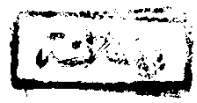
INTESTINAL POLYPOSIS

Essay

SUBMITTED FOR PARTIAL FULFILMENT

OF THE MASTER DEGREE IN

GENERAL SURGERY



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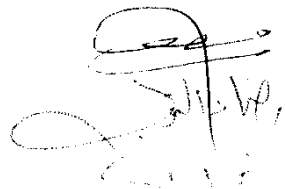

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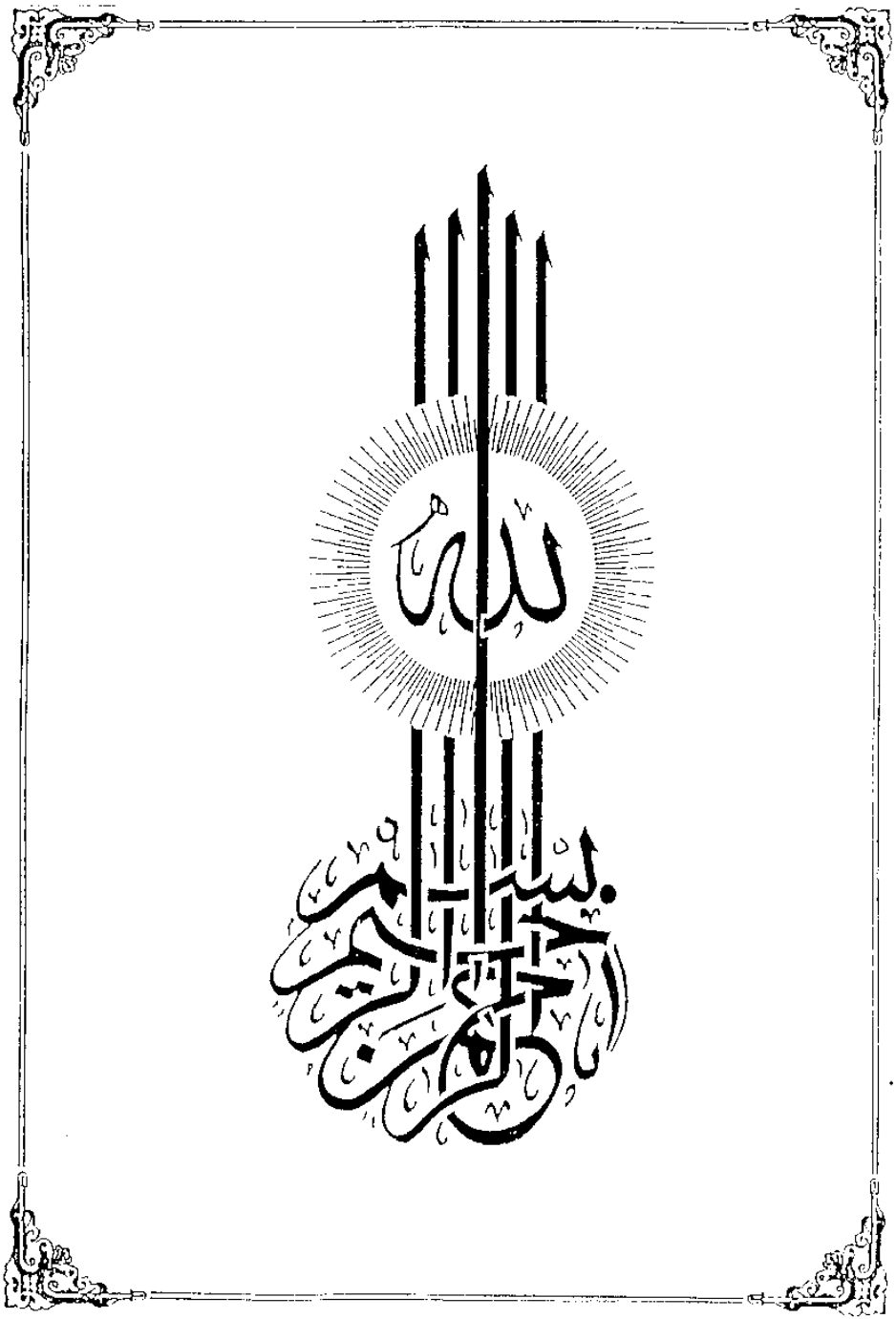

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INTRODUCTION

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Intestinal polyposis encompasses a spectrum of abnormal growths: hyperplastic polyps, the etiology and significance of which are virtually unkown; inflammatory polyps, which accompany non-hereditary conditions but do lead to cancers; hamartomas, both single and multiple which can follow a hereditary pattern with definable syndromes which are often associated with adenomatous polyps, in which include numerous inherited syndromes that usually appear to have a Mendelian dominant inherited pattern. Familial polyposis, Gardner's syndrome, and Turcot's syndrome are all parts of a systemic overgrowth pattern, and the associated soft tissue, bony, glandular and brain tumours illustrate the wide spectrum of this disorder (Watne, 1987).

There is large evidence supporting the importance of intestinal polyps as a precursor to the development of colorectal cancer. These precancerous polyps are tubular adenoma, villous adenoma, tubulovillous adenoma, familial polyposis coli, Gardner's syndrome and Peutz-Jeghers syndrome (Shinya and Wolff, 1979). The exciting thing about the intestinal polyposes is that, they allow us a human model to study the colonic mucosa through the polyp-cancer sequence, and thereby gain insight into the problem of cancer induction (Watne, 1987). Therefore, it

is useful to achieve an early detection of intestinal polyposis to prevent their complications especially malignant transformation and the methods of early detection should be considered.

CHAPTER ONE

HISTORICAL REVIEW

HISTORICAL REVIEW

Lesions of the large bowel consistent with polyposis coli were described by Menzel in 1721 in a soldier who ultimately died of chronic dysentry (**Barcon 1957**). In a lecture at Guy Hospital in 1824, Sir Astley Cooper described a case of polyps of the rectum in a young patient and stated that he had never before heard of such a disease (**Dukes, 1930**). In 1847, Corvisart described what ressembled polyps of the terminal ileum and colon. Chargelaigue in 1859 described a 16 year old girl and 21 year old man with polyadenomatosis (**Abramson, 1967**). In 1861, Luschka described a 30 year-old woman with myriads of colonic polyps. Also in 1861, Lebert published a report of a 32- year-old woman who suffered from diarrhea and at autopsy was found to have what was probably a true case of colonic polyps (Dukes, 1930). An excellent pathologic description of colon polyps was given by Virchow in 1863 based on material obtained from a 15-year-old boy. Virchow coined the term "colitis polyposis" implying an inflammatory nature to the polyps. In 1882 W. Harrison Cripps presented a paper before the Pathological Society of London establishing familial polyposis of the colon as a clinical entity and noting the familial nature of the disease and its malignatn potential. In subsequent papers Smith and Handford stressed the familial pattern of the

disease and the early malignant change of the polyps. In 1927 Cockayne was the first to state that multiple polyposis is inherited as a Mendelian dominant trait (Watne, 1987).

In a subsequent report Dukes and Lockhart-Mummery clearly established the autosomal dominant inheritance pattern for the disease, reporting its occurrence in ten families over three or four successive generations. Shortly thereafter McKenney described 21 patients in three families, all in their third generation of the disease. Friedell and Wakefield in 1943 described 49 members of a family descended to the third generation from a brother and sister with polyposis coli. In these families, 19 members had either polyposis coli or colorectal carcinoma. In 1947 Guptill collected 347 reported cases of polyposis coli from the world literature, to which he added five cases of his own. Four of Guptill's five patients had malignant degeneration and two had multiple cancer. Among the case reports of patients with polypsis coli was the occasional patient with associated extracolonic findings. The first report of such an association is credited to Devic and Bussey, who in 1912 described a woman with osteomas of the mandible, sebaceous cysts of the scalp, subcutaneous lipomas, and multiple polyposis coli. In 1935 Cabot presented the case of a 36-year-old man with multiple bony exostoses, a history of fibrosarcoma removed from three

ribs, multiple sebaceous cysts, and carcinoma of the ampulla of Vater. In 1937 Miller and Sweet described a 21-year-old woman with polyposis coli who had a desmoid tumor in the abdominal scar. Fitzgerald in 1943 described a 36-year-old woman with recurring desmoid tumors of the abdominal wall, bony exostoses, multiple compound odontomas, torus palatinus, and multiple polyposis of the bowel (Watne, 1987).

In a series of papers beginning in 1950, Eldon J. Gardner described the syndrome which bears his name. Gardner's syndrome includes soft tissue tumors (sebaceous cysts and fibromas), bony tumors (osteomas), and adenomatous polyps of the colon. The latter have the same growth pattern and malignant potential as the polyps seen in patients with familial polyposis. By 1967, MacDonald et al., were able to review 118 cases reported in the literature (MacDonald et al., 1967). They focused attention on the development of periampullary carcinoma as part of the syndrome.

In 1959 Turcot et al., described two siblings with documented familial polyposis of the colon: a 15 year-old boy who developed a medulloblastoma invading the medulla spinalis and a 13-year-old girl who died at age 21 from a left frontal glioblastoma; she also had a small chromophobe adenoma.

It is our belief now that the neoplastic polyposis syndromes, which include the classic Mendelian dominant familial polyposis of Cuthbert Dukes, Gardner's syndrome, Turcot's syndrome and discrete polyps and cancer, are all manifestations of an inherited neoplastic polyposis syndrome (**Watne, 1987**).

CHAPTER TWO

PATHOLOGY

OF

INTETINAL POLYPOSIS

Pathology of Intestinal Polyposis

Classification of intestinal polyps

Goligher et al., 1984 Classified intestinal polyposis into:

A. Neoplasms

Epithelial

Adenoma, including familial polyposis villous papilloma.

Other

Leiomyoma

Lipoma

Neurofibroma

Haemangioma

B. Hamartomas

Juvenile polyp

Polyps of Peutz-Jeghers syndrome.

C. Inflammatory

Ulcerative colitis

Segmental colitis

Crohn's disease

Dysenteries

Diverticulitis

Benign lymphoma

D. Unclassified

"Hyperplastic" or "metaplastic" mucosal polyp

Pneumatosis cystoides intestinalis

Hypertrophied anal papillae

Bland and Copeland 1986 Classified intestinal polyposis into:

I. Neoplastic polyps

A. Adenoma

Tubular adenoma (adenomatous)

Tubulovillous adenoma (papillary, mixed)

Villous adenoma

B. Familial Ademonatous Polyposis Syndromes

Multiple familial polyposis

Gardner's syndrome

Oldfield syndrome

Turcot's syndrome

Muir's (Torre's) syndrome

C. Non familial polyposis syndromes

Cronkhite-Canada Syndrome

II. Nonneoplastic polyps

A. Hamartomatous (mixed mesodermal) polyposes

Juvenile (mucous rentention) polyp