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A Case of Cushing's Syndrome in a Child, due to Adrenocortical Adenoma

by

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Introduction

Cushing's syndrome, though uncommon in infants and children, should be considered in the evaluation of children with obesity. The clinical features of this disorder are familiar to most physicians, not least of all the identical pattern as the results of prolonged therapeutic administration of cortisone.

The outstanding symptoms are obesity and the characteristic moon face. The face has ballooned and reddened cheeks, frequently there is a pustular acneiform eruption over the face. The obesity is in children and infants massive and generalized, involving the extremities as well as the trunk. It is different from the obesity with thin arms and legs of the adult with Cushing's syndrome. There are hypertension, glucosuria, a decreased glucose tollerance test, weakness, and virilizing symptoms. Purple striae are not seen in infants, mainly in older children. Osteoporosis appears to develop faster in infants than in adults.

An important sign in children is retardation in linear growth. The clinical features of the syndrome are due to the effects of excessive adrenocortical hormones, both 17-hydroxysteroids resulting from metabolism of glucocorticoids (cortisol) and 17-ketosteroids reflecting production of androgens.

The lesion in the adrenal gland may be a tumor (adenoma or carcinoma) or hyperplasia. Prolonged administration of glucocorticoids may produce a clinical syndrome similar to the spontaneous disease (Visser, 1966; Nelson, 1969; Wilkins, 1950).

In children tumors are the most common cause of Cushing's syndrome, many of them being carcinoids. Gilbert and Cleveland (1970) reviewed 92 cases of Cushing's syndrome in children under 15 years of age, reported between 1802 and 1969. Sixty-one patients had carcino-

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mas and 2 had neuroblastomas; only 14 patients had benign adenomas, and 18 had hyperplasia. The incidence of malignancy was somewhat lower under 1 year of age. Of 27 patients under the age of 1 year, collected from the literature from 1924 to 1969 by Gilbert and Cleveland (1970), carcinoma was the final diagnosis in 11 patients, adenoma in 10 and hyperplasia in 6. Only 12 of the 27 patients survived.

Since adrenal tumors cause Cushing's syndrome in over 80% of patients under the age of 15 years, the most important radiologic study is the intravenous pyelogram in order to localize the lesion (Darling, 1970).

Treatment in the situation of adrenal tumor consists of removal of the mass with proper preparation of the patient, especially since upon the removal of the tumor the patient will usually have little or no endogenous adrenal function. Preoperative administration of cortisone acetate is desirable, and following operation cortisone is given in gradually diminishing dosage. As it may be difficult to determine from either the gross or microscopic findings whether adrenal tumors are malignant or benign (Anderson, 1968) and even in the absence of demonstrable local invasion recurrence may take place, it is important to follow the patients carefully in order to discover clinical signs of a reappearing Cushing's syndrome and to measure excretion of steroids periodically.

Case Report

P, a 15-month-old female, was admitted to the Bethesda Hospital on 24-8-1972, because of obesity and moon face, present for about 5 months. Otherwise the child was well. She had not been ill yet. Appetite was normal.

The child was the 4th in a healthy family, born fullterm after a normal pregnancy. Birthweight was 3.5 Kg. On admittance the child was very obese. Bodyweight was 10.85 Kg.; length was 70 cm.

There was a marked general obesity with a buffalo hump, and the face was typical for the Cushing's syndrome, with reddened cheeks and acneiform eruptions over the forehead and cheeks. Hypertrichosis over the back and slightly on the cheeks. No pubic hair; slight clitoral enlargement. No striae were seen. On abdominal examination no masses could be felt. The child was weak and slept a lot.She could not stand up yet. Anterior fontanel was not yet closed (Fig. 1).

Laboratory data including complete blood count, urinalysis, serum electrolytes and fasting blood sugar were normal. Hemoglobin was 11.2 gm%. White blood count 5800/mm^3. Eosinophils 1%. Erythrocyte sedimentation rate 8-18 mm. Fasting blood sugar 96 mg% and 90 mg%.

FIG. 1 : *P. preoperative*

FIG. 2 : *Adrenal adenoma.*

FIG. 3 : *Microsc. (× 80) Thin capsule.*

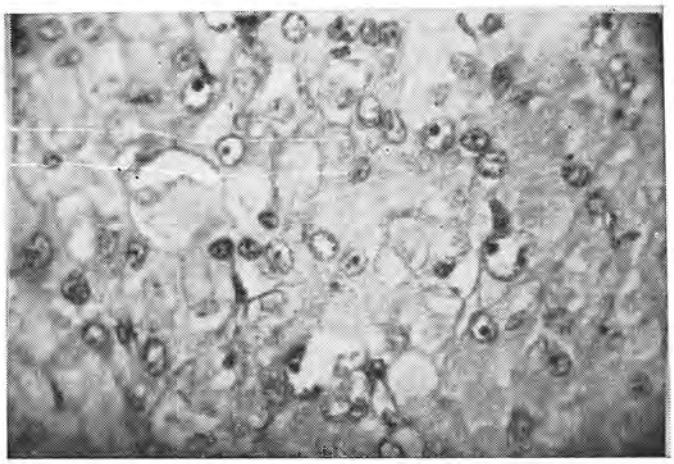

FIG. 4 : *Microsc. (× 360)) Atypic polymorph cells, vesic, nuclei, distinct nucleoli.*

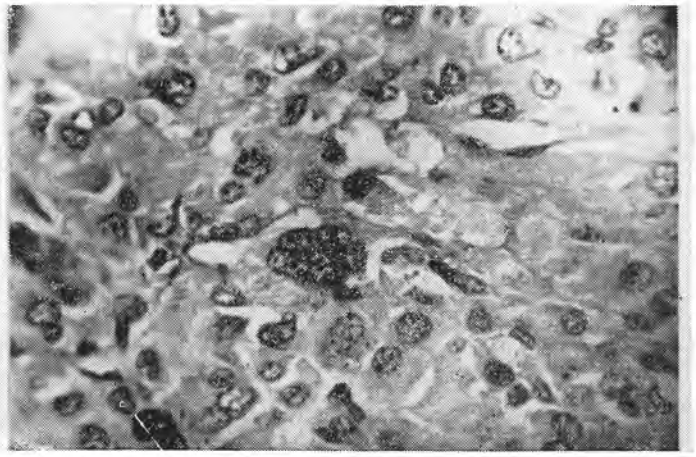

FIG. 5 : *Microsc. (× 360) Giant cell.*

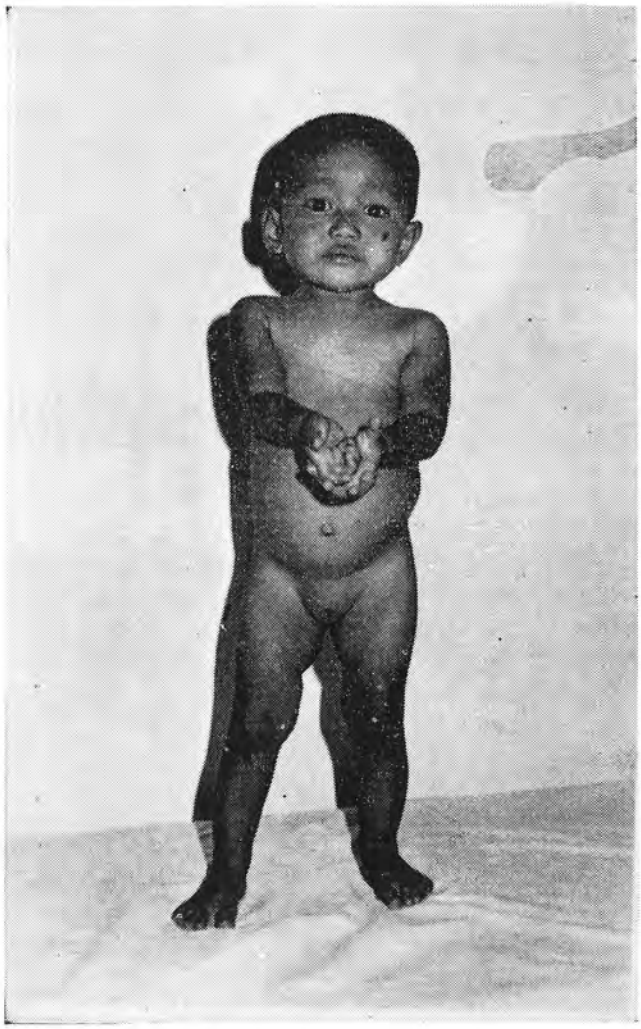

FIG. 6 : *P. at 2 years of age.*

Serum sodium 143.2 mEq./liter; serum potassium 4.8 mEq./liter and 5.7 mEq./liter; serum cholesterol 146.2 mg%.

The urinary 17-OHCS excretion was 5.0 mg/24 hours, a second time 3.2 mg/24 hours. The excretion of 17-ketosteroids was 4.1 mg and 6.6 mg/24 hours.

No ACTH-adrenal suppression test was performed.

Radiologic examination revealed diffuse osteoporosis. Carpals showed 2 primary ossification centers. The pituitary sella appeared normal.

An intravenous pyelogram after retroperitoneal insufflation of air demonstrated inferior displacement of the left kidney by a 2 cm in diameter suprarenal mass, with a normal urinary tract on the right side. *).

Hospital course:

Since a definite diagnosis of left adrenal tumor was made, suppresion test was not done. The child was prepared for operation by administration, of 50 mg cortison acetate intramuscularly, 6 hours prior to operation and antibiotic cover with gentamycin. Through a leftside lumbar incision and resection of the 12th rib, a 2 by 3 cm spherical, solid, bluish, well capsulated tumor in the left adrenal gland was easily removed, intact, where upon a left adrenalectomy was performed.

Postoperative course was uncomplicated. Gentamycin was discontinued on the 5th day after operation, and cortison was gradually reduced and completely discontinued 20 days after the operation (Aerography and operation were done by surgeon C.G.G. Boeken Kruger of the Bethesda Hospital).

Pathologic findings:

The gross finding was an encapsulated grayish mass measuring 3 by 3½ by 2 cm. (Fig. 2). Microscopic finding was an atypic epithelial polymorph tumor, diffusely growing and pressing the thin covering (Fig. 3). Tumor cells had granular eosinophilic cytoplasm with big, vesicular or lobed nuclei, containing irregular chromatin and sometimes distinct nucleoli. Mitosis was very rare. Some spindle cells were present as well as some giant cells with more than one nucleus. (Fig. 4, 5). Diagnosis was adrenocortical adenoma. (Sukarti/Bambang Soetarso/Soeripto, Department of Pathology, Gajah Mada University).

Subsequent course:

After the operation a gradual change in appearance occurred with the loss of typical features and a loss in weight. The excretion of 17-hydrocorticoids diminished and was normal (1.05 mg/24 hours) one month after

*) After the child had been sedated, insufflation was done by introducing a long needle post-anally into the presacral space, controlled by a finger in the rectum; 120 ml. air was injected with a 20 ml. syringe, after which the child remained in a semi-reclining position, and the next morning pyelography was performed.

operation. So were the 17-ketosteroids (0.86 mg/hour). The Cushingoid features and obesity totally disappeared within a 3-months period.

A last check-up was done on 12-1-1974, at the age of 2 years and 4 months. The child was healthy, active and gay. She could run, spoke rather well, and had no Cushingoid features at all. Appetite was moderate. Weight was 10.1 Kg; height 78 cm. Hemoglobin 11 gm%. (Fig. 6) *).

Summary

Description of a case of Cushing's syndrome in a young child due to a benign adenoma of the adrenal gland, in which surgical cure was achieved is presented.
Clinical picture was diagnostic, with typical facies and generalized obesity. Intravenous pyelogram indicated the presence and location of the tumor. Excretion of both 17-hydrocorticoids and 17-ketosteroids was increased and fell to normal after removal of the tumor.

Although the histological differentiation between adenoma and carcinoma can be difficult, and in our case the tumor cells were atypic and polymorph, and some mitotic figures were present, since there was no invasion to the capsule the tumor was diagnosed as an adenoma.

The tumor has been completely removed and the child has been free of any abnormal finding for about 15 months.

Acknowledgement

The author wish to thank Drs. Sukarti, Bambang Sutarso and Suripto from the Department of Pathology, Medical School, University of Gajah Mada, who had so kindly examined the histopathology and confirmed the diagnosis of adrenocortical adenoma.

*) Because of no availability of percentiles of weight and height of Indonesian babies, using the standard mean-values of measurements by Moh. Sugiono and Te, B.S. (1964), we can say that at the time of admission the child had the length of a child of 12 months and an overweight of 2.850 Kg (weight for height: 8 Kg). As at the age of 2 years and 4 months, height and weight were practically normal according to the standard measurements by Tan, E.D., R. Soekonto and J.H. de Haas (1938). (Mean value at 2 years: length 78.4 cm; weight 9.3 Kg).

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