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Abstract

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KEYWORDS: pseudomyxoma peritonei, ovarian origin

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PSEUDOMYXOMA PERITONEI OF OVARIAN ORIGIN —AN AUTOPSY CASE—

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Abstract. The autopsy case of an 80-year-old female with pseudomyxoma peritonei arising in the left ovary is reported. The patient was admitted with complaints of anorexia, sense of fullness and abdominal distension of two months' duration, and died of intestinal obstruction four months later. The autopsy revealed extensive tumor dissemination over the entire peritoneal cavity without any visceral invasion or distant metastasis. A part of the sigmoid colon showed marked stenosis and perforation with abscess formation. Histologically, the tumor was composed of various sized multiple cysts which were completely or incompletely lined by a layer of mucin-producing columnar epithelial cells with moderate nuclear hyperchromatism, and of a papillary pattern in some parts, indicating low grade malignancy.

Key words: pseudomyxoma peritonei, ovarian origin.

Pseudomyxoma peritonei is a relatively uncommon condition, which is characterized by massive accumulation of gelatinous ascites, lack of distant metastasis and long survival of the patients (1). Most reported cases have been of either ovarian or appendiceal origin (2-6). We present here the case of an 80-year-old woman with disease of ovarian origin, and report mainly the autopsy findings.

CASE PRESENTATION

Clinical Findings

An 80-year-old Japanese female was admitted to the Okayama Red-Cross Hospital on August 18, 1976 because of anorexia, general fatigue and a sense of abdominal fullness for the past two months. She had had hypertension pointed out five years earlier. The family history was not remarkable.

The physical examination on admission revealed intense abdominal distension with slight tenderness and many nodular masses palpable. X-ray examination with a retrograde procedure disclosed partial stenosis of the sigmoid colon due to compression by an external mass. From these findings, left ovarian

cancer with extensive dissemination was suspected. Cyclophosphamide therapy was initiated on the 34th hospital day, although the response was only temporary. She died of cachexia due to progressive enlargement of the tumor and of ileus on the 137th hospital day. The autopsy was performed two h after death.

Autopsy Findings (A.N. 1148)

Gross findings. The body was a female cadaver with cachexia and edema. The abdomen was highly distended, and large decubital wounds were noted in the back and sacral region.

On opening the pleural cavities, a clear yellowish effusion of 200 ml and partial fibrinous adhesion were noted in the left, and total fibrinofibrous adhesion in the right. The left lung weighed 330 g and the right 400 g. On sectioning, both atelectatic lower lobes contained a number of small hyperemic foci. The pericardial cavity contained a yellowish effusion of 30 ml. The heart weighed 250 g and showed slight concentric hypertrophy of the left ventricle. The myocardium was 1.4 cm in thickness with brown pigmentation.

Throughout the peritoneal cavity, massive tumor consisting of multiple mucinous nodules, 0.5 to 2 cm in diameter, involved all the organs such as liver, spleen and alimentary canal (Fig. 1), and filled all the free space of the cavity (Fig. 2). The descending colon was dilated as far as the stenotic part of the sigmoid colon with a perforation measuring 5 cm in diameter; this was accompanied by abscess, $12 \times 7 \times 6$ cm in size, and contained feces (Fig. 3). Near the abscess, the left ovary was entirely cystic measured $3 \times 2 \times 2$ cm with atrophic tuba (Fig. 4). This suggested that the left ovary was the probable primary site of the intraperitoneal mass. The right ovary and appendix were entirely embedded in the tumor mass. No direct invasion or distant metastasis of intra- or extraperitoneal organs was present.

The liver weighed 1000 g, and the cut-surface was rather cloudy and yellowish to reddish brown. Microscopically, central fatty change and congestion were observed. The pancreas and biliary tract were almost intact. The gallbladder contained a small pigmented stone. The wall of the gastrointestinal tract was generally edematous with mucosal hyperemia. The left kidney weighed 110 g, and the right 100 g. The surface was fine granular and reddish-purple. The cortical markings and corticomedullary borders were rather obscure. Each renal pelvis was highly dilated with intense mucosal hyperemia.

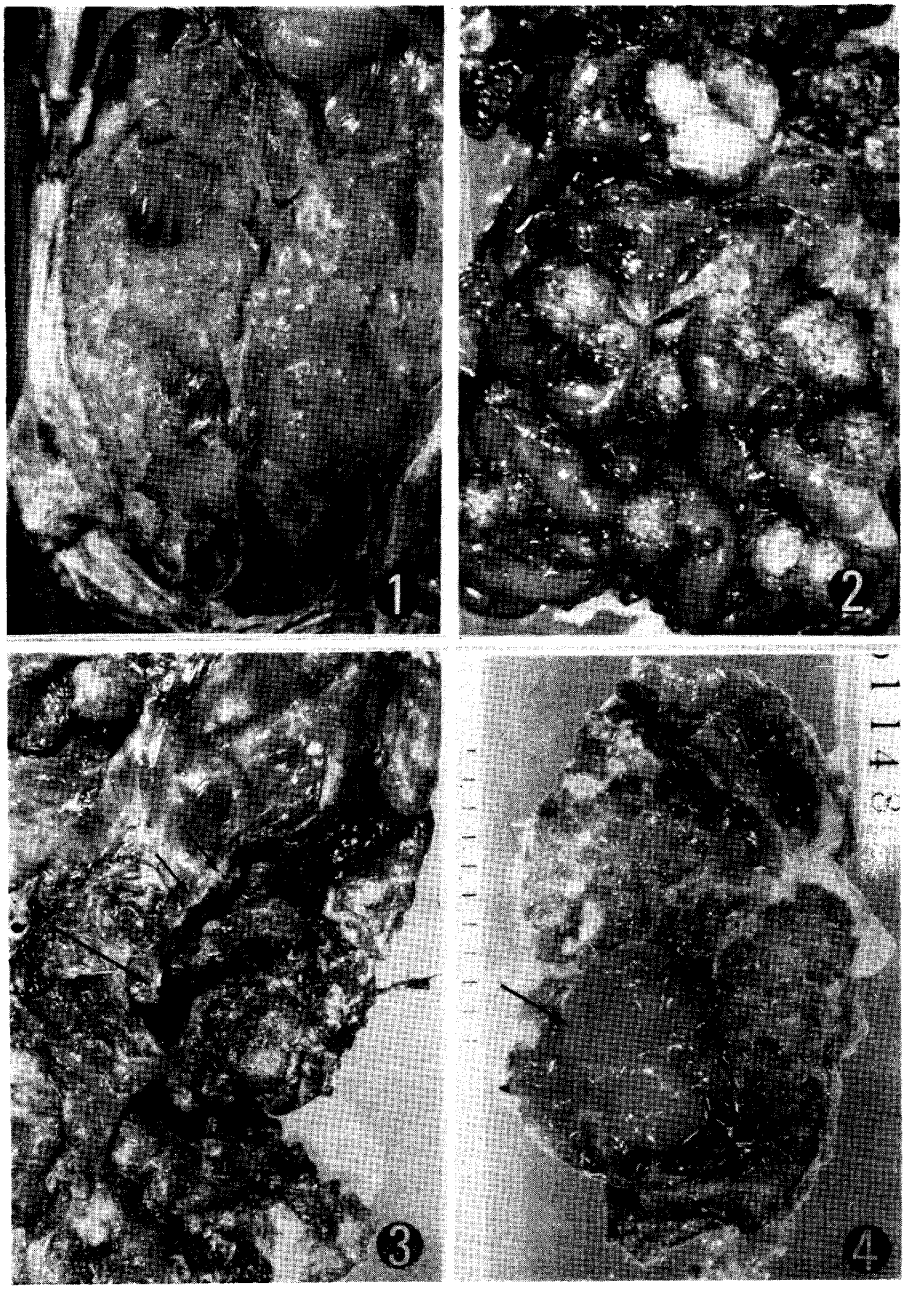
Fig. 1. In situ appearance of pseudomyxomatous tumor, occupying the entire peritoneal cavity.

Fig. 2. Pseudomyxomatous nodular growths, showing adherence to intraperitoneal organs.

Fig. 3. Sigmoid colon, showing a part with marked stenosis (short arrows) and a perforation (long arrow) with abscess formation.

Fig. 4. Left ovary (arrow), showing completely cystic changes.

Both ureters were also dilated with stenosis of the lower part due to compression by the tumor. The urinary bladder showed mucosal hyperemia. The



spleen weighed 100 g, showing lymph follicular atrophy and congestion. The lymph nodes were intact. The vertebral bones showed red marrows. The thyroid was rather small and contained a nodular adenoma measuring 1 cm in diameter in the right lobe. The left adrenal weighed 5 g and the right 3 g; each revealed lipid depletion of the cortex. The central nervous system was not examined.

Histological findings. The left ovarian lesion and intraperitoneally disseminated tumors were composed of various sized mucinous cysts, which were completely or incompletely lined with a single-layer of columnar cells (Fig. 5) partially showing papillary projections (Fig. 7). These neoplastic cells were rather differentiated with intensive mucin-production and scanty mitosis, but the nuclei were somewhat hyperchromatic (Fig. 6).

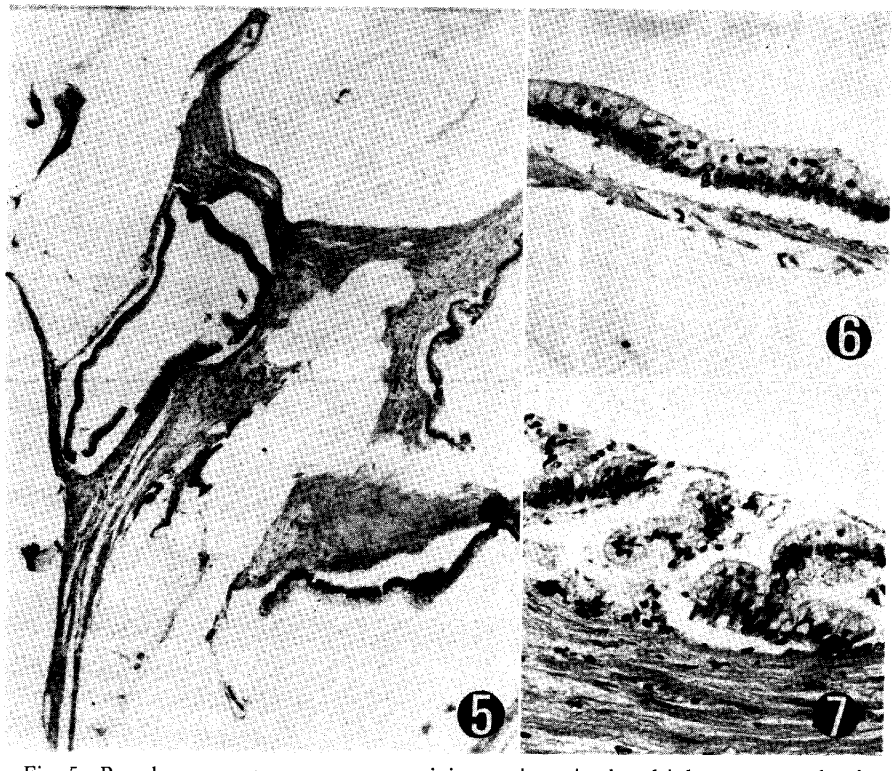


Fig. 5. Pseudomyxomatous tumor, containing various sized multiple cysts completely or incompletely lined by a single layer of columnar epithelium with mucinous accumulation. H-E. ×40.

Fig. 6. Mucin-secreting tumor cells, showing differentiated appearance but some hyperchromatism of the nucleus. H-E. ×400.

Fig. 7. Mucin-secreting tumor cells, showing a papillary pattern. H-E. ×400.

Findings were as follows: 1) diffuse extensive pseudomyxoma peritonei originating from the left ovary; 2) a perforation of a stenotic part of the sigmoid colon accompanied by abscess formation; 3) acute bronchopneumonia with atelectasis in both lower lobes; 4) hydronephrosis with slight acute pyelonephritis in both kidneys; 5) central fatty change and congestion of the liver; and 6) cachexia.

DISCUSSION

Since Werth (7) in 1884 first applied the term of pseudomyxoma peritonei to the secondary mucinous deposits arising in a rupture of ovarian cyst, and Fraenkel (8) in 1901 described the first case originating from a ruptured mucocele of the appendix in a male, many cases have been noted. In Japan, Amano (9) in 1899 first reported two cases of ovarian origin, and Abe (10) in 1910 first described a case of appendiceal origin. According to a recent review by Sotoyama *et al.* (6), 381 cases had been noted in Japan by 1975.

Although cases of pseudomyxoma peritonei of unusual origin including the uterine corpus, bowel, urachal cyst, umbilicus and common bile duct have been reported (11), most cases of the disease have been secondary to either ovarian or appendiceal lesions (1, 2, 6, 12). The appendix is the most common site of origin in men. In women, the ovary is most affected, and the bilateral nature of the ovarian origin (13) and concomitant ovarian and appendiceal involvement cannot be ruled out (3, 14). The precise site of origin is often difficult to decide in aggressive casses of the disease.

The clinical characteristics of the disease are summarized as follows (3-6, 12-18). Most patients are over middle-age, most commonly in the fifth and sixth decades. Females are more likely to be affected than male. The initial complaints are vague gastrointestinal symptoms of long duration such as anorexia, weight loss and a sense of a full abdomen. Marked abdominal distension with palpable nodules or massive ascites appears rather late. Laboratory findings are of little significance for the diagnosis, though anemia, high erythrocyte sedimentation rate and hypoproteinemia are often present. Aggressive surgical therapy results in a fairly improved prognosis. Intraperitoneal administration of anticancer agents may be of value, whereas radiotherapy and systemic chemotherapy are not usually effective. Some patients in the late stage have complications such as intestinal obstruction and peritonitis. Visceral invasion and distant metastasis are rare (19-22). The prognosis of the patient is relatively good. Long *et al.* (5). reported that the rate of five year survival was 45% and that of ten year survival 40%. Our case was a typical instance showing initial gastrointestinal symptoms, intestinal obstruction in terminal stage and extensive dissemination of the pseudomyxomatous tumor throughout the peritoneal cavity without distant

metatasis.

There are several theories on pathogenesis of pseudomyxoma peritonei including those based on inflammatory (8, 18), metaplastic, benign neoplastic and malignant neoplastic origins, of which the last seems to be generally accepted (2, 3, 18). Some authors have supported the theory of benign neoplasm because of long survival, no visceral invasion, no metastasis and differentiated histological features (3, 16, 23). A number of authors have stressed, however, that the ability of extensive spread of implanted cells from the original tumor must be evidence of malignancy and such phenomena questionably occur in benign neoplasm (2, 18). Although rare cases show true invasion, metastasis and a high degree of histological malignancy (9, 24), typical pseudomyxoma peritonei is a low grade of malignancy (3–5) and does not usually arise in any poorly differentiated tumor unable to produce massive mucinous ascites (3).

Our case was undoubtedly malignant because of the extensive intraabdominal dissemination and short course. Histologically, moreover, both primary and disseminated tumors showed moderate nuclear hyperchromatism of the tumor cells and a papillary pattern in some areas. Such histological findings, therefore, may be regarded as significant evidence of low grade malignancy in spite of the differentiated histology.

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