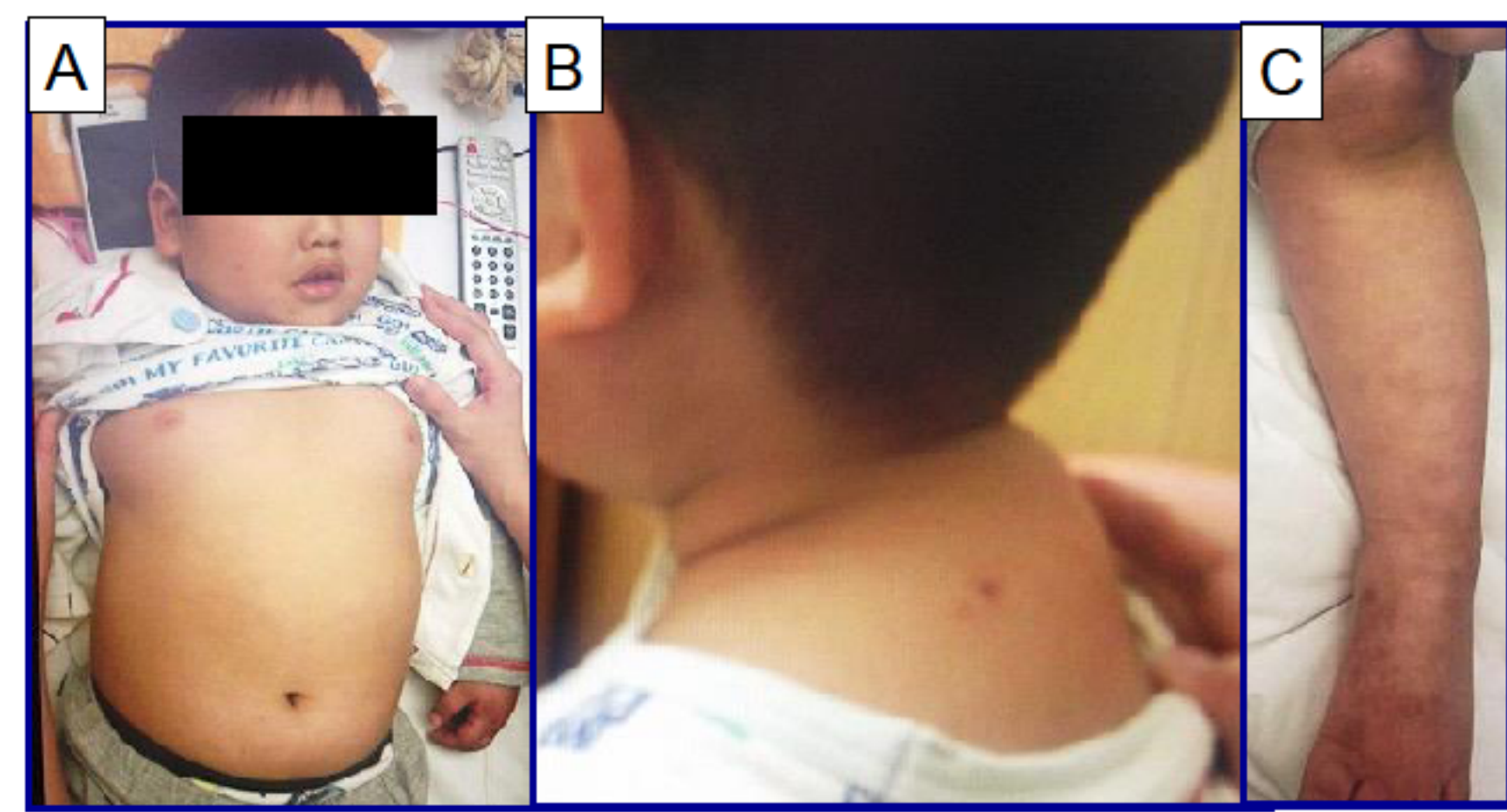
【case】 5Y1M, male

4Y6M body weight gain (7kg/2months)
diagnosed with primary obesity
started exercise therapy

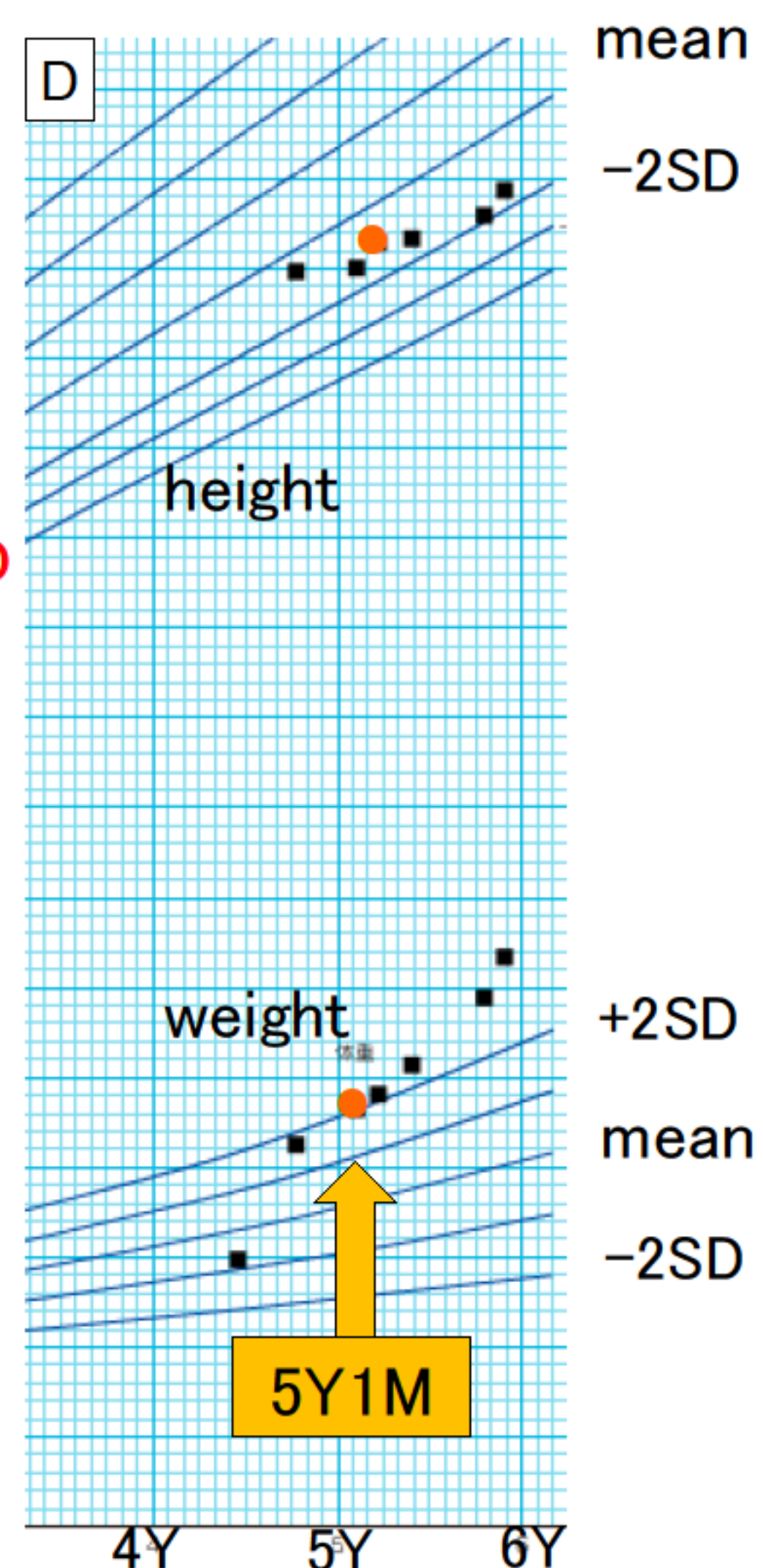
5Y1M referred to our emergency center due to near-drowning
referred to our department due to a decrease in growth
velocity with obesity.

【physical examination】

Height 100 cm (-1.7 SD)
Body weight 23.0 kg (+2.0 SD)
Blood pressure 128/71 mmHg
Head and neck moon face, buffalo hump
Skin hirsutism, pigmentation



A: moon face, obesity B: buffalo hump C: pigmentation



D: growth curve decreased in growth velocity with obesity

【laboratory data and imaging studies】

<blood examination (5Y1M) >

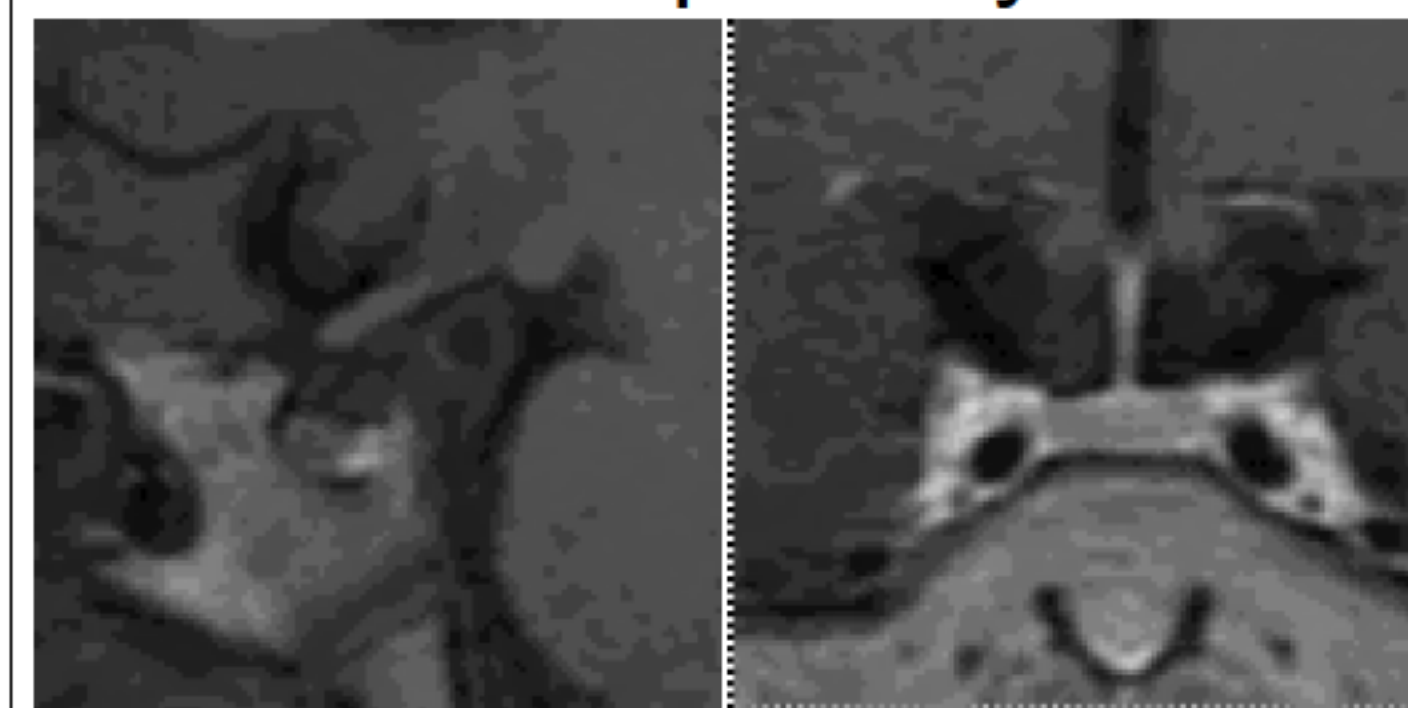
ACTH (8:00) 61.2 pg/mL
cortisol (8:00) 12.0 µg/dL
cortisol (24:00) 3.3 µg/dL
urinary cortisol 75.3 µg/m²/day
35.1 µg/m²/day
56.8 µg/m²/day
Low dose DEX (cortisol 8:00) 27.9 µg/dL

< bone age > 5 years old

<blood examination (5Y3M) >

cortisol (24:00) 7.1 µg/dL
urinary cortisol 88.6 µg/m²/day
71.7 µg/m²/day
60.5 µg/m²/day
Low dose DEX (cortisol 8:00) 21.2 µg/dL

< enhanced pituitary MRI >



no findings of adenoma

【laboratory data and imaging studies】

<blood examination (5Y1M) >

AST	31	U/L	K	4.8	mmol/L	LDL	151.8	mg/dL
ALT	29	U/L	Cl	111	mmol/L	TG	95	mg/dL
FBG	92	mg/dL	TC	222	mg/dL	IRI	7.4	µU/mL
Na	150	mmol/L	HDL	48.8	mg/dL	HbA1c	5.1	%
						IGF-I	125	ng/mL

<CRH test (5Y4M) >

CRH (1.5µg/kg)	basal	peak
ACTH (pg/mL)	22.5	50.9
cortisol (µg/dL)	21.6	33.6

【Results of bilateral inferior petrosal sinus samplings (5Y5M)】

CRH (1.5µg/kg)	ACTH (pg/mL)			
	basal	C/P	peak	C/P
Peripheral vein	40	-	58.7	-
Right inferior petrosal sinus	39.1	0.98	69.1	1.18
Left inferior petrosal sinus	429	10.7	2480	42.2

basal : C/P > 2
peak : C/P > 3

↓
Cushing's disease

C/P: central/peripheral

【Discussion】

Ninety percent of CD is caused by pituitary micro-adenoma. Conventional pituitary MRI can detect ACTH-secreting tumors in around 60%³. In pediatric CD, tumor size is relatively small⁴, therefore, we may not be able to detect pituitary tumor in this case. The median age at diagnosis with CD in prepubertal children (13 males and 4 females, range: 5.7-14.1 year-old) was 9.4 year-old, and median length of history prior to diagnosis was 2 year-old (range: 0.5-4.0 year-old)⁴. The our case was repeatedly performed using diagnostic tests, such as low dose DEX, urine cortisol level in 24 hours, and pituitary MRI, but we could not diagnose him with CD. The case was finally diagnosed with CD using by selective sinus sampling test because enhanced pituitary MRI was no findings of adenoma. The age at diagnosis with CD was 5 month, and length of history prior to diagnosis was 1.1 years. We believe that CD was diagnosed earlier using by selective sinus sampling test.

【Conclusion】

If pituitary tumors were not detected in pituitary MRI for children suspected with CD, we should perform a selective sinus sampling test to diagnose CD earlier even under 7-year-old.

references 1) Kishi K, et al. 2012, 2) Bas N. V, et al. 2013, 3) Kageyama K, et al. 2013, 4) Dias R. P, et al. 2010,

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Pituitary

Kikumi Ushijima

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