

Adenocarcinoma of the Appendix

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Ferro M, Anthony PP. Adenocarcinoma of the appendix. *Dis Colon Rectum* 1985;28:457-459.

Three cases of adenocarcinoma of the appendix are reported. All three patients presented with acute appendicitis and the tumors were diagnosed only on histologic examination of the excised appendix. The first patient subsequently had a right hemicolectomy and was proven to have a Dukes' B tumor. The second patient probably had a Dukes' B also, but no further surgery was performed because of advanced presenile dementia. Advanced disease was found in the third patient. Analysis of 145 cases reported over the last ten years suggests that, unless the tumor is in Dukes' A stage, right hemicolectomy should be carried out if the patient is fit for radical surgery. The overall prognosis appears to be the same as that for carcinoma of the colon. [Key words: Adenocarcinoma; Appendix; Treatment]

ADENOCARCINOMA of the appendix is an infrequently encountered surgical condition. In a recent review of the literature, some 250 cases were found to have accumulated since the first report in 1882.¹ Its incidence is quoted as 0.01 to 0.2 percent of resected appendices examined histologically, which is the same as that for small-bowel adenocarcinoma. Though the condition is rare, its recognition is important, as radical surgery may be life-saving. We wish to report three cases that occurred during a three-month period in the Exeter Health District (population, 292,000). This can only be a coincidence by chance, but the lessons to draw for the management of individual patients remain the same. To this effect, we analyzed 145 cases recorded in the ten-year period 1974 to 1984.¹⁻⁷

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Reports of Cases

Patient 1: A 54-year old man presented with a 24-hour history of pain in the right iliac fossa, associated with nausea and anorexia but no vomiting. On examination, he was pyrexial with tenderness and rebound in the right iliac fossa. Rectal examination also revealed tenderness on the right side.

Appendectomy was performed through a gridiron incision. A grossly inflamed appendix was found in a retrocecal position and had to be removed piecemeal. Postoperative recovery was uneventful. Histology showed gross inflammation with a coexistent, moderately differentiated, mucus-secreting adenocarcinoma that had breached the appendiceal wall and was present in the surrounding fat.

A right hemicolectomy was carried out five weeks later. Nodules in the perirenal fat and enlarged lymph nodes at the ileocecal junction were excised. Histology showed that the nodules in the perirenal fat consisted of residual tumor, but the mesenteric nodes were free.

The patient is well and free of disease ten months after hemicolectomy.

Patient 2: A 62-year-old woman was admitted as an emergency with a two-day history of pain in the right iliac fossa. She had had a similar episode of pain two weeks previously that had resolved spontaneously. She was known to suffer from severe presenile dementia, which made assessment of physical signs difficult. She was pyrexial with a temperature of 102.2°F and vague tenderness in the right iliac fossa.

She was diagnosed as having acute appendicitis and appendectomy was performed through a gridiron incision. The appendix was found to be inflamed and gangrenous with perforation at its junction with the cecum. Histology showed a grossly inflamed appendix. In addition, a well-differentiated adenocarcinoma was present, which had clearly arisen from a preexisting villous adenoma (Fig. 1). Invasion of the wall was present through to the subserosa, but there was no further spread. The proximal end of the appendix was tumor-free.

No further surgery was undertaken in view of her poor mental state. She remains free of disease six months after appendectomy.

Patient 3: A 64-year-old woman presented as an emergency with a 24-hour history of pain in the right iliac fossa associated with anorexia and nausea. The pain was aggravated by coughing and movement. Past medical history included a triple coronary artery bypass in the

Received for publication December 4, 1984.

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TABLE 1. *Clinical Presentation of 145 Cases of Adenocarcinoma of the Appendix*

Acute appendicitis	67
Appendiceal mass	25
Incidental discovery at surgery for other conditions	11
Intestinal obstruction	8
Ascites	6
Right pelvic mass with vaginal bleeding	3
Hematuria and fecaluria	3
Cutaneous mass in right iliac fossa	2
Krukenberg tumors	2
Postmenopausal bleeding	1
Umbilical mass	1
Virchow's node	1
Not stated	15
TOTAL	145

preceding year. This had improved her cardiac symptoms to a minimal degree, but she still had angina and dyspnea on mild exertion and was in congestive heart failure on admission. On examination, she had a temperature of 99.8°F with tenderness and rigidity in the right iliac fossa and tenderness on the right side of the pelvis on rectal examination.

Appendectomy was performed through a gridiron incision. A perforated gangrenous appendix was excised. Histology showed gross acute inflammation. A moderately differentiated adenocarcinoma had breached the wall, and tumor cells were present in the mesoappendiceal lymphatics.

Postoperative recovery was good; however, her cardiac state gave cause for concern and radical surgery was considered inadvisable. She was readmitted six months later with subacute intestinal obstruction. Disseminated abdominal carcinomatosis was found at laparotomy and no surgical procedure was carried out. She was discharged home for terminal care.

Analysis of the Literature

A computer-aided search of the literature for the years 1974 to 1984 revealed 31 reports from which 145 cases were extracted and analyzed. The list of references¹⁻⁷ contains major articles that include references to all the case reports.

The male-to-female ratio was 3 to 2 and the age range 18 to 88 years (average, 60 years). The clinical presentation is summarized in Table 1. The most common presentation was acute appendicitis or an appendiceal mass, which accounted for 92 of 130 cases where the mode of presentation was documented. Diagnosis was never made

prior to histologic examination, though the appendix was noted to be abnormal, in addition to being inflamed, in some cases. The pathology of the tumor was reported as colonic type adenocarcinoma in 84 patients, cystadenocarcinoma in 57, and there were four patients with diffuse infiltration consisting mainly of signet-ring cells (linitis plastica).

Analysis of the effectiveness of surgical treatment was carried out based on Dukes' staging system in 80 cases where this information was available (Table 2). Of the remainder, only palliative treatment was performed in 37 patients and they all died of the disease subsequently. Twenty patients were excluded due to insufficient information; seven patients died of other incidental causes and one tumor was first diagnosed at postmortem. In the analysis, local treatment refers to appendectomy alone and radical treatment to right hemicolectomy extended, in some cases, to removal of other involved viscera, e.g., bladder, uterus or ovary.

The mortality rate was low among patients with Dukes' A tumors: only one of 18 died. In the group of 46 patients with Dukes' B tumors, the mortality rate was higher among those who did not undergo radical surgery. The mortality rate in patients with Dukes' C tumors was highest, despite radical surgery. It must be admitted that descriptions of operative findings and procedures vary in details and that some patients, who had only local treatment, may, in fact, have had lymph node metastases. Overall, however, there does appear to be a correlation between Dukes' stage and prognosis and the figures are similar to those for adenocarcinoma of the colon,⁸ given the fact that survival up to five years was recorded in only approximately 50 percent of cases.

An associated malignant tumor was found in 16 cases (11 percent) at the same time as appendicectomy was performed. These were carcinomas of the colon (four), cervix (three), breast (two), prostate (two), esophagus (one), stomach (one), ovary (one), and bladder (one); there was also one case of lymphoma.

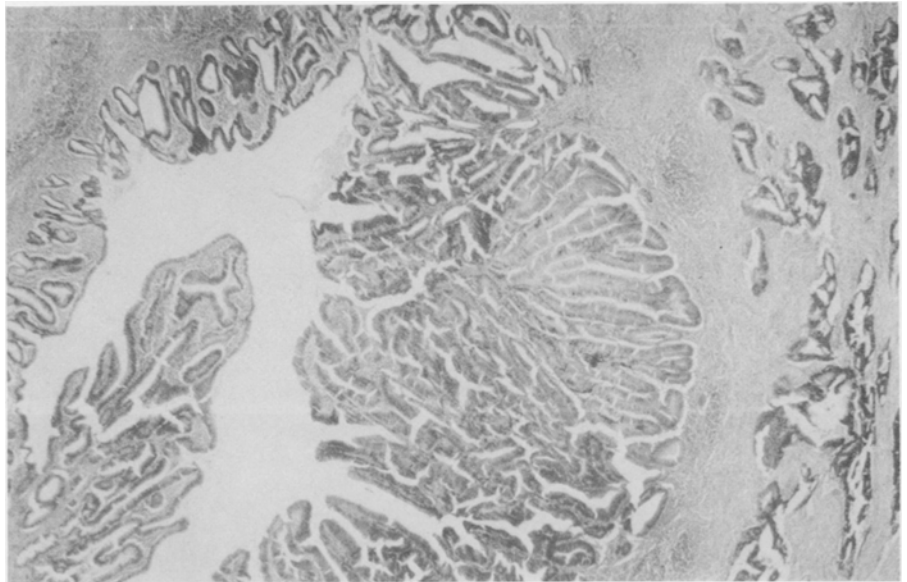
Discussion

These three cases conform with those already on record as regards age, sex, presentation, pathology, associated

TABLE 2. *Analysis of Results of Surgical Treatment by Dukes' Stages in 80 Cases of Adenocarcinoma of the Appendix*

Dukes' Stage	Number of Patients	Treatment		Deaths	Survival (Percent)
		Local	Radical		
A	18	9	—	—	100
		—	9	1	89
B	46	14	—	4	71
		—	32	4	87
C	16	—	—	—	—
		—	16	9	43

FIG. 1. Most of the lumen of the appendix is occupied by a villous adenoma. Malignant change has supervened with invasion of the muscle wall on the right (hematoxylin and eosin; $\times 25$).



malignancies, and treatment.¹⁻⁸ The condition is rarely suspected, let alone recognized, prior to histologic examination of the excised appendix. It is considered useful, therefore, to discuss briefly the implications of a chance discovery of adenocarcinoma of the appendix.

The most common presentation is appendicitis or an appendicular mass. It has not been possible to determine whether this, in itself, is a beneficial or adverse factor. The only possible conclusion is that, in the absence of an inflammatory episode, the tumor may remain asymptomatic until it presents with spread to some other abdominal or pelvic organ or, rarely, to skin.

As far as the limited information on Dukes' stage is concerned, most patients present with advanced disease. Appendectomy alone is probably sufficient in Dukes' A stage but, once the appendiceal wall has been breached, it is impossible to ascertain the presence or absence of ileocecal lymph-node involvement, *i.e.*, whether the tumor is Dukes' B or C; therefore, a right hemicolectomy should be carried out.

The pathology of adenocarcinoma of the appendix is similar to that of the colon.⁸ One of our three patients had a preexisting adenoma from which carcinoma arose: this has been noted previously in a number of cases.¹⁻⁴ The high incidence of cystadenocarcinoma is due to the anatomic peculiarities of the appendix: even small tumors may occlude the lumen, producing a cyst-like swelling due to continuing secretion of mucus. The rarity of other

histologic types, *i.e.*, signet-ring celled or diffusely infiltrating, corresponds to their rarity in the colon, also.⁸

The prevalence of associated malignant tumors is difficult to assess, but probably calls for thorough examination of the abdomen at the time of definitive surgery.

Though any surgeon is unlikely to encounter an appendiceal adenocarcinoma more than once, it should not be regarded in the same way as the much more common and usually benign carcinoid tumor, and early consideration should be given to radical right hemicolectomy once the diagnosis is known.

References

1. Gilhorne RW, Johnstone DH, Clark J, Kyle J. Primary adenocarcinoma of the vermiform appendix: report of a series of ten cases and review of the literature. *Br J Surg* 1984;71:553-5.
2. Quizilbash AH. Mucocoeles of the appendix: their relationship to hyperplastic polyps, mucinous cystadenomas and cystadenocarcinomas. *Arch Pathol* 1975;99:548-55.
3. Quizilbash AH. Primary adenocarcinoma of the appendix. A clinicopathological study of 11 cases. *Arch Pathol* 1975;99:556-62.
4. Gamble HA II. Adenocarcinoma of the appendix: an unusual case and review. *Dis Colon Rectum* 1976;19:621-5.
5. Wolff M, Ahmed N. Epithelial neoplasms of the vermiform appendix (exclusive of carcinoid): I. Adenocarcinoma of the appendix. *Cancer* 1976;37:2493-510.
6. Didolkar MS, Fanous N. Adenocarcinoma of the appendix: a clinicopathologic study. *Dis Colon Rectum* 1977;20:130-4.
7. Waizbard E, Michowitz M, Baratz M, Papo J, Stavorovsky M. Unusual presentation of carcinoma of the vermiform appendix: a report of two cases. *J Surg Oncol* 1984;25:263-7.
8. Morson BC, Dawson IM. *Gastrointestinal pathology*. 2nd ed. Oxford: Blackwell Scientific Publications, 1979:447-5.