

# Primary Peritoneal Carcinoma Initially Presenting as Atypical Cervical Lymphadenopathy

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## Key Words

Cervical lymphadenopathy · Primary peritoneal cancer

## Abstract

Primary peritoneal carcinoma (PPC) is a rare cancer arising from the extraovarian peritoneum and is of müllerian origin. PPC and epithelial ovarian carcinoma share similar clinical, histopathological, and immunohistochemical features. Clinical symptoms and findings include abdominal distension and ascites. We experienced 2 cases of PPC which initially presented with cervical lymphadenopathy. Here, we report the 2 cases and review the literature.

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## Background

Primary peritoneal carcinoma (PPC) is a rare cancer arising from the extraovarian peritoneum and is of müllerian origin. PPC and epithelial ovarian carcinoma (EOC) have a number of clinical, histopathological, and immunohistochemical (IHC) features in common. However, PPC has minimal or no ovarian involvement, with no obvious precursor lesion [1]. The mainstream treatment for PPC is cytoreductive surgery followed by platinum-based chemotherapy. Treatment and response to chemotherapy are similar to those seen in EOC. PPC patients diagnosed with International Federation Gynecology and Obstetrics stage III–IV disease show a worse survival than patients diagnosed with an earlier-stage disease. In the past, PPC and EOC have been considered a single entity, but molecular and epidemiological findings have demonstrated that PPC is distinct from ovarian carcinoma [1–4]. According to

several reports, symptoms and clinical findings include abdominal distension, a palpable abdominal mass, and a large amount of ascites [1, 5–8]. To the best of our knowledge, there have been no reported clinical findings to date of PPC presenting with massive cervical lymphadenopathy. Here, we report the cases of 2 PPC patients who presented with cervical lymphadenopathy and review the current literature.

## Case Presentation

### Case 1

A 59-year-old woman was admitted to our clinic with a palpable mass on the left side of her neck. She had undergone a total hysterectomy with bilateral salpingo-oophorectomy 5 years earlier due to uterine leiomyoma. There was no other medical or gynecological history. CT scans of the neck, chest, abdomen, and pelvis identified a left pararenal mass and multiple sites of lymphadenopathy. Gastroscopy and colonoscopy examination results were normal. Laboratory findings showed a high level of serum CA-125 (1,710 U/ml). We performed a cervical lymph node biopsy. Histopathological findings revealed a metastatic serous adenocarcinoma of ovarian origin. IHC staining was positive for cytokeratin (CK) and negative for thyroid transcription factor 1 (TTF-1), vimentin, and CK20 (fig. 1).

### Case 2

A 76-year-old woman presented with persistent pain in her left flank. Multiple bilateral palpable neck masses were detected on physical examination. She had no relevant medical or gynecological history. CT scans of the neck, chest, abdomen, and pelvis showed a left pararenal mass and multiple sites of lymphadenopathy. Gastroscopy and colonoscopy examination results were normal. Laboratory findings revealed a high level of serum CA-125 (3,205 U/ml). We performed a cervical lymph node biopsy. Histopathological findings showed a metastatic serous adenocarcinoma of müllerian origin. IHC findings were positive for CK7 and epithelial membrane antigen and negative for TTF-1, vimentin, and CK20 (fig. 2).

In both cases, the following features were found: tumor involvement of extraovarian sites, ovaries of normal size, no microscopic ovarian components, and histological characteristics of serous adenocarcinoma. We diagnosed both cases as FIGO stage IV primary peritoneal carcinoma. Both patients were treated with combination chemotherapy of paclitaxel and carboplatin according to the treatment protocol used with EOC combination chemotherapy. Following the completion of 6 cycles of chemotherapy, both patients showed a partial response, and the serum CA-125 levels dropped to 28.34 U/ml in case 1 and to 164.6 U/ml in case 2. The patients survived for 13 (case 1) and 10 (case 2) months after the initial diagnosis.

## Discussion

Serous primary ovarian carcinoma (OC), serous primary fallopian tube carcinoma, and serous PPC were, until recently, considered a single entity – EOC. These diseases have similar clinical characteristics, but their diagnostic criteria are different. The diagnostic criteria for PPC according to the Gynecologic Oncology Group (GOG) are as follows: (1) both ovaries must be of normal size or enlarged by benign processes; (2) the involvement of the extraovarian sites must be greater than the involvement on the surface of either ovary; (3) microscopically, the ovarian component must (a) be nonexistent, (b) be confined to ovarian

surface epithelium with no evidence of cortical invasion, or (c) involve the ovarian surface epithelium and underlying cortical stroma with a given tumor size of less than 5 × 5 mm [1].

The 2 cases reported here presented initially with cervical lymphadenopathy and showed no ascites. The most common clinical symptoms and findings for PPC include abdominal distension, abdominal pain, a palpable abdominal mass and ascites. Although PPC and OC have similar clinical characteristics, 2 studies have reported that PPC patients present with abdominal discomfort more frequently than OC patients do [1]. According to GOG diagnostic criteria, our 2 cases are consistent with PPC, but the clinical characteristics of the 2 cases were not typical for PPC. CT scans of the cases showed pararenal masses and multiple sites of lymphadenopathy. Both cases showed only a partial response to chemotherapy. Overall, the clinical response to platinum-based chemotherapy in PPC is 63–88%, similar to that in OC [1]. The overall survival for PPC ranges from 7.8 to 25 months, also similar to that for OC [1]. The patients in our 2 cases have survived until 13 and 10 months after diagnosis.

PPC and OC have similar clinical and pathological findings. However, the origin of PPC and OC is different. PPC has a multifocal origin, but OC has a unifocal origin [1, 2]. Two studies have reported on protein expression or loss of heterozygosity (LOH) in different tumor sites from the same patient; they showed discordant expression of P53, HER2/NEU, BCL2, and NM23H1 proteins in 48% of patients or discordant LOH patterns on chromosome 6q in 97% of patients [1, 9, 10].

PPC and OC share a common embryological origin, and both diseases have many similar characteristics. However, they have molecular and epidemiological differences. In clinical practice, it is difficult to distinguish between primary unknown carcinoma and primary peritoneal carcinoma, particularly since PPC does not have any ovarian involvement, but its histology is that of serous adenocarcinoma. Many cancers share these features. Most PPC studies to date have been small, case-control, retrospective studies. Therefore, we have limited information regarding clinical findings, symptoms, and prognosis. Our cases did not display the typical clinical patterns of PPC, and it was difficult to diagnose PPC. Therefore, we reported them as PPC initially presenting with atypical cervical lymphadenopathy. Further international studies and analyses are required to collect more information on PPC.

### Statement of Ethics

Our institution approved the writing of this case report.

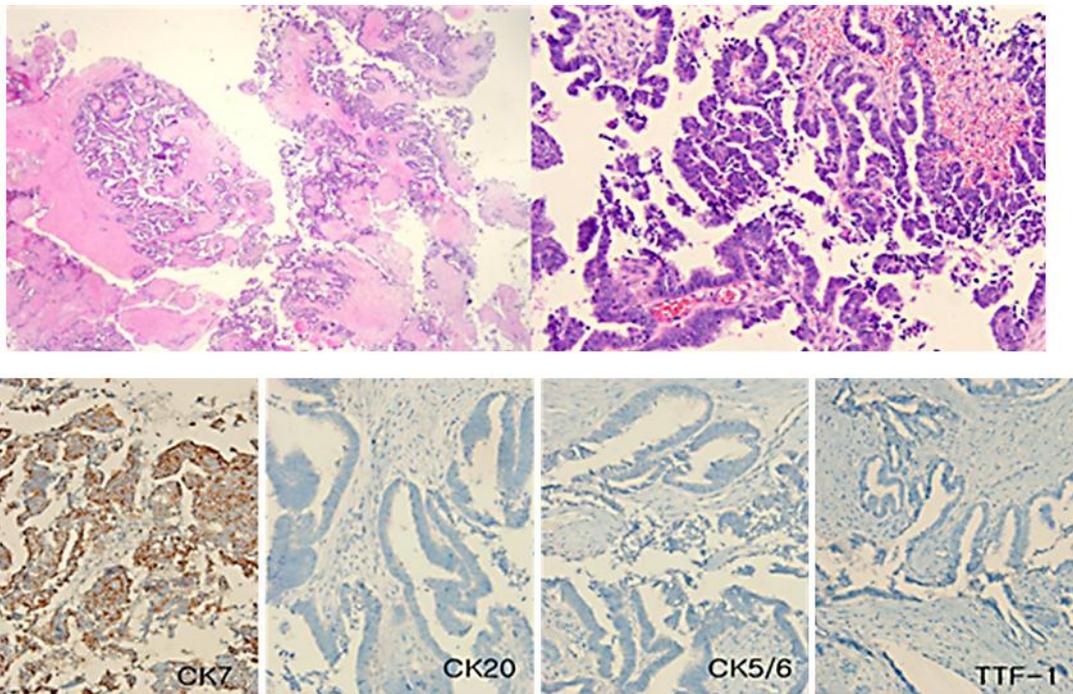
### Disclosure Statement

The authors have no conflict of interest to declare.

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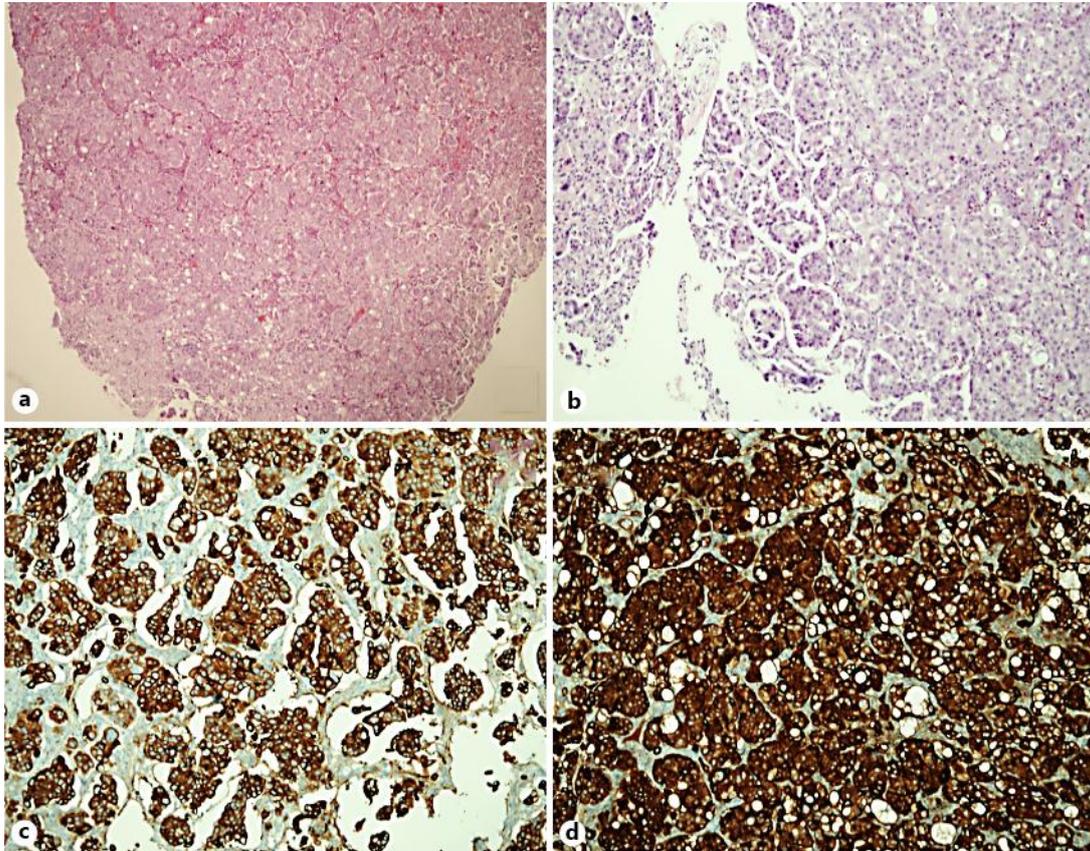
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**Fig. 1.** Microscopic findings of the left neck mass. The tumor tissue shows papillary growth with fibrotic stroma, and nuclear atypia is conspicuous (upper lane, HE stain). The tumor cells are positive for CK7, but negative for CK20, CK5/6, and TTF-1, which are more suggestive of müllerian origin than of gastrointestinal or lung origin (lower lane, immunohistochemical stains).

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**Fig. 2.** **a** The lymph node is entirely replaced by cancer cells (HE.  $\times 40$ ). **b** The cancer cells are arranged in a nested pattern with a glomerulus-like pattern at the periphery (HE.  $\times 100$ ). **c, d** The cancer cells show positive staining for CK7 and epithelial membrane antigens.