Carcinoma of the Colon, Ampulla of Vater and Urinary Bladder Associated with Familial Multiple Polyposis: A Case Report*

LT. COL. WILLIAM F. CAPPS, JR., USAF (MC), † MARTIN I. LEWIS, M.D. ‡
D. A. GAZZANIGA, M.D. §

Los Angeles, California

The association of multiple familial polyposis with other body lesions has been thoroughly explored by many investigators. Through a series of reports on familial association of tumors, Gardner's name was given to the syndrome linking multiple polyposis, exostoses, and subcutaneous lesions.5–7 11 Familiar polyposis, when associated with brain tumors, is known as Turcot's syndrome. Recently, Collins3 examined the relationship between familial polyposis and other body tumors and found that there was no definite consistent relationship. He agreed with Gardner and Plenk3 that a single pleomorphic gene may be responsible for the association of these multiple lesions and might represent a genetic linkage between two dominant genes.

While this definite relationship linking familial polyposis with other body lesions has not been established, it is most interesting to note that there are strong existing correlations. Polk and his associates10 reported that in approximately 50 per cent of all patients with primary cancers of the colon and rectum, a tumor will develop ultimately. Approximately 50 per cent of these secondary neoplasms will be carcinomas of the skin. In the remaining 50 per cent, the lesion will involve a wide variety of organ systems.

Further correlation is exemplified by a report from the Mayo Clinic, covering a ten-year period from 1944 to 1953 inclusive, analyzing 6,012 cases of carcinoma of the colon and rectum. This report shows the presence of multiple simultaneous primary malignant lesions of the colon in 157 cases (2.6 per cent). Twenty-four patients in this series had associated multiple familiar polyposis. 9 Bochetto and associates1 emphasized the well-known fact that, in almost 100 per cent of patients with neglected and untreated multiple polyposis, carcinoma of the colon will develop.

Not associated with familial polyposis, but pertinent to the case presented here-with, is the interesting phenomenon of spontaneous tumor regression. Everson and Cole,4 in 1956, reviewed medical literature on the subject of spontaneous regression of known carcinomas. They selected 47 cases from more than 600 published reports and documented examples of spontaneous regression of known neoplasms. They attempted to find a common factor which could explain or help predict spontaneous tumor regression, and concluded that tumor

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† Medical Service, USAF; presently assigned as Resident, Colon and Rectal Surgery, Queen of Angels Hospital.
‡ Department of Surgery, Queen of Angels Hospital; Attending Instructor, Queen of Angels Hospital Surgical Clinic; Ross-Loos Medical Group, Department of Surgery.
§ Department of Surgery, Queen of Angels Hospital; Section of Colon and Rectal Surgery, Service of Ross-Loos Medical Group.
Fig. 1. Shows strong family history of patient demonstrating four generations of both carcinoma of colon and polyps of colon.

Report of a Case

In 1954, a white boy, aged 14 years, was examined at a hospital in St. Louis because he was bleeding from the rectum. Sigmoidoscopic examination and x-ray of the colon after a barium enema revealed multiple polyposis. He had a very interesting family history of both multiple polyposis and carcinoma of the colon (Fig. 1). In October

Fig. 2. One of five adenocarcinomas found in resected colon was an adenocarcinoma in base of polyp with invasion of muscularis mucosae. Low-power view shows small area of adenocarcinoma at base of polyp (hematoxylin and eosin; X10).