Primary Adenocarcinoma of the Vermiform Appendix:

Report of Seven Cases and Review of the Literature*

G. BRUCE HOPKINS, M.D., RICHARD H. TULLIS, M.D.,
KAI A. B. KRISTENSEN, M.D.

From the Departments of Pathology and Surgery, Scripps Memorial Hospital, La Jolla, California

EPITHELIAL TUMORS of the appendix are classified as three types: adenocarcinoma, carcinoid, and mucocele. Of these, adenocarcinomas are by far the rarest. Since Beger's first description in 1882, only 158 cases have been recorded. This present report documents an additional seven cases and reviews the relevant literature.

Clinical and Pathologic Data

From January 1958 to December 1971, seven of 6,578 appendectomy specimens excised at Scripps Hospital were found to include primary adenocarcinomas. Clinical data are summarized in Table 1. The mean age of the patients at time of diagnosis was 63 years, with a range of 44 to 75 years. Six patients had signs and symptome of acute appendicitis or appendiceal abscess when first seen. One patient was seen because of progressive weight loss and inanition; peritoneal carcinomatosis of appendiceal origin was discovered at laparotomy. The diagnosis was not suspected preoperatively in any case.

Four of the seven tumors arose in polypoid fashion near the base of the appendix (Figs. 1 and 2). Three were primarily ulcerative and occurred near the tip. All were invasive and histologically resembled colonic adenocarcinoma. Continuity of the tumor with the appendiceal mucosal surface was established in each case.

General Discussion

Primary adenocarcinomas of the appendix have variously been reported to be present in 0.03 to 0.08 per cent[2] of all appendices removed. The incidence in our series was 0.1 per cent. It is primarily a disease of older age groups, with the sixth decade of life predominating (Table 2). Of the 165 patients reported, 102 (62 per cent) were male. One hundred ten patients (67 per cent) were first seen because of symptoms of acute appendicitis or appendiceal abscess; in 11 cases (7 per cent) the tumors were found following appendectomies incidental to other procedures (Table 3). The diagnosis has not been established preoperatively in any reported case.

Adenocarcinomas may occur at the base or tip of the appendix, although they are more frequent at the base. Grossly, they are polypoid or ulcerative, and microscopically they resemble adenocarcinoma of the colon. Primary cecal tumors or mucoceles are excluded by demonstrating invasive malignant glands lying in continuity with the appendiceal mucosal surface.[3]

Like other colonic carcinomas, these adenocarcinomas metastasize by local invasion, via lymphatics and by the blood stream. However, since the muscle layers

* Received for publication March 6, 1972.

Address reprint requests to: G. Bruce Hopkins, M.D., Pathology Department, Scripps Memorial Hospital, La Jolla, California 92037.

TABLE 1. *Clinical Summary of Seven Cases*

	(Years), Sex Age	Preoperative Diagnosis	Operative Findings	Operative Treatment	Results
Patient 1	56, M	Acute appendicitis	Appendicitis, appendiceal adenocarcinoma	Appendectomy, right colectomy a week later	Alive and well nine years after operation
Patient 2	72, M	Acute appendicitis	Appendiceal abscess, appendiceal adenocarcinoma with peritoneal carcinomatosis	Appendectomy palliative right colectomy two weeks later	Died of metastases three weeks after operation
Patient 3	75, M	Acute appendicitis, partial small bowel obstruction	Appendiceal abscess, partial ileal obstruction due to serosal infiltration of appendiceal adenocarcinoma	Appendectomy, partial small-bowel resection	Died of metastases six months after operation
Patient 4	52, M	Malignancy, origin unknown	Appendiceal adenocarcinoma with peritoneal and mesenteric carcinomatosis	Palliative appendectomy	Lost to follow-up
Patient 5	74, M	Appendiceal abscess	Appendiceal abscess, appendiceal adenocarcinoma	Appendectomy, right hemicolectomy three weeks later	Alive and well a year after operation; lost to follow-up since
Patient 6	44, F	Appendiceal abscess	Appendiceal abscess, appendiceal adenocarcinoma	Appendectomy, right hemicolectomy three months later	Alive and well nine months after operation
Patient 7	68, F	Acute appendicitis	Appendicitis, appendiceal adenocarcinoma	Appendectomy	Alive and well nine months after opertion

of the appendix may be incomplete or absent, direct extension into adjacent structures can occur quite early. In some instances submucosal invasion is actually

TABLE 2. *Age Distribution, 165 Patients*

Age (Years)	Number of Patients
10–19	1
20–29	4
29–39	14
39–49	32
49–59	28
59–69	49
69–79	32
79–89	5

TABLE 3. *Preoperative Symptoms or Diagnosis, 165 Cases*

Acute appendicitis	77
Appendiceal abscess	33
Incidental appendectomy	11
Abdominal cutaneous fistula	6
Carcinoma of the cecum	6
Right lower quadrant pain	6
Metastatic carcinoma	3
Right lower quadrant mass	3
Carcinoma of the prostate	2
Uterine fibroid	2
Hydronephrosis	2
Cholecystitis	2
Ovarian tumor	2
Other	10

FIG. 1. Polypoid adenocarcinoma arising from the mucosal surface. Note the resemblance to colonic adenocarcinoma (hematoxylin and eosin, × 100).

subserosal extension.[7] The initial lymphatic spread of these tumors is to the ileocolic nodal basin, infraduodenal and para-aortic nodes.[5]

In contrast, carcinoids rarely metastasize, and fewer than 10 per cent of mucoceles manifest "malignant" characteristics.[6]

Simple appendectomy for primary ap-

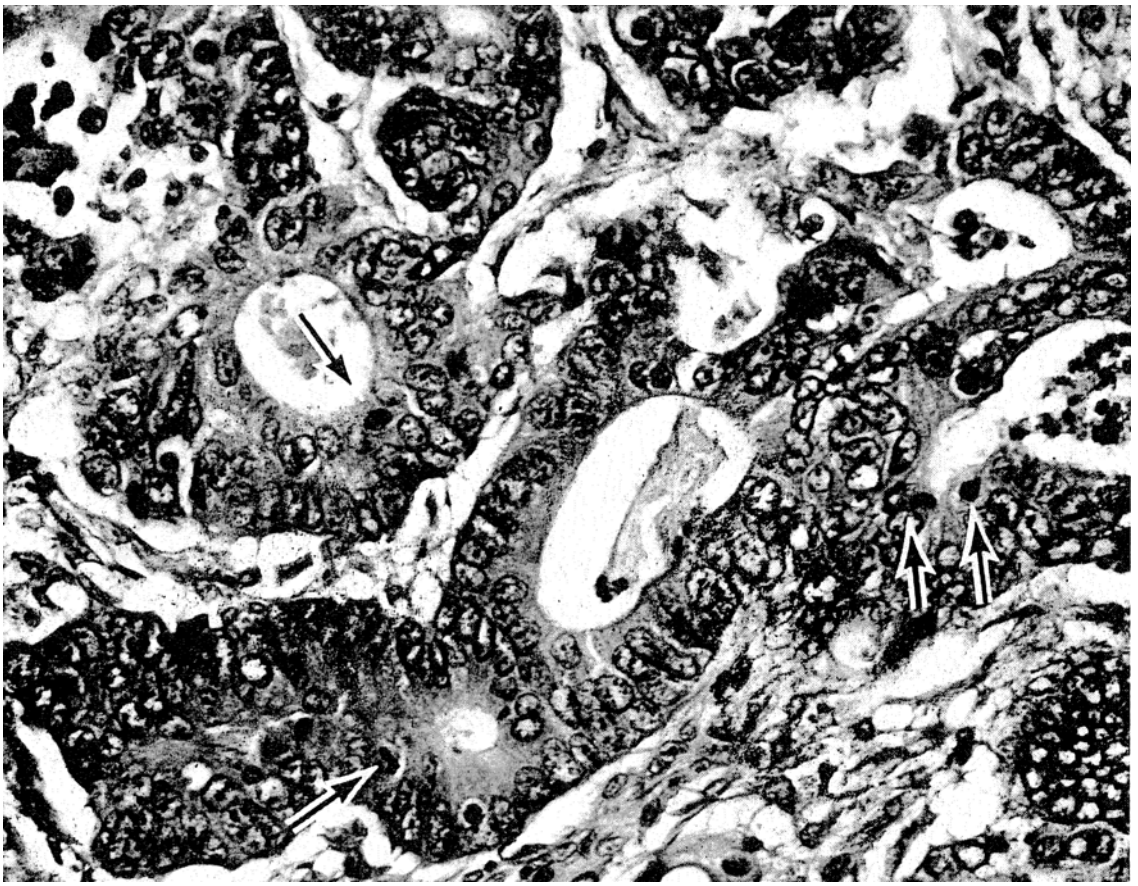

FIG. 2. Higher magnification of adenocarcinoma shown in Figure 1, showing the malignant glands. Note the numerous mitotic figures (hematoxylin and eosin, × 450).

pendiceal adenocarcinomas has proven inadequate. Death from metastases has occurred in 48 per cent of those treated with appendectomy, as opposed to 18 per cent of those treated with appendectomy and right hemicolectomy (Table 4). Perforation does not appear to alter survival figures significantly. Thus, once a diagnosis of adenocarcinoma is established, the procedure of choice is either primary or secondary right hemicolectomy with node dissection.

TABLE 4. *Survival of 165 Patients Following Treatment for Adenocarcinoma of the Appendix*

	Number of Patients*	Patients Surviving Five Years		Deaths from Metastases	
		Number	%	Number	%
Appendectomy	65	13	20	31	48
Appendectomy and hemicolectomy	71	32	45	13	18

* Patients with metastases on initial presentation were excluded from survival and death figures.

Summary

There have been 158 recorded cases of adenocarcinoma of the appendix reported in the literature. Seven additional cases are documented. The great majority of patients have symptoms of acute appendicitis or appendiceal abscess when first seen.

Appendectomy with right hemicolectomy yields the best long-term survival rates.

Acknowledgment

The authors wish to express their appreciation for the invaluable assistance of Mrs. Lois Bennett and Miss Kay McGuire in the preparation of this manuscript.

References

1. Beger A: Ein Fall von Krebs des Wurmfortsatzes. Berlin Klin Wochenschr 19: 616, 1882
2. Collins DC: 71,000 human appendix specimens: A final report, summarizing forty years' study. Am J Proctol 14: 365, 1963
3. Guarino GB, Chitwood EM Jr: Adenocarcinoma of the appendix: With a review of recent literature. Am J Surg 87: 293, 1954
4. Hesketh KT: The management of primary adenocarcinoma of the vermiform appendix. Gut 4: 158, 1963
5. Jamieson JK, Dobson JF: The lymphatic system of the caecum and appendix. Lancet 1: 1137, 1907
6. Raiford TS: Treatment of tumors of the appendix, Treatment of Cancer and Allied Diseases. Second edition. Volume 5. Edited by GT Pack, IM Ariel. New York, Paul B. Hoeber, Inc., 1962, p 296
7. Sieracki JC, Tesluk H: Primary adenocarcinoma of the vermiform appendix. Cancer 9: 997, 1956