

Primary Peritoneal Carcinoma: A Case Report of Primary Peritoneal Papillary Serous Adenocarcinoma in a 59 Year Old Postmenopausal Woman

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Abstract Primary peritoneal carcinoma is rare and may present with symptoms similar to epithelial ovarian carcinoma which has some histological similarities. This is a case report of a 59 year old woman Para 2 who presented with massive ascites, abdominal pain, difficulty in breathing, abdominal mass and weight loss of three months duration. No conclusive diagnosis could be made after an ultrasound scan and CT-scan investigations showed the ovaries were of normal size but serum CA125 level was markedly elevated. No organ including the ovaries was identified as primary source of the tumour at an initial laparotomy. Specimens were however taken from multiple areas including the peritoneum and the omentum for histopathology. Reports of cytology and histology specimens indicated papillary serous adenocarcinoma with likely primary source being the ovary. Two cycles of neo-adjuvant chemotherapy was administered followed by re-laparotomy where total abdominal hysterectomy plus bilateral salpingoophorectomy, omentectomy and surgical reduction of tumour seedlings. Final histopathology ruled out papillary serous adenocarcinoma of the ovary which has similar histological features with papillary serous adenocarcinoma of the peritoneum. Four cycles of adjuvant platinum plus paclitaxel based chemotherapy was then administered and serum CA 125 declined from 214u/ml to 49.5u/ml with no clinical signs of residual disease on completion of chemotherapy. The patient is now on followed-up. **Conclusion:** Primary peritoneal carcinoma though rare, should be suspected if the peritoneum is involved in a malignancy of an unknown primary source. Management of primary peritoneal carcinoma should involve surgical debulking and appropriate chemotherapy as in ovarian carcinoma.

Keywords: *primary peritoneal carcinoma, carcinoma of unknown primary origin, peritoneal serous papillary adenocarcinoma, primary peritoneal malignancies*

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1. Introduction

A primary peritoneal carcinoma (PPC) is a malignant process predominantly involving the peritoneum [1]. Malignancies that arise mainly from the peritoneum were previously referred to as malignancies of unknown origin probably because the peritoneum is mostly noted to be affected by tumours from other organs in the abdominal cavity. PPC is a rare tumour and histologically identical to epithelial ovarian carcinoma, it is differentiated from epithelial ovarian carcinoma based on extent of gross ovarian involvement and microscopic invasion of the cortex [1,2,3]. When the tumor involves the extraovarian peritoneum significantly and the ovarian surface minimally or not at all, it is generally considered to be of peritoneum in origin [1,2]. Due to a common embryonic origin of the ovary and the peritoneum, carcinoma of the ovary and primary peritoneal carcinoma have much

histological similarity [4]. Epithelial ovarian cancers and primary peritoneal cancers arise from the germinal epithelium which develops from the coelomic epithelium [1,4].

Whilst the early stages of PPC may be asymptomatic, symptoms of the advance stages of the disease include abdominal distention, non-specific abdominal pain and dyspnoea all as a result of massive ascites which could be mistaken for an ovarian carcinoma or other conditions that present with similar symptoms. Investigations using abdominal ultrasound or CT-Scan may show presence of ascites, abdominal masses "omental cakes" and tumour nodules on various peritoneal surfaces with normal size ovaries but elevated tumour makers CEA and CA-125. Epithelial ovarian carcinoma has similar clinical features and elevation in levels of tumour makers CEA and CA-125. The main treatment for PPC which has been successful included debulking surgery in combination with platinum plus taxane combination chemotherapy [2,5].

In view of the difficulties in arriving at a diagnosis of primary papillary serous adenocarcinoma of the peritoneum, its diagnosis may be missed or may be considered as carcinoma of unknown primary origin. This report would help draw attention for PPC to be considered in the working diagnosis when patients present similarly.

2. Case Report

A 59 year old Para 2 postmenopausal woman was first seen in March 2014 at our gynaecology clinic with complaints of abdominal distention, abdominal pain, with difficulty in breathing which was of gradual onset and of three days duration. Three months prior to her presentation to the gynaecology clinic with the above complaints she had perceived abdominal fullness and a general sense of ill health for which she has been investigated in another hospital. She developed obvious abdominal distention with time which was associated with abdominal pain. She could also feel a vague mass in her abdomen just days before presentation at the gynaecology clinic. Multi-disciplinary consultation became necessary as no specific diagnosis could be made.

She had no dizziness, palpitations, headache, nausea, vomiting, diarrhoea nor constipation. She had no vaginal bleeding or discharge, and had no dysuria, frequency or urgency. She however had dyspnoea, weight loss and early satiety. There was no past medical and family history of significance. There was no positive history of abuse of alcohol, drugs or smoking. Her menarche was at

16 years and coitarche was at 23 years. She had regular 28 days menstrual cycles with normal menstrual flow lasting 4 days with no dysmenorrhoea. She had never used any form of modern contraception and never had any form of screening for gynaecological cancers.

She looked chronically ill and anxious, not febrile, not pale, not jaundiced. She was 162cm tall and weighed 64kg, a body mass index (BMI) of 24.4kg/m² and body surface area (BSA) of 1.7. She had mild pedal oedema with bilateral inguinal lymphadenopathy largest measuring about one centimeter. The thyroid gland and the breasts were normal. Both the cardiovascular system, respiratory system, the liver, spleen and both kidneys were all normal. The abdomen was grossly distended with oedematous abdominal wall and positive fluid thrill sign. There was a vague mass palpable in the mesogastrium with no area of tenderness. Speculum examination of the vagina showed healthy vulva, vagina and cervix. Rectal examination showed external haemorrhoids with contact bleeding but normal anal sphincter tone. The cardinal and utero-sacral ligaments were normal. The pouch of Douglas had no obvious masses noted.

The patient was informed of a provisional diagnosis of ovarian carcinoma after clinical examination. She gave her consent for investigations, surgical management and chemotherapy after counselling. Results of some initial investigations conducted and histopathology reports of biopsies taken during the first laparotomy are summarised in the [Table 1](#) below.

Table 1. Summary of some radiological, endoscopy and pathological investigations

INVESTIGATIONS	REPORTS
Ultrasound	The liver appears slightly shrunk and course. No cyst or calcification seen. No other masses seen. The spleen, kidneys and gallbladder are all normal. No pleural or pericardial fluids seen. The pelvic organs appear normal. Significant ascites noted. Diagnosis: ? Chronic Liver Disease, ?? hollow viscous perforation
CT-Scan 1	There is an ill-defined hypodense collection within the peritoneum, (hepato-renal space and spleno-renal space) extending into the region around the bowel system and bladder. Ascites is present. Observation of other organs reported as normal.
2	There is an ill-defined lobulated/polypoid and contrast enhancing mass seen to arise from the anterior bladder wall of the recto-sigmoid colon. It measures 7x6cm and extends/infiltrates the posterior bladder wall. Observation of other organs reported as normal. Diagnosis: Recto-sigmoid CA
Endoscopy	Normal upper gastrointestinal tract, rectum; 3 rd degree haemorrhoids
Ascitic fluid cytology	Numerous malignant cells, showing metastatic adenocarcinoma Psammoma bodies. Large amount of serous material
Initial laparotomy Biopsy report	Consistent with a papillary adenocarcinoma, likely primary source being the ovary.

During the first laparotomy the findings included massive ascites about 4 litres, the uterus was normal at the fundal area but there were nodular tumour seedlings of various sizes ≥ 2 centimetres on the bladder peritoneum and pouch of Douglas both adhering to the lower parts of the uterus, both ovaries and fallopian tubes were normal in size with intact surface without any macroscopic tumour on their surface. The parietal peritoneum was thickened and most of its surface was seeded with tumour sizes ≥ 2 centimetres. The gastrium, small bowel, the upper part of the rectum and sigmoid colon were normal. There was an "omental cake" and extensive tumour seedling on the upper parietal peritoneum and under surface of the

diaphragm, around the transverse colon which extended to the splenic flexure. There was no evidence of involvement of the pancreas, gall bladder, the spleen and liver parenchyma, but the under surface of the liver had tumour nodules ≥ 2 centimetres. Because the primary tumour could not be identified with any organ in the abdomen, biopsy was only taken from the bladder surface, the pelvic peritoneum in the pouch of Douglas, the parietal peritoneum, the "omental cake" and some para-aortic lymph nodes were taken for histopathology. There was however some suspicion the primary tumour may involve the transverse colon but that was not possible to confirm. A colostomy was done as a palliative and preventive

treatment of any large bowel obstruction that may arise from the progression of any tumour involving the transverse colon. An abdominal drainage tube was left in-

situ for two weeks to drain the ascitic fluid during the post-operative period. Some findings at the initial exploratory laparotomy are included in [Figure 1](#) a-c below.

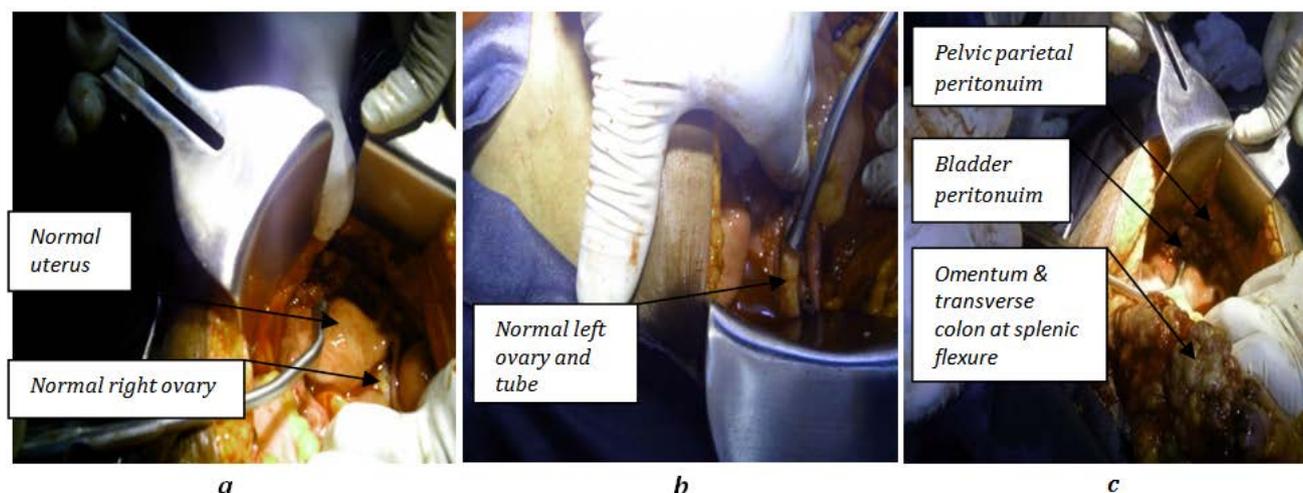


Figure 1. a: Findings at first laparotomy, **b:** Findings at first laparotomy, **c:** Structures with Tumour nodule

On account of the cytology and histopathology reports, and findings at surgery, two cycles of neo-adjuvant chemotherapy of Paclitaxel $135\text{mg}/\text{m}^2$ with Cisplatin $100\text{mg}/\text{m}^2$ was given to downstage the tumour. Shrinkage of the “omental cake”, cessation of ascites and a drop in the CA 125 values were observed. A re-laparotomy was performed at which total abdominal hysterectomy plus bilateral salpingo-oophorectomy, omentectomy and surgical reduction of the tumour on the peritoneal surfaces was done with the primary tumour considered to be peritoneal carcinoma in origin. The colostomy was also then reversed as the transverse colon and rest of large bowel were normal. Except for the ovaries that appeared normal, the finding at laparotomy on both occasions was equivalent to figo stage III C ovarian carcinoma. There

was adequate pre-operative preparation of the bowel, administration of broad spectrum antibiotics and blood transfusion on all two occasions that laparotomy was performed. The uterus together with the tubes and ovaries, the “omental cake”, other tumour mass and lymph nodes removed during the re-laparotomy are shown in [Figure 2](#) a-c below. Re-laparotomy pathology reports summary showed macroscopy:-normal size uterus, ovaries and tubes. Omental tissue 17cm long with lymph nodes and resected bowel about 6 cm long. Microscopy showed tumour deposits and infiltration of the uterus, omentum, large bowel and lymph nodes is composed of papillary structures. There were tumour deposits on the serosa of ovaries, with corpora albicantia.

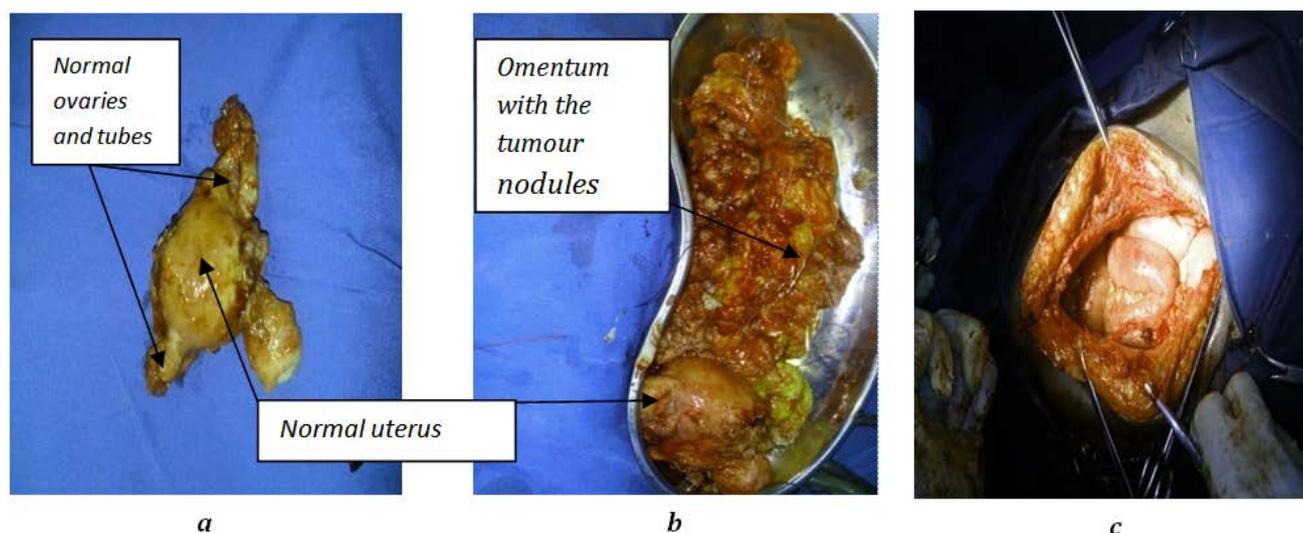


Figure 2. a: Findings after hysterectomy (at re-laparotomy), **b:** Showing tissues after debulking (at re-laparotomy), **c:** Abdomen just before closure after debulking surgery

She received the third and fourth cycles of adjuvant chemotherapy of Paclitaxel $135\text{mg}/\text{m}^2$ with Cisplatin $100\text{mg}/\text{m}^2$ at three and six weeks respectively after the re-laparotomy was performed. Cisplatin $100\text{mg}/\text{m}^2$ was replaced with carboplatin $250\text{mg}/\text{m}^2$ for the fifth and sixth cycles based on deteriorating GFR of $50\text{ml}/\text{min}$

from $>89\text{ml}/\text{min}$ at the start of chemotherapy. The urea and creatinine also deteriorated from $2.9\text{mmol}/\text{l}$ to $4.6\text{mmol}/\text{l}$ and $49\text{mmol}/\text{l}$ to 127mmol respectively. The electrolytes, liver function and white cell counts fluctuated within the normal range whilst thrombocytosis of $757 \times 10^9/\text{l}$ at the initial state to $467 \times 10^9/\text{l}$ at completion

chemotherapy was observed. Her haemoglobin fluctuated from 11.4g/dl to 8.1g/dl, she was given adequate haematinics, blood transfusion and Erythropoietin for any anaemia that was observed. Other investigation which included chest X-ray, ECG, HBsAg, HCV, HIV I&II, sickling test, fasting sugar, urine and stool routine examinations were all normal during the treatment. The tumour marker CA 125 declined from 214u/ml before debulking surgery to 49.5u/ml by time adjuvant chemotherapy was completed. Her condition has been stable and satisfactory during follow-up after completion of chemotherapy.

3. Discussion

The management of stage IIIC primary peritoneal papillary serous adenocarcinoma in a 59 year old woman, para 2 and 5 years postmenopausal was presented to help draw attention for PPC to be considered in the working diagnosis of patients who present with clinical features similar to that of ovarian carcinoma. Primary peritoneal carcinoma is a rare malignancy with isolated cases being diagnosed and treated even in large oncology centers across the world [2,6,7]. The incidence of ovarian cancers is high in the age range of 50 to 70 years, within the range of mean age of 57.6 and 64.4 years in reviews of 27 and 23 patients respectively with primary peritoneal carcinomas [1,5,7].

PPC presents with symptoms similar to that of ovarian carcinoma. Therefore investigations that are conducted are usually similar but making a diagnosis of primary peritoneal carcinoma before surgery if the ovaries are not the source of the carcinoma is quiet difficult. However if the cytology is suggestive of carcinoma of the ovary but ultrasound and CT-Scan finding are normal, then the suspicion should be high especially if cytology of ascitic fluid is positive and CA 125 values are high as in the case presented. In one review of 55 cases of carcinoma of the ovary, only 5.8% had positive malignant cytology while all 23 cases with PPC had positive cytology [7]. In the present case the diagnosis of PPC was not made after ultrasound and CT- scan investigations had been done. Experience in the radiological diagnosis of primary papillary serous carcinoma of the peritoneum has shown that the presence of peritoneal masses, extensive omental calcifications, and the absence of an ovarian mass on CT-particularly in postmenopausal women is highly suggestive of primary papillary serous carcinoma of the peritoneum and should alert the radiologist to the possibility of this diagnosis[8]. Primary peritoneal serous papillary adenocarcinoma is similar to papillary serous adenocarcinoma of the ovaries but the ovaries are not involved in the former and appear normal at surgery [9] as was found in this case. Given the clinical presentation of this case, the diagnosis of PPC could have been an earlier consideration if the patient had a history of bilateral salpingoophorectomy.

The management of PPC is surgical debulking plus platinum and taxane based chemotherapy as in ovarian cancer [2,5]. The challenge is delaying debulking surgery because of uncertainty in diagnosis or when the ovaries are not known to be involved as may be suggested by radiological investigations. In the case presented, a

clinical diagnosis at initial exploratory laparotomy was difficult to establish as no primary tumour was identified at surgery. Despite the normal findings of the ovaries at exploratory laparotomy, based on ascitic fluid cytology reports and histology reports of biopsies that were taken, neo-adjuvant chemotherapy was initiated. During the re-laparotomy, debulking surgery was performed; however some tumour deposits on the surface of the liver, inaccessible areas on the under surface of the diaphragm and extensive lymph node dissection was not done. The findings at surgery and the clinical stage IIIC provided followed the figo staging for ovarian cancer; which has similar clinical features and pattern of spread in advanced stage of the disease except for the ovaries which remain grossly normal findings in PPC. In patients with type IIIC or IV ovarian cancer, neo-adjuvant chemotherapy could reduce postoperative morbidity and mortality without improving survival [10,11]. However, accurate tumour typing and grading of ovarian cancer is impossible when preoperative chemotherapy is used. It can be difficult to confirm the presence residual tumour, making it imperative that pre-chemotherapy tissue biopsies are obtained [12]. In the case presented, no tissue was taken from the ovaries for biopsy at the initial laparotomy because the ovaries looked grossly normal in the presence of extensive tumour nodules on the peritoneum, omentum, undersurface of the liver and the diaphragm.

The papillary serous adenocarcinoma type which was present in this patient are responsive to platinum based chemotherapy [2,13]. The general condition of patient improved following surgical cytoreduction and continuation of platinum based chemotherapy. CA-125 values were declining but after the fourth cycle, carboplatin was substituted for cisplatin because of observed deterioration in renal function.

Prognosis and survival data is still limited on PPC [2]. Our patient presented with an advanced disease her prognosis and survival still remains unknown. However a review by Nam JH et al showed a median overall survival time of 41 months and 5 year survival of 18.1% [5]. Survival rates however may be less than Survival rates for similar stage ovarian cancer [10,14].

4. Conclusion

The diagnosis of a primary peritoneal cancer may be initially difficult because the peritoneum is often involved in metastasis from other organs in the peritoneal cavity. Primary peritoneal cancer though rare, should be suspected if the peritoneum is involved in a malignancy of an unknown primary source. Management of primary peritoneal cancer should involve surgical debulking and appropriate chemotherapy as in ovarian carcinoma.

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