Case Report

Feminizing adrenal tumor: A case report and review of the literature

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ABSTRACT:
We present a case of Feminizing adrenal tumor with high estradiol who presented with fatigue, anorexia, decreased libido, and gynecomastia. He had bilateral gynaecomastia without galactorrhea. Estradiol was high (1722 pg/ml) with low testosterone (0.47 ng/ml:n=2.4-8.3). Abdominal ultrasound showed a huge tumor above the right kidney that was compressed and was confirmed by CTscan.At surgery, a well-limited much vascularized mass weighing 2.5 Kg was removed successfully. (Rawal Med J 201;42:127-129)
Keywords: Adrenal, feminizing tumour, gynaecomastia.

INTRODUCTION

The term of feminising adrenal tumors (FATs) is used to describe tumors secreting estrogens, especially in men.[1,2] These tumors are uncommon as they account for only 0.37-2% of all adrenal tumors. Clinical manifestations aregynecomastia or breast tenderness with or without other manifestations of gonadal deficiency. Sometimes a huge abdominal tumor is observed.[3]Gynecomastia without galactorrhea is generally the first symptom. Its association with erectile troubles is classical, even in subjects with normal testosterone rates. As the tumor is retroperitoneal, it is rarely found by palpation. Feminization results from the imbalance between high estrogen and low free androgens.[3]

FATs are characterized by secretion of estrogens or by a mixed secretion.[4] Their association to overt or sub clinical hypercortisolism is more common than androgen.[5-7] The hormonal secretion is generally autonomous as in other adrenal tumors but in some cases, it is stimulated by ACTHand suppressed by dexamethasone.[8]These neoplasms are usually seen in adult men over 70 (7%) than in those under 30 years old (1%).[1] They are very rare in women and children.

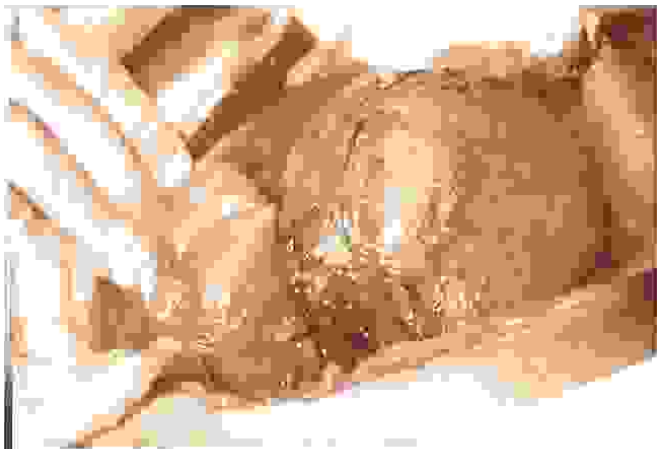
CASE PRESENTATION

A male patient aged 36 year,teacher presented with feeling enlargementof breasts six months ago and gradual distention of abdomen for the past two months.He also noticed that he was becoming obese gradually and had gained about 5 kg of weight. He noticed purplish striaeour abdomen, easily fatigued and dyspneic with his routine job. Five months ago bilateral mamoplasty was done at a hospital for bilateral gynecomastia.

He had central obesity, with moon-shaped rounding of the face, thickening of the fat pads in the supra-clavicularfosae. There were two scars of bilateral mastectomy. Height was 5 feet 7 inch and 70kg Weight.The abdomen was distended with large, livid purplish striae on both sides. A firm smooth non tender mass moving slightly and filling whole of right hypochondrium and lumbar region was palpable. There was no loss of axillary or pubic hair and the testes were atrophic.

CBC was normal and ESR was 45.X-ray chest (PA) view, blood sugar, blood urea, urine examination, liver function tests and ECG were normal.Imaging showed left kidney was normal and right kidney was displaced by extensive soft tissue mass. Estradiol was high (1722 pg/ml) and testosterone (0.47 ng/ml:n=2.4-8.3) was low.

Figure. A huge tumor (2.5 kg) at operation.



Removal of the tumor done under general anaesthesia using thoracoabdominal approach.The mass (2.5 kg) was well capsulated and right kidney was pressed down. Pedicle had infilterated inferior vena cava.Post operatively he was given antibiotics and corticosteroids and analgesics. He made a good recovery. Histologic diagnosis wasfeminizing adrenal carcinoma.

DISCUSSION

Feminizing adrenal cortical tumors account for less than 2% of all adrenal tumors.[9]They are characterized by secretion of estrogens.These neoplasms are usually seen in adult men and are very rare in women.Some authors propose a mutation in suppressive gene tumor located in 17p13 chromosome.[10]The main clinical signs in men are gynecomastia, decreased libido and erections, or ejaculation problems.A very high estradiol and estrone, androgen precursors, and mixed secretion spell worst prognosis.[10]

Cushing'syndrome resulting from adrenal tumor occurs in approximately 1 ofevery 10,000 hospital admissions.Most common sign is gynecomastia.[11] The diagnosis is made by the observation of other signs of adrenal over activity, e. g. hypertension, demonstration of excessive estrogen in the urine, and demonstration of a supra-renal mass by intra-venous pyelography (IVP) or arteriogram. This patient was operated 5 months ago for bilateral gynecomastia by the surgical specialist but nobody investigated the cause of it.

The only completely satisfactory treatment of adrenal tumor is the surgical removal. It is essential that the patient be supported with gluco-corticoids during and for several days after the operation, because chronic suppression of ACTH production, which can result in atrophy of the non- tumorous adrenal. Some physicians use anti cortisol products such as OP'DDD (dichlorodiphenyldichloroethane) or mitotane and aminoglutetimide.[10]Some authors suggest use of new aromatase inhibitors to reduce aromatase activity and estradiol production, but this new therapy needs time to be evaluated.[3]In summary,adrenal tumor is a rare and can present with various endocrinopathies. The most common is Cushing's syndrome, whereas the most rare is feminization.Cure is only possible with complete resection.

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