Primary Papillary Serous Carcinoma of the Peritoneum, A Retrospective Analysis of 18 Cases

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OBJECTIVE: Primary papillary serous carcinoma of the peritoneum (PPSCP) is an uncommon tumor. The aim of this study was to review of the patients with PPSCP.
STUDY DESIGN: 18 women with PPSCP in gynecologic oncology clinic between 1995 and 2005 were retrospectively reviewed.
RESULTS: At diagnose, one patient (5.5 %) had stage IV and other patients (94.5 %) were IIIC. Six patients received neoadjuvant chemotherapy (paclitaxel/platinum in 5 patients and cyclophosphamide/platinum in the other). Initial or interval optimal/suboptimal debulking surgery was performed in all patients. After surgery, all of the patients received adjuvant chemotherapy including paclitaxel/platinum (n=16) and cyclophosphamide/platinum (n=2). In 6 months after adjuvant chemotherapy, persistent, progressive or recurrence disease was detected in 8 patients (44.4%). In 9 patients (50%) recurrence was detected after 6 months. Clinical complete response to chemotherapy was 55.5%. Survival rate was 90%.
CONCLUSION: PPSPC are frequently indistinguishable from advanced serous papillary carcinoma of the ovary. The treatment choice is optimal cytoreductive surgery with adjuvant chemotherapy, but although the radical treatment is used the survival rate is poor.
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Key Words: Primary Papillary Serous Carcinoma, Peritoneum

Primary papillary serous carcinoma of the peritoneum (PPSCP) is an uncommon tumor. The incidence of the PPSCP has been reported to be 6.5-9% of all cases with clinically suspected serous papillary carcinoma of the ovary.[1] The clinical presentation and histological appearances are frequently indistinguishable from advanced serous papillary carcinoma of the ovary.[1-3] Diagnosis and treatment of this malignancy are also similar to those of the serous papillary ovarian carcinoma.[3,4]

The aim of this study was to review the clinical features, the diagnosis, and the treatment of the patients with primary papillary serous carcinoma of the peritoneum.

Material and Methods

The clinical characteristics and treatment outcome of 18 women with PPSCP in gynecologic oncology clinic between 1995 and 2005 were retrospectively reviewed. The criteria for diagnosis of PPSCP are due to the Gynecologic Oncology Group (GOG) as the following:

1. Both ovaries are either normal in size or enlarged by a benign process and their largest diameter does not exceed 5 cm;

2. The involvement in the extra ovarian sites must be greater than the involvement on the surface of either ovary;

3. Microscopic examination of the ovarian component must be one of the following;

a) No tumor.

b) Confined to ovarian surface epithelium with no evidence of cortical invasion or superficially invading the underlying cortical stroma with any given tumor size less than 5 mm in any area.

4. Histological and cytological characteristics of the tumor and peritoneal disease are similar to that of ovarian serous papillary adenocarcinoma of any grade.

The stage at diagnose was defined as IIIC in patients whom had peritoneal disease greater than 2 cm and as IV whom had peritoneal disease with pleural effusion, according to the 1987 FIGO staging system.

Results

The median age was 61.8 years (range 52-75). At diagnosis, the most common symptom of the patients was abdominal distention (66.7%). The mean value of CA 125 at diagnosis was 1026.7 IU/ml with a range of 49-6000 IU/ml. One patient (5.5%) had pleural effusion positive for malignant cells diagnosed by thoracentesis and was stage IV and other patients (94.5%) were classified as stage IIIC. Diagnostic procedures included ultrasonography, Computed Tomography (CT) in all patients, paracentesis in 4 patients, co-

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lonoscopy and eusophagogastroduodenoscopy in 12 patients, 6 for each procedure. In our patients, the greatest ascites volume at surgery was 14000cc (Table I). The most common sites of disease at laparotomy were peritoneal surfaces, omentum and superficial involvement of the ovaries.

Table 1. Clinical characteristics, diagnostic procedures, volume of ascites and operative complications of PPSCP patients

	No.	%
Main presenting symptom		
Abdominal distention	12	66.7
Gastrointestinal symptoms	6	33.3
Diagnostic procedures		
Pelvic ultrasonography	18	100
CT	18	100
Colonoscopy	6	33.3
Eusophagogastroduodenoscopy	6	33.3
Paracentesis	4	22.2
Volume of ascites		
≤1000 cc	5	38.5
>1000 cc	13	61.5
Operative Complications		
Infection	3	16.6
Lymphocyst	3	16.6
Wound dehiscence	1	0.6

For treatment modalities; in patients determined as unresectable, optimal debulking could not be performed, neoadjuvant chemotherapy was given initially. Six patients received neoadjuvant chemotherapy with three courses (paclitaxel/platinum in 5 patients and cyclophosphamide/platinum in the other). Initial or interval optimal/suboptimal debulking surgery was performed in all patients. Optimal debulking surgery defined as a residual tumor diameter below 0.5 cm. One patient's initial surgery was assessed as suboptimal debulking surgery. A large proportion of patients underwent an extensive surgical procedure including total abdominal hysterectomy (TAH) + bilateral salpingo-oophorectomy (BSO) (n=18), omentectomy (n=18), appendectomy (n=16), paraaortic-bilateral pelvic lymphadenectomy (PAB-PLND) (n=16). In one patient, low rectal anostomose was performed as a result of segmental resection of the adherent bowel infiltrated with tumor.

After surgery in 6 weeks, all of the patients received adjuvant chemotherapy including paclitaxel/platinum (n=16) and cyclophosphamide/platinum (n=2). In 6 months after adjuvant chemotherapy, persistent or progressive disease was detected in 8 patients (44.4%). In 9 patients (50%) recurrence was detected after 6 months. In 17 of 18 patients (94.4%), recurrence was seen within 2 years. Clinical complete response to chemotherapy was 55.5% (n=10). Fifteen patients (83.3%) had taken second line, 9 patients (50%) had taken third line chemotherapy. Second look laparotomy (SLL) was performed in 3 patients (16.7%). SLL was positive in 2 patients and negative for the other one. In our cases, the median follow up time is 35.5 months (12-41). Eight patients had lost to follow-up. One of the remaining 10 patients died.

Discussion

Primary papillary serous carcinoma of the peritoneum (PPSCP) is an uncommon tumor; it was first described by Swerdlow in 1959.[1] In 1977, Kannerstein et al. reported 15 cases and attracted attention for separation of serous papillary carcinoma from malignant mesothelioma with epidemiological, histopathological, immunohistochemical differences and suggested different types of therapy.[5] These tumors' clinicopathologic features resemble that of serous ovarian tumors. As in serous ovarian tumors, there are variants such as primary peritoneal serous borderline tumors, low grade peritoneal serous carcinomas and malignant form that is referred to as serous surface papillary carcinoma or as extra-ovarian peritoneal serous papillary carcinoma. In borderline forms, many cases are discovered incidentally at operation for other reasons. Low grade peritoneal serous carcinomas have invasion pattern with multifocal or diffuse widespread peritoneal involvement. The average age increases to 57 years so was 35 in borderline forms.[6] In a study by Look et al, the mean age of the patients with PPSCP was 54 years. But, Ransom et al, found the median age older similar to our group (61 and 61.8 respectively).

Patients with PPSCP usually present with nonspecific complaints of abdominal pain, distention and anorexia.[1] as did most of our patients. These patients with nonspecific complaints needed advanced investigation to prove the diagnosis of PPSCP. The most common method used was CT survey. Chiou et al. concluded that the presence of diffuse peritoneal disease is strongly suggestive for PPSCP.[1] In our series we also used CT to diagnose the disease. Ascites, peritoneal or omental thickening, nodularity or masses, associating with normal size ovaries or the absence of an ovarian mass on CT are used as diagnosis criteria and at least one of these was seen in all of our patients. Elevation of serum CA-125 level may also help in the diagnosis of PPSCP. In all studies, it was shown that CA-125 levels are higher than the base line level of 35 IU/ml, as in our series.[2,3,7]

As in other studies, all our PPSCP patients had stage III or IV disease at diagnosis.[2,3] Because of the advanced stage, the rate of the optimal cytoreduction in PPSCP has been variously reported 33% to 70%[3] and the rate in our study (61.1%) is within this range but when we included the 6 patients who received neoadjuvant chemotherapy, this percentage will be higher. Optimal cytoreductive surgery for PPSCP can be effective when adjuvant chemotherapy was added. Chen and Flam reported that with combination chemotherapy consistent of Adriamisin and Cisplatin, the patients had no evidence of disease 5 or more years after diag-

nose.[8] Similar to this study Ransom et al. reported that by debulking surgery with platin based chemotherapy, patients have had long term survival.[9] We also used platin based adjuvant chemotherapy as a part of treatment in all patients. Instead of the radical surgery and adjuvant chemotherapy, survival rate of the disease is poor as 10 months.[10] Fromm et al, reported 74 patients with serous carcinoma of the peritoneum. In their group, the average age was 57.4 years, 89.1% patients' disease involved the omentum, and the median survival for the total group was 24 months. They reported that neither patient age nor presence of residual disease after cytoreduction predicted survival but only the absence of mitoses improved survival. Combination chemotherapy was better than that of the treatment with single agent regiments.[11] But one point that should be strongly determined was that in 17 of 18 patients, recurrence was seen within 2 years (94.4%). In our study, survival rate was 90% in 35.5 months which is median follow-up time.

In conclusion, PPSCP is a rare disease with a frequency of 6.5-9%.[1] The clinical presentation and histological appearances are frequently indistinguishable from advanced serous papillary carcinoma of the ovary. The possibility of the disease should be considered in elderly or middle aged patients with peritoneal carcinomatosis in the absence of an enlarged ovary. The optimal treatment choice is cytoreductive surgery with adjuvant chemotherapy, but although the radical treatment is used, the survival rate is poor.

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