An 18-Month-Old Boy with Decreasing Height Velocity

Lana Soylu, MD, FAAP; and Vonita Chawla, MBBS

An 18-month-old boy presented to our clinic for a routine well visit. His mother was concerned that, although he was cruising at age 12 months, he was still not walking or talking.

He was born at 39 weeks and 5 days via cesarean delivery secondary to breech presentation. His APGAR (Appearance, Pulse, Grimace, Activity, Respiration) scores were 9 and 9 at 1 and 5 minutes, respectively. Antenatal course was only pertinent for gestational hypertension. His birth weight, length, and head circumference were 3,770 g (65th percentile), 52.7 cm (76th percentile), and 36.3 cm (49th percentile), respectively. His hospital course was uneventful and his newborn metabolic screen was normal.

His immunizations were up to date and his medical history and family history were noncontributory. His diet was appropriate for age.

A review of his growth parameters revealed a decline in linear growth velocity starting after age 6 months (from 76th percentile to <1st percentile) (Figure 1), a cessation of head growth starting at age 9 months (from 36th percentile to 2nd percentile), and a slight decline in weight percentile beginning at age 12 months (from 95th percentile to 73rd percentile).

His vital signs were within normal limits with the exception of blood pressure of 130/90 mm Hg. On physical examination, he was noted to be happy, interactive, and in no acute distress. His skin was slightly dry and he had blonde lanugo hair throughout. HEENT (Head, Eyes, Ears, Nose, Throat) examination was only pertinent for chubby cheeks (Figure 2). His abdomen was soft, nontender, non-distended without organomegaly or masses, and with normal bowel sounds. Genitourinary examination revealed a Tanner 1 circumcised boy with bilaterally descended testes. The remainder of the examination was unremarkable.

Due to his decline in length and head velocity, the patient was referred to endocrinology. Initial laboratory results from endocrinology showed a normal thyroid panel, renin activity, aldosterone, 24-hour vanillyl mandelic acid, metanephrines, dehydroepiandrosterone, insulin-like growth factor-1 (IGF-1) and insulin-like growth factor–binding protein 3; a decreased adrenocorticotropic hormone (ACTH) of 7 pg/mL; and increased serum cortisol (21.1 mcg/dL) and 24-hour urinary cortisol (65 mcg/24 h).

Given the above results, a 12-hour overnight dexamethasone suppression test (DST) was completed and resulted in a serum cortisol of 25.6 mcg/mL (which is elevated). Subsequently, an adrenal ultrasound revealed a left adrenal mass, which was confirmed by magnetic resonance imaging (MRI) of the abdomen to be a solid left adrenal mass measuring 3.9 cm x 5.2 cm, suggestive of left adrenal adenoma. MRI of the brain was normal. After a normal metaiodobenzylguanidine scan, a left adrenalectomy was performed and a tumor weighing 54.4 g was resected.

Lana Soylu, MD, FAAP, is an Assistant Professor of Pediatrics, University of South Florida. Vonita Chawla, MBBS, is a Medical Graduate, Pravara Institute of Medical Sciences.

Address correspondence to Lana Soylu, MD, FAAP, University of South Florida Health South Tampa Center for Advanced Healthcare, 2 Tampa General Circle, 5th floor Pediatrics, Tampa, FL 33606; email: lmccaule@health.usf.edu.

Disclosure: The authors have no relevant financial relationships to disclose.

doi: 10.3928/00904481-20151012-04

Editor’s note: Each month, this department features a discussion of an unusual diagnosis. A description and images are presented, followed by the diagnosis and an explanation of how the diagnosis was determined. As always, your comments are welcome via email at pedann@Healio.com.

continued on page 420
Diagnosis: Cushing’s Syndrome

The histopathology was consistent with a benign adrenal adenoma. The patient received stress doses of hydrocortisone during surgery and was placed on a slow, 6-month long hydrocortisone taper. He has since showed marked improvement in his linear growth velocity and head growth and was walking within 6 months of treatment. An abdominal ultrasound at the 6-month follow-up did not show any new masses.

DISCUSSION

Adrenal cortical tumors are neoplasms arising from the cortical layer of adrenal glands, which, when functional, can lead to an oversecretion of mineralocorticoids, glucocorticoids, and/or androgens. The incidence of adrenal cortical tumors in the pediatric population is extremely low (0.3 per 1 million children).\(^1\)\(^2\)

Prolonged exposure to excess corticosteroids manifests as a constellation of clinical features known as Cushing’s syndrome (CS). In children, these features include a decrease in linear growth velocity, weight gain, obesity, and facial plethora.\(^3\) The growth retardation is explained by two mechanisms. First, for reasons not completely understood, prolonged excess of glucocorticoids causes suppression of growth hormone production, or the glucocorticoid receptor represses activator protein-1, a transcription factor that mediates the effects of IGF-1 and is responsible for the differentiation, proliferation, and apoptosis of cells.\(^4\)

Cushing’s syndrome is either ACTH-dependent (most common) or ACTH-independent.

Aldosterone excess increases salt retention, leading to hypertension and low potassium, and in cases of excess androgens, genital virilization and hirsutism may be seen. Any of these signs or symptoms should prompt further evaluation.

The initial testing is to establish the presence of elevated cortisol. That can be done with any of the following methods: two measurements of 24-hour urinary free cortisol, two measurements of late night salivary cortisol, or an overnight 1-mg DST.

Any abnormal test result (ie, cortisol >50 nmol/L after overnight DST, late-night cortisol >4 nmol/L, or cortisol greater than the upper limit of normal in a 24-hour urinary free cortisol measurement) should prompt further evaluation to identify ACTH dependency and referral to an endocrinologist.\(^5\)\(^6\) MRI of the brain to look for pituitary tumors along with surgical consultation may also be warranted.

Although unclear, multiple studies have shown that none of the patients diagnosed with a benign lesion has died of the disease.\(^7\)\(^8\) Treatment is aimed at removing the source of excessive cortisol, so surgery continues to be the first-line treatment. Patients require intraoperative and postoperative steroid replacement followed by a taper over a period of 6 months.\(^3\)\(^7\)\(^9\) Additional steroid bursts may be necessary during illness or stress. Patients will need ongoing monitoring of blood pressure, routine blood chemistries and hormone levels, renal and pelvic ultrasounds, and physical examinations to evaluate for reoccurrence of the tumor. Over the long term, these children typically do well and resume

Figure 1. Decline in linear growth velocity starting after age 6 months, from the 76th percentile to <1st percentile.
growth and development once the exogenous source is removed (Figure 3).

CONCLUSION

Although pediatric adrenocortical tumors are rare, when functional they can lead to significant disruption in growth and developmental delay. Glucocorticoid excess can present with growth failure and facial plethora, and mineralocorticoid excess can lead to hypertension and low potassium. If either of these excesses are expected, the source needs to be determined, followed by appropriate management. For benign adrenal cortical tumors, resection remains the standard of care followed by steroid replacement with a gradual taper. These patients will also require regular evaluations for recurrence. Patients with benign tumors have a good prognosis.

REFERENCES