

ADRENOGENITAL SYNDROME IN A FEMALE WITH ADRENAL CARCINOMA

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ABSTRACT : Adrenogenital syndrome due to an adrenal carcinoma is a very rare presentation. This is the case report of a 22 year old female who presented with classical features of the syndrome and underwent adrenalectomy.

KEY WORDS : Adrenal Hyperfunction, Hyperandrogenism, Amenorrhoea, Adrenogenital Syndrome, Adrenal Neoplasms, Adrenalectomy.

INTRODUCTION

Tumours of the adrenal gland are of rare occurrence. They may arise from the adrenal cortex or the medulla^{1,2}. Tumours arising from the adrenal medulla include pheochromocytoma, ganglioneuroma and neuroblastoma while those arising from the cortex produce classical syndromes like Cushing's, Conn's and adrenogenital syndrome depending upon the production of glucocorticoids, mineralocorticoids and sex hormones respectively. A small number of nonfunctioning tumours have been found at autopsy or discovered accidentally (incidentalomas)¹ on CT scan, MRI or ultrasonography performed for some other condition. Adrenal¹ tumours may be benign or malignant, infiltrating surrounding structures or invading the renal vein and metastasizing.

The adrenogenital syndrome is due to overproduction of adrenal cortical androgens and may be congenital or acquired³. The congenital type is the result of an inborn defect of normal steroid synthesis causing ACTH hypersecretion, and ultimately excessive secretion of the cortical androgens. The acquired variety in children is always due to an adrenal cortical tumour⁴ while in adults it may be due to a tumour⁴ or cortical hyperplasia seen in cases of Cushing's syndrome⁵. This report describes the cases of an adult female who presented with classical features of adrenogenital syndrome due to an adrenal cortical carcinoma.

CASE HISTORY

A house wife, aged 22 years was admitted with history of amenorrhoea for 9 months after taking some family planning medicine from a general practitioner. There was no menstrual irregularity prior to this. Six months ago she started developing pigmentation all over the body which steadily increased ever since. Along with the pigmentation excessive hair started growing all over the face and she also complained of acne. With the passage of time she developed ever increasing hair all over the body. She also complained of swelling of face, loss of weight and weakness.

The past history was insignificant. She was happily married with two children, a daughter and a son. Apart from betel nuts she is not addicted to anything. Socio-economically she belonged to the poor class.

On examination the patient was a female of small built, wasted, co-operative, having facial hirsutism and acne (Fig. 1), with a coarse voice. She had a rapid pulse and a blood pressure of 190/140 mmHg. General examination revealed pigmentation all over the body and oedema of the face.

Her chest examination revealed atrophied breasts, pigmentations and tachycardia. The abdomen (Fig. 2) was slightly protuberant with pigmentation and android-type obesity and hair distribution. The limbs also



Fig. 1.



Fig. 2.

appeared wasted, pigmented and hairy (Fig. 3).

A provisional diagnosis of adrenogenital syndrome was made and investigations including complete blood picture, urine analysis, blood sugar and urea, liver function tests, thyroid profile, and serum - calcium, electrolytes, creatinine, acid phosphatase, aldosterone, cortisol and testosterone were carried out. X-ray chest, ultrasound abdomen and pelvis, and a CT scan of the abdomen were also performed.

The results of the various investigations were normal except for serum testosterone which was > 1230 ng/dl (range 30-140) and serum cortisol - 39.9 $\mu\text{g}/\text{dl}$ (range 6-25); serum calcium and potassium were found just below the normal range. Ultrasonography revealed

hypoechoic non-homogenous mass, $86 \times 80 \times 65$ mm in size, displacing the kidney downwards. CT scan (Fig. 4) findings were similar with no invasion of the surrounding tissues or the renal vein.

After stabilizing the patient medically on minipress, tenormin and acetopril, and thorough preoperative preparation, the patient was operated through a classical Rutherford Morrison incision with resection of the 12th rib. The kidney and the tumour (adrenal) were identified. The tumour was then separated from the kidney and adrenalectomy performed after ligating the vessels in the pedicle. Haemostasis secured and the wound closed with tube drainage from a separate stab incision. Postoperatively the patient was put on antibiotics and hydrocortisone which were gradually



Fig. 3.



Fig. 4.

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