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A case of primary retroperitoneal mucinous cystadenocarcinoma.

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Abstract

A rare case of primary retroperitoneal mucinous cystadenocarcinoma in a 44-year-old woman is reported. The cystic tumor was delineated by CT and echography. The tumor was removed intact in the presence of bilateral normal ovaries and demonstrated an infiltrating malignant process. This neoplasm may have arisen from a supernumerary ovary. The patient died of recurrence 4 months after surgery. A comparison of the known cases indicates that aggressive treatment by hysterectomy with bilateral salpingo-oophorectomy in addition to cyst extirpation may improve prognosis.

KEYWORDS: retroperitoneal cystadenocarcinoma

A Case of Primary Retroperitoneal Mucinous Cystadenocarcinoma

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A rare case of primary retroperitoneal mucinous cystadenocarcinoma in a 44-year-old woman is reported. The cystic tumor was delineated by CT and echography. The tumor was removed intact in the presence of bilateral normal ovaries and demonstrated an infiltrating malignant process. This neoplasm may have arisen from a supernumerary ovary. The patient died of recurrence 4 months after surgery. A comparison of the known cases indicates that aggressive treatment by hysterectomy with bilateral salpingoophorectomy in addition to cyst extirpation may improve prognosis.

Key words : retroperitoneal cystadenocarcinoma

Primary retroperitoneal mucinous cystadenocarcinoma is exceedingly rare. This is the fifth case in the English literature to date.

A 44-year-old woman was admitted to the Iwakuni National Hospital with an enlarged asymptomatic abdominal mass in March 1988. She expressed no abdominal complaints, change in bowel habits, or change in menstrual cycles.

Physical examination was normal except for a palpable mass extending 5 finger-breadth in the right lower quadrant. Pelvic examination by a gynecologist showed normal female genitalia. An ultrasound study delineated a large cystic mass in the right lower abdomen with an echogenic solid nodule (Fig. 1-a). CT scan also demonstrated a large cystic mass (Fig. 1-b). Laboratory evaluation, including carcinoembryonic antigen and alpha-fetoprotein level, was within normal limits.

At laparotomy, in April 1988, a large cystic mass arising from the retroperitoneum at the right

iliac fossa was found. There was no pedicle, and the cyst was removed in toto by blunt dissection. The uterus, both ovaries and ureters were normal in appearance. Origination from the kidney, adrenal, or appendix was also excluded. There were no peritoneal implants or no ascitic fluid. The cyst, removed intact, measured 5.0 × 10.0 × 12.5 cm. On gross examination, the external surface of the cyst was smooth and glistening. The cyst exhibited a solid mass projecting into the lumen and contained dark gray-colored fluid (Fig. 2).

Microscopic examination revealed a mucinous cystadenocarcinoma with fibrous wall. This tumor showed a varying degree of differentiation. One area was lined with a single layer of benign appearing mucinous columnar cells (Fig. 3-a). In contrast, the papillary nodule protruding into the cystic lumen was composed of atypical adenocarcinoma cells (Fig. 3-b), and some areas demonstrated infiltration into the connective tissue wall by carcinoma cell nests surrounded with a sar-

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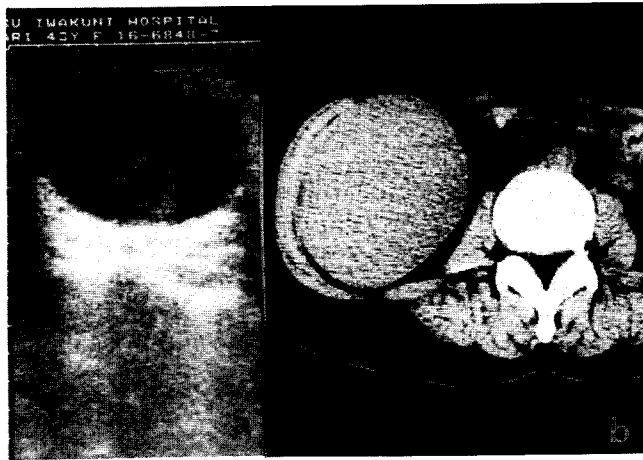


Fig. 1 Imaging techniques for the diagnosis of the cystic tumor. (a) ultrasonography, (b) CT scanning.



Fig. 2 The gross appearance of tumor. The inner surface exhibited a solid mass projecting into the lumen.

comatous element (Fig. 4). These histologic features raised the possibility of malignant transformation of a benign mucinous cystadenoma. Ovarian tissue was not identified in the wall.

The patient's post-operative course was unremarkable and she was discharged from the hospital. She was treated with an anticancer

agent; Tegafur-Uracil 300mg daily after operation. But, in July 1988, she was re-admitted complaining chiefly abdominal distension and weakness. CT scan demonstrated ascites with multiple metastatic nodules. Laboratory data at this admission were within normal limits except for white blood cell counts of $11,300/\mu\text{l}$ and a

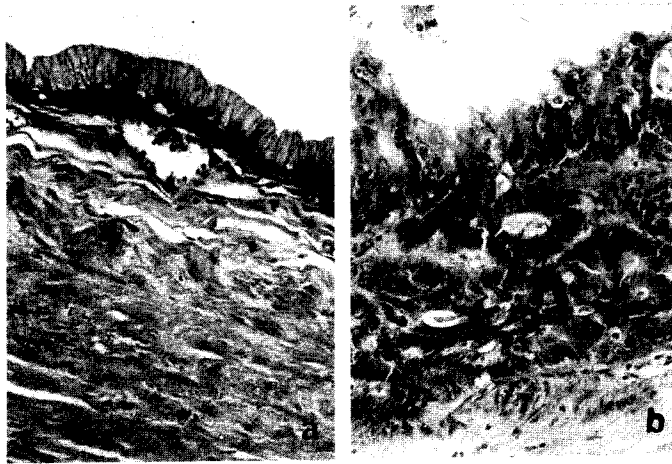


Fig. 3 Photomicrographs of (a) cyst wall showing the appearance of mucinous cystadenoma, and (b) nodule showing mucinous cystadenocarcinoma.

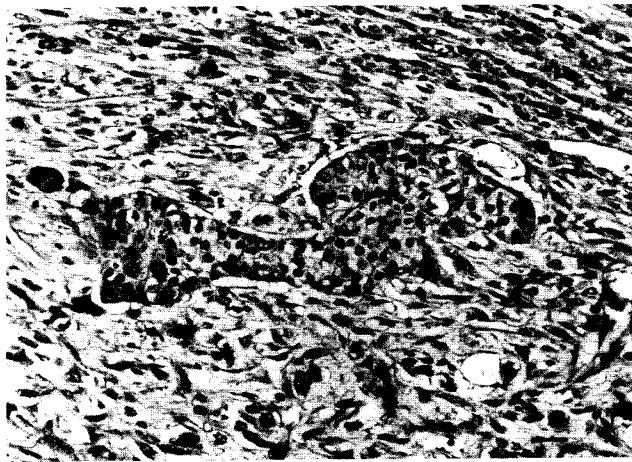


Fig. 4 An area of stromal invasion by nests of carcinoma cells surrounded by a sarcomatous element.

cancer antigen (CA) 125 of 592 U/ml. The cytological test of ascitic fluid proved to be adenocarcinoma. Her condition rapidly deteriorated despite the treatment with anticancer drugs (Cisplatin, Mitomycin C) and she died four months after the removal of the primary tumor. Necropsy was performed and the pathologic

examination proved the multiple nodules to be a recurrence of mucinous cystadenocarcinoma.

Primary retroperitoneal mucinous cystadenocarcinoma is very rare. The English literature to date contains only 4 other cases of this neoplasm (1-4). The patients, including this case, were all female. At the time of surgery

both ovaries appeared normal in all patients. It is of interest that the disease recurred in all 3 patients who underwent only cyst extirpation. Two of the 3 patients died of widespread metastasis 4 and 6 months after surgery, respectively. In contrast are the other 2 patients, on whom hysterectomy and bilateral salpingoophorectomy were performed in addition to tumor extirpation, although the ovaries were seemingly normal. Only these 2 patients lacked evidence of recurrence 22 and 36 months after operation, respectively. Despite its rarity, a supernumerary ovary has been considered the basis for the development of benign or malignant retroperitoneal cystadenomas that are documented in the presence of two normal ovaries (4). According to Lachman (5), there have been 16 cases of supernumerary ovary reported from 1959 until now. And two of them had an associated tumor of a mucinous cystadenoma. Retroperitoneal cystadenocarcinoma is thus thought to arise from supernumerary ovary and to be under the influence of female hormones. Under the same hormonal circumstance, the original ovaries may have neoplastic potential like a supernumerary ovary. Alternatively the supply of female hormone from the

original ovaries may promote the recurrence of the tumor. Considering the fact that the patients whose operation was confined to tumor extirpation often died of recurrence, the additional and aggressive surgery of hysterectomy and bilateral salpingoophorectomy may be necessary to improve the outcome.

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