MANAGEMENT OF ADRENOCORTICAL TUMOURS

ESSAY

SUBMITTED FOR PARTIAL FULFILMENT
OF MASTER DEGREE IN GENERAL SURGERY

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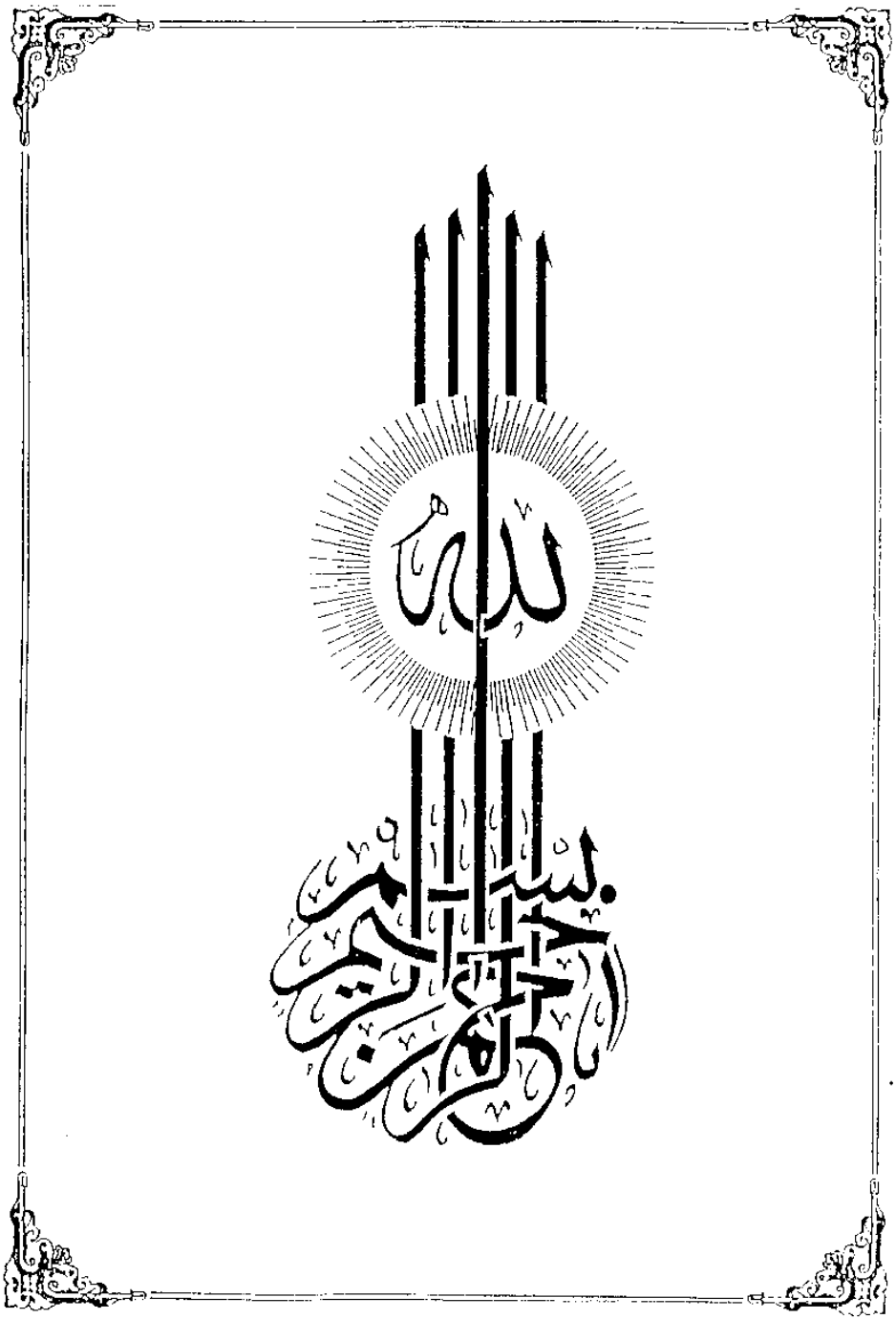
1992

ACKNOWLEDGEMENT

I would like to express my deepest appreciation and thanks to PROFESSOR DR. IBRAHIM KAMEL EL-WEKIL, Professor of General Surgery, Ain Shams University, for his kind supervision and persistent encouragement. His continuous guidance and assistance were much valuable.

Also, I wish to acknowledge my sincere gratitude to PROFESSOR DR. AMIR ISMAIL EL- LAMEI, Professor of General Surgery, Ain Shams University. He has given me so much of his experience and time, and provided me with a lot of valuable comments and suggestions.

My obligation to DR. HOSAM EL-DIN EL-AZZAZI, Lecturer of General Surgery, Ain Shams University, for his unique cooperation, suggestions and encouragement. I feel fully indebted to him for the time he generously devoted to review this manuscript.



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INTRODUCTION

The surgeon must have a working knowledge of adrenal pathophysiology and the essential clinical and laboratory evaluations of the patient, how to differentiate between the surgical and non- surgical adrenal diseases and point the way to appropriate surgical procedure.

The surgical treatment of diseases of the adrenal glands has evolved during the past half century, during which time the endocrinology of the adrenal cortex and the adrenal medulla were elucidated. In a sense, surgery of the adrenal glands is the treatment of functional and neoplastic disorders of the cortex and the medulla.

Currently, adrenal operations are done primarily for the treatment of a variety of endocrinopathies caused by tumours or hyperplasias of functionally different cells within the cortex and medulla.

Advances in adrenal surgery occurred as the adrenal cortical hormones and catecholamines were identified and synthesized. The ability to detect their presence in blood and urine by biochemical and immunoassay testing has made the diagnosis of adrenal disorders a practical reality. Furthermore, the operative treatment of functional adrenal tumours has been greatly aided by localization techniques such as computed tomography, adrenal scintigraphy, magnetic resonance imaging and selective plasma hormone sampling. In

addition, the use of pharmacologic agents to counter the deleterious effects of hormone excess, before operation, has made formerly hazardous procedures safe and almost routine today.

With the recent revolutionary advances in the diagnostic techniques, adjuvant with progress in anaesthesiology and advent of life reserving substitution therapy, safe removal of the adrenal gland becomes feasible.

AIM OF THE WORK:-

The aim of this work is to lay stress on the pathology, clinical picture, diagnosis and management of adrenocortical tumours that are commonly overlooked with review of the most recent advances in the laboratory investigations and the precise localization of these tumours by the different radiographic techniques.

EMBRYOLOGY AND SURGICAL ANATOMY

EMBRYOLOGY OF THE ADRENAL GLAND

The adrenal cortex and medulla originate separately. The cortex develops from mesodermal elements and the medulla from ectodermal cells of the neural crest (O'Neal, 1968).

THE ADRENAL CORTEX:-

It develops by proliferation of cells derived from the mesoderm of the coelomic mesothelium. During the fourth week of embryonal life, mesothelial buds appear at the level of upper third of the mesonephron and project into the celom at each side of the root of the dorsal mesentery. Eventually, they coalesce to form a compact mass of cells, the adrenal cortex, lateral to the aorta (Dluhy et al., 1979).

After vascularisation and encapsulation by surrounding mesenchyme, these mesothelial cells proliferate and grow to differentiate into large acidophilic cells forming the foetal cortex. Later, a further mesothelial proliferation occurs forming smaller cells that cover the outer surface of the foetal cortex.

As development proceeds, the outer smaller cells become arranged into zona glomerulosa and zona fasciculata of the definitive cortex. The surrounding mesenchyme forms the capsule and connective tissue of the cortex.

After birth the fetal cortex retrogresses and its involution is largely completed in the first few weeks of life (Snell, 1981).

The zona glomerulosa can be identified shortly after birth, the zona fasciculata appears by the third week and the zona reticularis by three to six months of age (O'Neal, 1968).

The cortical buds that do not join the main cellular mass disappear or may be left behind in various locations to form accessory adrenal cortical tissues. These are most commonly found close to the adrenal, within the kidney, in the celiac plexus, in the spermatic vessels, testis, in the broad ligament of the ovary, in the ovarian pedicle,in the ovary itself or in the para-aortic areas (Dluhy et al., 1979).

The anomalous locations of the adrenal cortex are clinically important for the following reasons:-

1- Hyperplasia in the accessory adrenal tissue may produce continued adrenal activity after adrenalectomy for Cushing's syndrome or for metastatic cancer.

2- Adrenal insufficiency occasionally develops when misplaced normal adrenal glands are inadvertently removed during nephrectomy.

3- Neoplastic transformation of heterotropic or accessory adrenal tissue may take place (Schechter,1968).

THE ADRENAL MEDULLA:-

It is ectodermal in origin. It develops by migration

of sympathetic primordial cells (Sympathogonia) from the neural crest where they cluster ventrolateral to the spinal cord to form the sympathetic ganglion primordia and ventrolateral to the aorta to form the preaortic ganglion primordia. Some of these cells differentiate into chromaffin endocrine cells and penetrate the capsule of the proliferating adrenal cortical primordium from the medial side, thus forming the adrenal medulla (O'Neal, 1968) about the 7^{th} week of fetal life (Dluhy et al., 1979).

The medulla remains relatively undeveloped at birth and attains maturity during the first five years of postnatal life (Snell, 1981), but it remains illdefined until puberty (Smith et al., 1984).

Numerous small groups of chromaffin cells are also found in association with the prevertebral sympathetic ganglia, the celiac, mesenteric, renal, adrenal and hypogastric plexuses. A large collection above the aortic bifurcation is called the organ of Zucherkandl. In the bladder chromaffin cells are found within the sympathetic nerve fibers. Pheochromocytomas frequently develop in these accessary sites.

Accessory adrenal tissue may consists of cortical or medullary tissue alone or may contain both (Dluhy et al., 1979).

The function of the fetal cortex is not completely known. Several investigations have shown that the fetal cortex can synthesize hydrocortisone, corticosterone and

other steroids.

At birth the adrenal glands are relatively large but later on,the fetal cortex involutes and the gland becomes reduced in size.

In a foetus of 2 months, the adrenals are larger than the kidneys. By the end of the 6^{th} month, the kidneys become about twice as larger as the adrenals. At birth, the adrenal gland is about one third the size of the kidney, while in the adult it is only about one - thirtienth (Di George, 1983). This change in proportions is not only due to renal growth,but also due to involution of the foetal cortex after birth (Warwick and Williams, 1973).