Case Report

A Case of Primary Mucinous Cystadenocarcinoma of the Appendix with Elevated Serum Carcinoembryonic Antigen (CEA)

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We report a 69-year-old female patient with mucinous cystadenocarcinoma of the appendix and elevated serum carcinoembryonic antigen (CEA). Serum CEA levels were 27.6–37.8 ng/ml. An oval shaped cystic mass at the ileocecal region was consistently seen by several radiologic procedures such as, barium enema, ultrasonography, computed tomography, and magnetic resonance imaging. The tumor was resected, and the histopathologic findings indicated mucinous cystadenocarcinoma of the appendix. CEA was demonstrated in the neoplastic columnar cells in immunohistochemical studies. The literature on the determination of serum CEA level in adenocarcinoma of the appendix is reviewed.

Key words: Immunohistochemical studies

Primary mucinous cystadenocarcinoma of the appendix is rare (1, 2) and pre-operative diagnosis is difficult (3, 4). To date, only 16 patients with adenocarcinoma of the appendix with determination of serum carcinoembryonic antigen (CEA) (4–18) have been reported in Japan. We report a patient with mucinous cystadenocarcinoma of the appendix with elevated serum CEA; the tumor production of CEA was demonstrated by immunohistochemical techniques.

CASE REPORT

A 69-year-old woman was admitted to Gifu University Hospital on April 21, 1988, because of traumatic right femoral neck fracture. During the postoperative period she frequently complained of diarrhea, and an elevated serum CEA level (29.0 ng/ml, normal range: less than 5 ng/ml) was found for the first time on May 9, 1988. Serum CEA levels gradually increased to 37.8 ng/ml. A smooth and tender mass (6 cm) was palpated in the right ileac region. Although other tumor markers were un-

remarkable, repeated serum CEA determination consistently showed an elevated value (27.6 ng/ml, Table 1). Barium enema examination demonstrated a smooth-surfaced defect at the ileocecal region, and the appendix was not demonstrated (Fig. 1). Ultrasonography (US) of the lower abdomen showed an oval shaped cystic mass with papillary growth of the cystic wall at the same area (Fig. 2). By computed

Table 1. Laboratory data on admission.

RBC ($\times 10^{12}/l$)	4.16	Na (mmol/l)	146
WBC ($\times 10^9/l$)	5.8	K (mmol/l)	3.5
Plt ($\times 10^9/l$)	164	Cl (mmol/l)	103
TP (g/l)	68	ESR (mm/h)	9
Alb (g/l)	40	CRP	(-)
AST ($\mu\text{kat}/l$)	0.28	CEA (ng/ml)	27.6
ALT ($\mu\text{kat}/l$)	0.16	AFP (ng/ml)	< 5.0
LDH ($\mu\text{kat}/l$)	5.5	CA19-9 (U/ml)	6.5
ALP ($\mu\text{kat}/l$)	3.7	CA125 (U/ml)	11
TBil ($\mu\text{mol}/l$)	17.1		
Tchol (mmol/l)	3.9		
TG (mmol/l)	1.0		
FPG (mmol/l)	4.7		

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Fig. 1. Barium enema demonstrating the defect (4.5×4.1 cm) in the ileocecal region. Appendix was not detected.

Fig. 2. Ultrasonography of the lower abdomen demonstrating a low echoic, heterogeneous, and cystic mass at the ileocecal region.

Fig. 3. Computed tomography of the lower abdomen demonstrating a low density, homogeneous, and cystic mass with capsule enhanced by contrast at the ileocecal region.

Fig. 4. Magnetic resonance imaging of the lower abdomen delineating a cystic mass that shows low intensity on T1-weighted sequences and high intensity on T2-weighted sequences.

tomography (CT), the tumor presented as a homogeneous cystic mass with a capsule which was enhanced by contrast medium (Fig. 3). Magnetic resonance imaging (MRI) delineated the cystic mass,

showing low intensity on T1 weighted sequences and high intensity on T2 weighted sequences (Fig. 4). A diagnosis of ileocecal tumor was made and the tumor was resected. In the gross findings, the tumor was solid with a smooth surface and $7.5 \times 3.5 \times 3$ cm in size, originating from the appendix. There was no connection with the cecum nor adhesion to the colon. The distended appendiceal lumen was filled with mucinous material. On histological examina-

tion, neoplastic columnar cells formed papillary structures and showed evidence of mucin secretion (Fig. 5A). Histological diagnosis was mucinous cystadenocarcinoma of the appendix. The neoplastic columnar cells were positive for CEA by immunohistochemical techniques according to avidin-biotinyl enzyme complex method (Fig. 5B). One week after operation, the serum CEA value decreased to 7.6 ng/ml. The patient and her family did not consent

Fig. 5A. Microphotograph of the tumor showing neoplastic columnar cells forming papillary structure and evidence of mucin secretion ($\times 100$).

Fig. 5B. Columnar cells, positive for CEA by immunohistochemical technique. The cells were stained brown ($\times 200$).

to right hemicolectomy. She is being followed up regularly and is well until the present time, and her serum CEA value is 3.8 ng/ml.

DISCUSSION

Primary malignant epithelial tumors of the appendix are uncommon (19) and primary adenocarcinoma of the appendix are rare (1). Due to the rarity of the disease and the difficulty to pinpoint the anatomical origin of the tumor of the appendix, often preoperatively a misdiagnosis is made as in the present case. Here, the diarrhea and elevated serum CEA led to the detection of an ileocecal tumor. The clinical significance of the latter in the diagnosis of adenocarcinoma of the appendix is still unclear. To date there have been sixteen reported cases in Japan in whom the serum CEA level was determined

(4–18). As shown in Table 2, preoperatively 4 of these 16 cases had an elevated serum CEA (6, 14, 16, 18). Among them, one (14) was the colonic type, one (6) the cystic type and the other two (16, 18) the cystic type complicated with pseudomyxoma peritoneal (Table 2). There was no correlation between the serum CEA level and histologic classification of the tumor. Immunohistochemical studies, although performed only in our case and in one other study (13), identified the presence of CEA in the resected tumor cells in both cases. In our patient, in addition to the evidence of tumor expression of CEA, the decrease in the level of serum CEA immediately after surgical resection strongly indicates that the elevated serum CEA was secreted from the tumor tissues. An normal serum CEA level was reported in the other patient with expression of

Table 2. Cases in Japan of adenocarcinoma of the appendix including evaluation of the serum CEA level.

Case No.	Age (year)	Sex	Histological diagnosis	Tumor size (cm)	Serum CEA (ng/ml)
1(5)*	56	M	cystic	10×10	1.7
2(6)	69	M	cystic	32×24×8	25.0
3(7)	61	F	colonic	3.6×3.3×2.5	1.1
4(8)	72	F	cystic	3.5×1.5	4.4
5(9)	31	M	colonic	3	not elevated
6(10)	53	F	cystic	6×2	5.0
7(11)	54	F	cystic	4.5×4.5	1.8
8(12)	72	M	cystic	3	2.2
9(13)	49	F	pseudomyxoma peritonei	ND	4.2
10(14)	59	M	colonic	5×6	10.8
11(4)	65	F	colonic	2	1.9
12(15)	30	F	colonic	3	1.0
13(16)	72	F	pseudomyxoma peritonei	ND	slightly elevated
14(17)	67	F	colonic	5	1.2
15(17)	63	F	cystic	not noted	0.9
16(18)	43	M	pseudomyxoma peritonei	ND	118.6

M, male; F, female; ND, not determined

*Reference number.

CEA in the tumor. Since other pathophysiological processes such as the release of CEA into the surrounding tissue (20), the entrance into the lymphatics and blood stream (21) are known to be involved in the mechanism of elevated serum CEA and the metabolic degradation and excretion by the liver (22) also influence the serum levels of CEA, the normal serum CEA levels reported in the majority of the patients does not necessarily exclude the tumor expression of CEA in these patients. Judging from the incidence of a positive test in the cases reported in Japan, the measurement of serum CEA does not seem to be sufficiently sensitive as a tumor marker for adenocarcinoma of the appendix. However, it seems important to take adenocarcinoma of the appendix into consideration in the differential diagnosis of neoplastic disorders associated with an elevated serum CEA. Right hemicolectomy is indicated for the treatment of adenocarcinoma of the appendix (2, 23). Unfortunately, in this patient right hemicolectomy was not performed, because she and her family did not consent to this therapy. She is well now but careful follow-up will be necessary.

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