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Peritoneal papillary serous carcinoma is a rare tumor that involves the surface of the pelvic and/or abdominal peritoneum. Long-term survival among patients with this tumor has been rare. Most patients with this cancer have been treated with debulking surgery and postoperative chemotherapy. A case of incompletely resected peritoneal papillary serous carcinoma with a complete response to cisplatin-based chemotherapy is reported. Subsequent laparotomy revealed no residual tumor. This case suggests that primary chemotherapy may be successful in treating unresectable primary papillary serous tumors of the peritoneum. (Henry Ford Hosp Med J 1992;40:136-8)

Papillary serous carcinoma (PSC) of the peritoneum was first described by Swerdlow (1) in 1959. It involves diffusely the peritoneum, histologically resembles PSC of the ovary, but either spares the surface of the ovary or involves it only microscopically (2). About 130 cases have been described under a variety of names including mesotheliomas, mesodermoma, peritoneal PSC, papillary carcinoma from extraovarian tissue, serous surface papillary carcinoma of the ovary, and peritoneal carcinomatosis of unknown primary site. There is still no consensus regarding nomenclature, histogenesis, immunohistochemical characteristics, clinical behavior, and treatment.

Case Report

A 75-year-old white female, gravida 3, para 3, underwent surgery in a local hospital for possible incarcerated inguinal hernia. There was a four-month history of gradually increasing abdominal girth, and on admission her abdomen was markedly distended. She denied any change in weight. Physical examination confirmed the presence of abdominal distension and tympany and hyperactive bowel sounds and disclosed a 3 cm ovoid nontender, nonreducible mass in the right inguinal area. On admission, x-rays and laboratory tests were reportedly normal. With the patient under spinal anesthesia, a right incarcerated femoral hernia sac was opened. During exploration, multiple polypoid implants suggestive of carcinomatosis along with a large amount of brown-colored peritoneal fluid and necrotic tumor material were encountered. There was no evidence of bowel in the hernia. The incision was extended and abdominal exploration revealed marked ascites, diffuse tumor infiltration of the entire pelvis and small bowel, and "caking" of the lower third of the omentum. The liver and upper abdominal structures were not involved, and there was no evidence of bowel obstruction. After multiple biopsies were taken from the omentum, hernial sac, and pelvic peritoneum (Figure), the incision was closed as the tumor was considered to be unresectable. The postoperative course was uneventful except for mild ileus. On the recommendation of the medical oncologist, the patient received nine courses of cytoxan and carboplatin over a total of eight months following the usual protocol for ovarian carcinoma. Follow-up visits revealed that the patient was doing well with no clinical evidence of disease. The carcinoembryonic antigen (CEA) and CA 125 levels were consistently normal. After completion of the chemotherapy, abdominal computed tomography (CT) revealed a 1 cm soft tissue density at the level of the cecum, which was unchanged from a previous CT one month earlier.

The patient was referred to Henry Ford Hospital for a second exploratory laparotomy 11 months after the original surgery. She underwent lysis of adhesions, cytology washings, total abdominal hysterectomy and bilateral salpingo-oophorectomy, selective pelvic and paraaortic lymph node dissection, omentectomy, and repair of right inguinal hernia. There were dense adhesions around the bowel and adnexae. Both ovaries were atrophic, about 1 cm in greatest diameter. A 2 cm nodule was found in the right pelvic side wall, posterior to the cecum, with multiple small yellowish nodules on the pelvic floor and peritoneal surface. Overall there was no gross evidence of disease. The pelvic washings and multiple biopsies were negative. The patient was alive and free of disease 11 months after her second exploration.

Discussion

Many previous studies indicate that patients with primary PSC have a worse prognosis than do those with ovarian cancer. It is unclear whether this worse prognosis is due to the advanced disease which is frequently found in this tumor or to unknown inherent biological characteristics. We present a case of primary PSC and extensive peritoneal involvement with absence of disease surgically documented after chemotherapy.

Malignant tumors involving the peritoneum are usually due to metastatic ovarian carcinoma. In rare cases, no primary tumor

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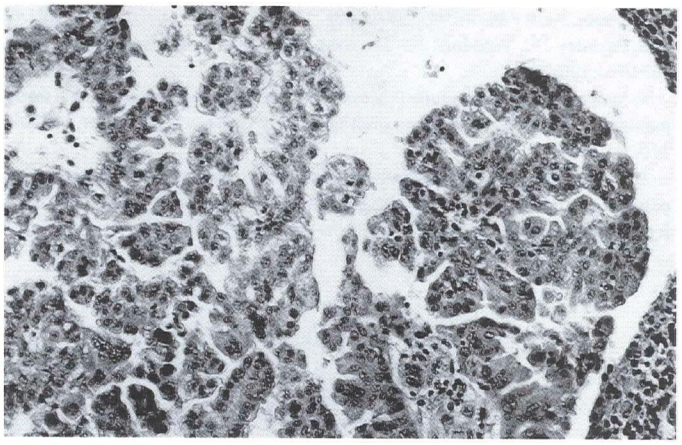
can be found and a primary tumor of the peritoneum must be considered. Of course other primary tumors must be ruled out. As in ovarian serous tumors, benign, borderline, and malignant variants of papillary serous tumors have been reported (3). Benign or borderline peritoneal tumors must be differentiated morphologically from mesotheliomas (4). PSC is frequently described microscopically as showing mitotic figures and many psammoma bodies (5,6). Most PSC tumors have at least a few cells that show cytoplasmic or surface mucin and stain positively for keratin and epithelial membrane antigen. Other markers such as Leu-M1, B72.3, vimentin, HPLAP, and CEA are found in a small percentage of cells (6,7). In fact, on the basis of light microscopy and histochemical and immunohistochemical characteristics, PSC cannot be differentiated from ovarian carcinoma. This lack of cytochemical discrimination is explained by the common histogenesis of these neoplasms. Both the peritoneal mesothelium and the surface epithelium of the ovary develop from the embryonic coelomic epithelium. The commonly accepted theory for the origin of peritoneal papillary carcinoma is that it arises directly from the mesodermal lining of the abdominal cavity in response to an unknown oncogenic stimulus (4,8). On the other hand, some believe that PSC arises from embryonic rest cells (5). In any event, the coelomic origin of these tumors explains their resemblance in many respects to ovarian cancer. The fact that PSC has occurred in patients after bilateral salpingo-oophorectomy supports the concept of its peritoneal origin (9).

PSC affects only women, whose median age is about 60 years (4,10). Abdominal pain and distension are the most common presenting symptoms (6,9) while a palpable mass is uncommon (4). Radiological studies including CT are usually not helpful in diagnosis (6). In most cases the entire mesothelial surface is involved and the tumor is rarely predominant in or limited to the pelvis. Generally the ovaries are involved only superficially and are of normal size. When the tumor invades the ovaries, one encounters superficial involvement as well (6). PSC frequently involves the submesothelium, but deep invasion into the abdominal or pelvic organs or either local or distant metastases are rare (5). Spread of the disease outside the abdominal cavity producing malignant pleural effusion has been described (9). Ascitic fluid, when present, usually contains malignant cells.

The median survival time of patients with PSC is between 11 and 24 months, similar to those of the same age group who have ovarian cancer (2,9). Some have found that peritoneal tumor has a worse prognosis than ovarian cancer (11,12). While most series report only a few long-term survivors, Ransom et al (13) described three patients who lived over six years. Chen and Flam (3) also reported long-term survival after cisplatin-based chemotherapy. These patients had undergone optimal debulking surgery followed by cytoxan-cisplatin chemotherapy.

There is no consensus as to the best therapy for peritoneal carcinoma, but most authors prefer to treat it in the same manner as ovarian cancer (2,4,9). Complete clinical response to postoperative chemotherapy has been reported in 20% to 25% of patients (2,4,6), but not all respond so favorably (13). Patients treated with combination chemotherapy do better than those treated with single agents and cisplatin-based regimens seem to be more effective (4). Contrary to other authors, Fromm et al (4) in their large series of 74 patients found that neither chronologic age nor the presence of residual disease after cytoreduction determined prognosis. This finding is in contrast with that found in ovarian cancer. Fromm et al concluded that the absence of mitoses represented a favorable prognostic factor. Lele et al (2) described one patient with complete response to cisplatin, bleomycin, and cytosine arabinoside administered intraperitoneally. Most studies concerning PSC of the peritoneum are retrospective and there is no staging system. Few reports allow conclusions about therapy, most focus on the pathology, and all are retrospective in nature. Only prospective studies conducted in multiinstitutional clinical trials can provide reliable data about the natural history of this disease. The value of intraperitoneal chemotherapy must be carefully investigated. Our case suggests that chemotherapy may be successful in treating unresectable primary PSC of the peritoneum.



Figure—Papillary serous carcinoma of the peritoneum (hematoxylin-eosin stain, X50).

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