Primary Carcinoid Tumor of the Uterine Corpus  
- A Case Report -

Sung Ran Hong · Hy Sook Kim  
Jae Uk Shim¹

Department of Pathology, ¹Department of Obstetrics and Gynecology, Samsung Cheil Hospital, Sungkyunkwan University School of Medicine, Seoul, Korea

Corresponding Author
Sung Ran Hong, M.D.
Department of Pathology, Samsung Cheil Hospital & Women’s Healthcare Center, Sungkyunkwan University School of Medicine, 1-19 Moakjung-dong, Chung-gu, Seoul 100-380, Korea
Tel: 02-2000-7661
Fax: 02-2000-7779
E-mail: sungran@samsung.co.kr

Carcinoid tumors are neoplasms of a neuroendocrine origin that rarely affect the genital tract. Most carcinoid tumors in the female genital tract arise in the ovary or in the cervix.¹-⁴ The uterine cervix is the most frequent site of genital neuroendocrine tumors, some of which are small cell carcinomas having an aggressive behavior.⁵ Only a few carcinoid tumors of the uterine corpus have been described in the English medical literature.⁶-⁸ We have recently experienced a case of typical carcinoid tumor of the uterine corpus, and this is considered to be the first case reported in Korea.

CASE REPORT

A 61-year-old woman was admitted in December 1992, with an impression of uterine myoma. She complained of vaginal spotting for 10 days and she also had a past history of chronic gastritis. Her physical examination revealed a 1.5 cm sized pedunculated mass in the uterine cervix. The papanicolaou smear was normal. Her routine laboratory and radiologic examinations preparing her for an operation were within normal ranges. The patient received a total hysterectomy and bilateral salpingo-oophorectomy, and this was done under the clinical impression of cervical myoma.

On gross examination, the endometrial cavity showed a relatively well demarcated, elevating mass involving nearly the full thickness of the underlying myometrium (Fig. 1). The mass was solid, about 5 × 5 × 4 cm in dimension, with a superficial friable consistency and a deep calcified area. The lower uterine segment revealed a pedunculated solid mass, 3 cm in diameter, which obstructed the cervical os. Microscopically, the endometrial mass involving the underlying myometrium was that of a typical carcinoid tumor. Upon pathological analysis, the mass revealed a characteristic organoid arrangement with trabecular, follicular, rosette, and solid patterns in a relatively scanty fibrous stroma (Fig. 2A). Individual tumor cells were round and uniform, and they demonstrated mild pleomorphism, finely stippled to slightly coarse chromatin pattern, inconspicuous nucleoli and abundant granular pinkish cytoplasm with rare mitotic activities (Fig. 2B). These cells showed positive reactions for neuron-specific enolase (NSE) and chromogranin (Fig. 2C). The pedunculated mass of
the lower uterine segment was of a leiomyoma classification.

The clinical evaluation was done and we tried to look for other primary sites of the endometrial carcinoid tumor. Her facial flushing for 2 years was recognized. However, the postoperative 24-h urinary 5-hydroxyindolacetic acid (5-HIAA) level was normal. Gastric endoscopy showed a small 7 mm sized mucosal elevation at the posterior wall of the mid-antrum. The gastric lesion was a well differentiated adenocarcinoma (Fig. 3) and the immunohistochemistry for NSE and chromogranin was negative. Other studies including chest, abdominal and pelvic CT scans were all normal. No further treatment was done.

The patient was lost to follow-up until June 1999, when she complained of a vaginal nodular mass. The vaginal mass, approximately 4 × 4 × 3 cm, involved the right pelvic cavity. The pathologic findings of the vaginal mass were similar to those of the previous endometrial tumor. However, the tumor's cellular pleo-

Fig. 1. A relatively well demarcated solid mass, 5 cm in diameter, involves the full thickness of the uterine wall.

Fig. 2. The uterine corpus tumor shows trabecular growing pattern (A), round and uniform tumor cells characterized by the stippled chromatin pattern (B), and positive reaction for chromogranin (C).
morphism and mitotic activity were slightly more severe in degree than the previous ones, and this mass resembled an atypical carcinoid tumor (Fig. 4). At that time, she did not have any gastric symptoms. Radiation therapy was recommended, but the patient refused. Approximately 2 years later, the patient was admitted to another hospital due to right lower abdominal pain. Sonogram and CT scan showed a huge vaginal mass, approximately 9.8 × 5.8 cm, that was associated with a right pelvic mass approximately 5 × 8 cm in size. Ascites, markedly dilated small bowel, collapsed large bowel, and multiple metastatic lesions in right upper lung and lymph nodes of right lower mesentery and right iliac arterial area where all noted. The patient died of the mechanical intestinal obstruction by the carcinoid tumor with distant metastasis.

DISCUSSION

Because primary typical carcinoid tumors occurring in the endometrium are exceptionally rare, the central, critical question is whether this lesion is a primary or metastatic carcinoid tumor. We considered this case to be a primary uterine corpus tumor rather than a metastatic carcinoid tumor, although a small gastric lesion was presented. The gastric lesion was pathologically different from the endometrial carcinoid tumor: the gastric lesion was a well-differentiated adenocarcinoma lacking the typical features of carcinoid tumor and it was negative for NSE and chromogranin. The small gastric adenocarcinoma might have been an unusual variant of minute early gastric carcinoma that could have regressed after endoscopic biopsy. Her endometrial carcinoid tumor was positive for NSE and chromogranin, and her clinical findings of this case were also consistent with a primary endometrial carcinoid tumor. If this endometrial tumor was a metastatic one from the gastrointestinal lesion, it might be an unusual finding that the uterine body was the only metastasized focus without metastasis to any other organs at that time of total hysterectomy.

Fig. 3. Endoscopic biopsy of gastric antrum reveals a well differentiated adenocarcinoma.

Fig. 4. The recurred tumor shows trabecular or solid arrangement (A), and round and uniform tumor cells having slightly atypical nuclei and increased mitotic activity (B).
terectomy. Therefore, it was inferred that this case was a primary carcinoid tumor of the endometrium. The argyrophilic cells are reportedly present in normal endometrial gland, although the frequency and number of argyrophilic cells are higher in endometrial glandular adenocarcinoma than in normal endometrium. Since the initial description of endocrine tumors of the uterine cervix, subsequent publications have employed many diverse terms to describe the broad morphologic spectrum of these tumors; carcinoid tumor, argyrophil cell carcinoma (apudoma), small cell carcinoma, endocrine carcinoma of intermediate cell type, neuroendocrine carcinoma, and adenocarcinoma with carcinoid features. With these diverse terms, it has been difficult to compare their incidence, clinicopathologic features, biological behavior and natural history of these unusual tumors. For this reason, there was a consensus workshop to recommend the following four general categories for endocrine tumors of the uterine cervix: typical (classical) carcinoid tumor, atypical carcinoid tumor, large cell neuroendocrine carcinoma and small (oat) cell carcinoma. We also suggest the application this of this terminology should be used for the neuroendocrine tumor of the uterine cervix and for endometrial tumors. The present case belongs to the typical (classical) carcinoid tumor that is characterized by typical organoid patterns, round small uniform neoplastic cells, finely granular chromatin, inconspicuous nucleoli and rare mitotic figures.

The prognosis of carcinoid tumors is better than that of most carcinomas. A review of the literature concerning typical carcinoid tumor of the uterine cervix revealed that two patients (approximately 22%) died of disease within two years, and the remaining patients were alive and disease free at follow-up intervals of 2.5 to 24 months. The present patient showed a recurrence of the carcinoid tumor approximately eight and a half years after the initial detection of the tumor, and she died of mechanical intestinal obstruction by the carcinoid tumor.

REFERENCES