Colonic Type Adenocarcinoma of the Appendix: Report of a Case and Review of Japanese Literature

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Citation

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Abstract

We report a case of “colonic type” adenocarcinoma of the appendix in a 35-year-old man. The preoperative diagnosis was acute appendicitis and emergent appendectomy was performed. Appendiceal cancer was diagnosed pathologically, and the stump was found to be positive for cancer. Right hemicolectomy was undertaken as a secondary procedure after the first operation due to the involvement of the cecum and lymph nodes. The histopathological diagnosis was well and moderately differentiated adenocarcinoma of the appendix. After the operation, the patient underwent chemotherapy. He has survived for 30 months after the colonic resection without any signs of recurrence.

INTRODUCTION

Primary adenocarcinoma of the appendix is relatively rare, contributing to less than 0.5% of all gastrointestinal malignancies (1,2) and with an incidence of approximately 0.08%-0.1% of appendectomy specimens (3,4). A search of Ichushi Web, a Japanese medical database, revealed 77 Japanese patients diagnosed postoperatively with “colonic type” adenocarcinoma in the 10 years between 1999 and 2008 (Table 1). Median survival after abdominal surgery for 16 of the 77 cases was 587.3 days (range 48-2129 days), and the prognosis was not good. Since appendiceal cancer is difficult to diagnose preoperatively, postoperative histopathologic examination may be used to establish a definite diagnosis. We report here a case of colonic type adenocarcinoma of the appendix diagnosed after appendectomy.

CASE REPORT

A 35-year-old man was referred to our hospital in April 2008 with suspected acute appendicitis. On the day of admission, he had consulted another hospital with a history of right lower abdominal pain.

He was 167cm in height, weighed 112kg, and had a body mass index (BMI) of 40.2kg/m^2. He had been smoking 20 cigarettes daily for the past 15 years. Physical examination revealed right lower abdominal tenderness. His body temperature was 37.4□. His white blood cell (WBC) count was 15,000/□L and C-reactive protein (CRP) was 12.1mg/dL. Abdominal ultrasonography showed an enlarged appendix measuring 3×3cm in diameter and an abdominal computed tomographic (CT) scan revealed appendiceal swelling, spanning an area of 7×3cm at the cecal region (Fig. 1). We suspected that the preoperative diagnosis was acute appendicitis.

Emergent appendectomy was performed on April 11, 2008. The appendix measured 9×5cm and a tumor measuring 6×3cm was revealed in the mucosa (Fig. 2A, B).

Microscopic examinations demonstrated both well and moderately differentiated adenocarcinoma (Fig. 3A, B).

The tumor was initially positive on the edge, and thus, right hemicolectomy was performed on May 12, 2008. Macroscopy revealed a tumor, which invaded concentrically from the orifice of the appendix (Fig. 4). The histological diagnosis was well and moderately differentiated adenocarcinoma similar to that observed on the appendix (Fig. 3C, D). Finally, it was diagnosed as appendiceal adenocarcinoma. Pathology revealed lymph node metastasis, which was staged as T2N1M0 (stage IIIa).

Following colonic resection, chemotherapy was administered. The patient has survived for 30 months since

the right hemicolectomy.

Figure 1

Figure 1: Abdominal computed tomography scan finding. An enlarged appendix (white arrows), 7×3cm in size was found in the right pelvic cavity.

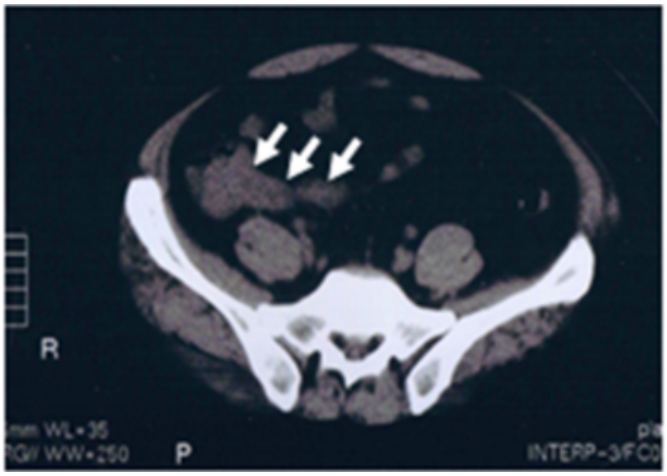

Figure 2

Figure 2: The surgical specimen of the appendix. The elevated tumor was 6×3 cm in size at the mucosal level (white arrows) (A). The tumor after formalin fixation (black arrows) (B).

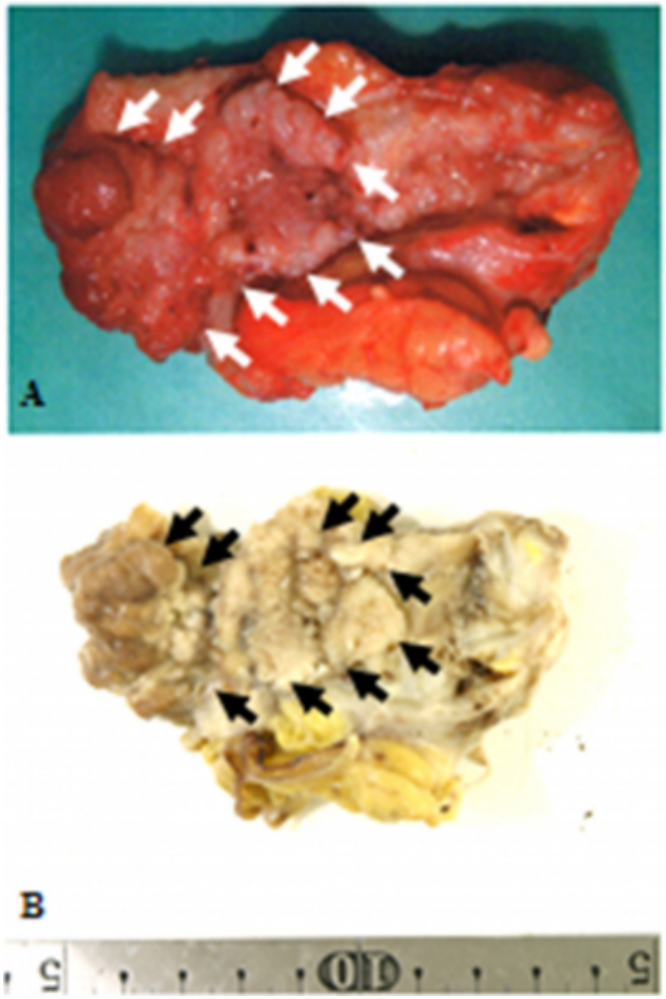

Figure 3

Figure 3: Histological view. Microscopy of the appendiceal tumor with hematoxylin and eosin stain (Ã—12.5) (A). Pathology revealed both well and moderately differentiated adenocarcinoma on the appendix (Ã—200) (B). Microscopy of the cecal tumor (Ã—12.5) (C). This cecal tumor showed the same pathology as the appendix (Ã—200) (D).

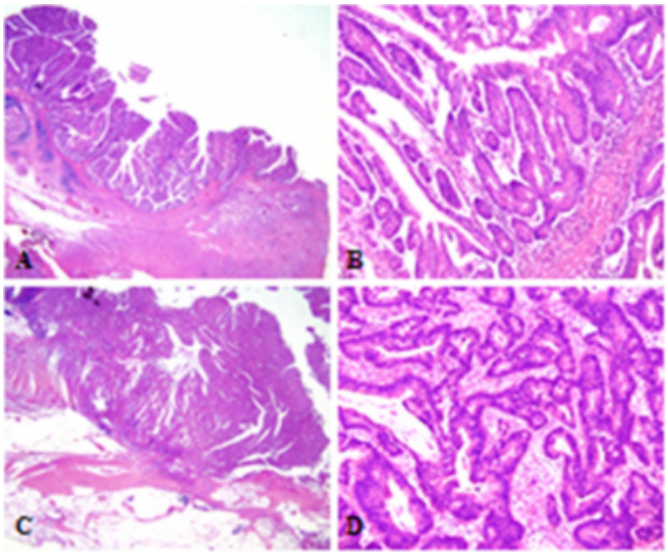

Figure 4

Figure 4: Macroscopic finding after right hemicolectomy. A tumor (white arrows), 3.5×3 cm in diameter at the cecum, invaded concentrically from the orifice of the appendix.

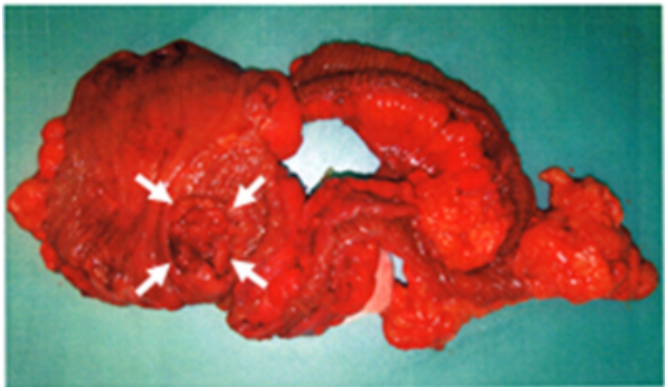

Figure 5

Table 1. Japanese cases of colonic type appendiceal cancer between 1999 and 2008 based on a search of Ichushi Web

Case numbers	77				
Gender	M 27, F 50				
Age (years) (range)	63.2±15.0 (21-88)				
Chief complaint (%)					
Rt.lower abd. pain	29 (38)	Appetite loss	3 (4)		
FOBT positive	6 (8)	Anemia	3 (4)		
Screening	4 (5)	Others	15 (19)		
Epigastralgia	4 (5)	U	13 (17)		
Body temper(℃) (range)	37.0±0.7	(35.9-38.6)			
WBC (/μL)	11.5±7.0×10³	(2.9-35.8)			
CRP (mg/dL)	9.3±9.0	(0.1-32.5)			
CEA (ng/mL)	55.7±189.6	(0.7-958)			
CA19-9 (U/mL)	150.1±267.1	(2.0-1051)			
Preoperative diagnosis (%)					
Appe. cancer	25 (32)	Asc. colonic ca.	4 (5)		
Acute appendicitis	24 (31)	Ovarian tumor	3 (4)		
Cecal tumor	12 (16)	Others	9 (12)		
Type of surgery (%)					
Cecectomy	36 (47)	Rt.hemicolectomy	11 (14)		
Appendectomy	18 (23)	Others	12 (16)		
Lymph nodes involved (%)					
Negative	55 (71)				
Positive	20 (26)				
U	2 (3)				
Histology (%)					
Well differ. adenocarcinoma	37 (48)				
Moderately differ. adenocarcinoma	18 (23)				
Poorly differ.adenocarcinoma	14 (18)				
Others	8 (11)				
pStaging (%)					
0	12 (16)	IIIb	3 (4)		
I	11 (14)	IV	16 (21)		
II	18 (23)	U	7 (9)		
IIIa	10 (13)				
Prognosis (U: 20) (median; range)					
Alive	41	(727.9±833.1; 6-4380 days)			
Death	16	(587.3±527.0; 48-2129 days)			
Cause of mortality (16 cases)					
P.C.	7	Brain meta.	1		
Hepatic meta.	1	Jejunal meta.	1		
Pulmonary meta.	1	U	5		
pStaging for mortality (16 cases)					
II : 4, IIIa : 2, IV : 8, U : 2					
0-II : 4, III-IV : 10 (71.4%)					
pStaging for survivor (41 cases)					
0 : 6, I : 8, II : 12, IIIa : 4, IIIb : 3, IV : 4, U : 4					
0-II : 26, III-IV : 11 (29.7%)					
(20 cases unknown on alive or dead)					

M, male; F, female; Rt, right; Abd, abdominal; FOBT, fecal occult blood test; U, unknown; Temper, temperature; Appe, appendiceal; Asc, ascending; Ca, cancer; Differ, differentiated; P, pathological; P.C, peritoneal carcinomatosis; Meta, metastasis

DISCUSSION

The reported incidence of adenocarcinoma of the vermiform appendix represents approximately 0.08%-0.1% of appendectomy specimens (3,4). Only about 250 cases of primary adenocarcinoma of the appendix have been described since Berger first recognized the neoplasm in 1882. The International Classification of Diseases for Oncology Group classifies adenocarcinoma of the appendix into three categories: colonic, mucinous, and signet ring cell carcinoma (5,6). The colonic type adenocarcinoma is the least common of the three types. Its mucosa consists of acinar, tubular, or papillary structures that resemble adenocarcinoma of the colon (7-9), and in gross appearance, it is polypoid or ulcerative (10,11).

Right lower abdominal pain is the frequent chief complaint and clinical symptom in Japanese patients with colonic type adenocarcinoma of the appendix (Table 1). In colonic type adenocarcinoma, symptoms of acute appendicitis may be due to obstruction of the lumen by the tumor, infiltration by the tumor, infection superimposed on the wall, obstruction of lymphatic channels, obstruction of vasculature, or intussusception (12).

Preoperative diagnosis of adenocarcinoma of the appendix is difficult because of the lack of definite diagnostic, clinical, sonographic, or radiological findings characteristic of this disease (2, 13). Of the recorded Japanese cases, appendiceal carcinoma has been diagnosed preoperatively only in 32% of 77 patients (Table 1).

Hemicolectomy has been reported to be the most curative operation. Simple appendectomy is inadequate for primary appendiceal adenocarcinomas. The 5-year survival in 165

patients with primary adenocarcinomas of the appendix was reported to be 20% for appendectomy and 45% for hemicolectomy. Death from metastases occurred in 48% of 65 patients treated by appendectomy compared to 18% in those treated by appendectomy and right hemicolectomy (11, 14). Median survival after abdominal surgery for 16 of the 77 recorded Japanese cases was 587.3 days (range 48-2129 days), and the prognosis was not good. The percentage of the III and IV stages among the 16 patients was 71.4% in the reported Japanese cases (Table 1).

In conclusion, adenocarcinoma of appendix is relatively rare and often presents at an advanced stage. Despite surgery and adjuvant treatment, the prognosis remains poor. In cases of acute appendicitis, it is important to consider the possibility of appendiceal adenocarcinoma.

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