

Primary peritoneal carcinoma: an uncommon entity

Royal College of Surgeons in Ireland Student Medical Journal. 2011;4(1):28-30.

Background

Primary peritoneal carcinoma (PPCa) is a rare neoplasm originating from the cells of the peritoneal cavity, first described in 1959 by Swerdlow.¹ It is less common than peritoneal carcinomatosis, an advanced stage of abdominal malignancy with extensive metastasis of tumours to the peritoneum. PPCa was also previously known as extra-ovarian primary peritoneal carcinoma or primary serous papillary carcinoma of the peritoneum.

Histologically, PPCa cannot be differentiated from primary epithelial ovarian carcinoma, as the ovary and the peritoneal epithelium share a common embryological origin. As such, the diagnosis of PPCa is based on minimal or non-involvement of the ovaries.² Overall, PPCa is thought to account for 10% of ovarian malignancies. Previous reports have identified women with primary peritoneal cancer as being older and with late menarche.⁴ PPCa has been reported most frequently in postmenopausal women with a mean age of 64.4 years.⁴

The case

We report the case of a 58-year-old woman who presented with a three-day history of progressive generalised abdominal pain slowly localising to the left iliac fossa and the back. The pain was associated with constipation and abdominal distension. The patient had a background history of hypothyroidism and hypercholesterolaemia. Her social and family history was non-contributory. On physical

examination, she was in tachycardia but was otherwise haemodynamically stable. There was generalised abdominal guarding and focal tenderness over the left iliac fossa. Bowel sounds were present and there was no evidence of ascites or bleeding per rectum. Clinical suspicion at that time was of a perforated diverticulum with diverticulitis causing an acute abdomen.

Further investigations showed significant raised C-reactive protein (248mg/L). Chest and abdominal x-ray did not suggest any abnormality. An abdominal CT scan demonstrated a large inflammatory mass in the lower abdomen and left iliac fossa. The mass was thought to represent multiple loops of markedly thickened small bowel with adjacent inflammatory change (**Figure 1**). There was no evidence of obstruction. A diagnosis of probable intra-abdominal malignancy was made.

The patient underwent laparotomy. During the operation, a large mass was identified adjacent to the mesentery of the small bowel and transverse colon (**Figure 2**). The mass was well circumscribed with a smooth outline. Thickened loops of small bowel were also seen. The mesenteric mass was excised along with a short segment of adherent small bowel. Both ovaries had a normal gross appearance. Since abdominal lymphoma is a common small bowel malignancy, it was deemed the most likely diagnosis.

The patient's post-operative course was uneventful. The resected mass and small

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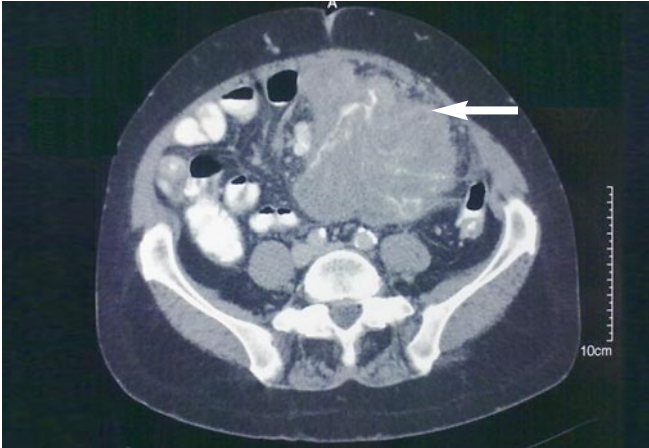


FIGURE 1: Computed tomography (CT) scan showing a left abdominal mass (arrow).

bowel were sent for histological evaluation. The mass measured 155 x 120 x 80mm with attached mesenteric fat, and in aggregate weighed 570g. The external aspect of the mass was smooth, and the cut surface was heterogeneous with lobules of creamy material and areas of haemorrhage. No cystic areas were seen.

The resected length of small bowel measured 280mm and appeared to be dusky and haemorrhagic with areas of exudate on the surface. The mucosa was friable in areas. The appearance was consistent with the clinical suspicion of a lymphoma.

Microscopically, the tumour was well circumscribed and composed of solid sheets of malignant cells lining micro-papillary structures (Figure 3a). Abundant psammomatous calcification and necrosis was identified (Figure 3a). Vascular invasion was evident.

The serosal surface of the small bowel was infiltrated with tumour but none of the 11 mesenteric lymph nodes retrieved

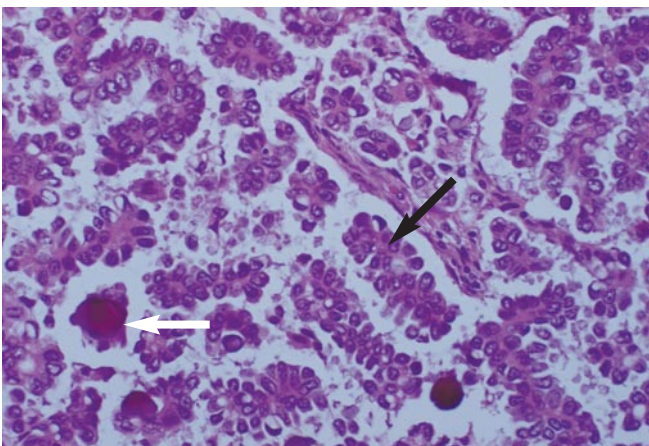


FIGURE 3a: Microscopic findings of tumour cells showing micro-papillary structures (black arrow) and psammoma bodies (white arrow) (H&E staining, magnification x400).

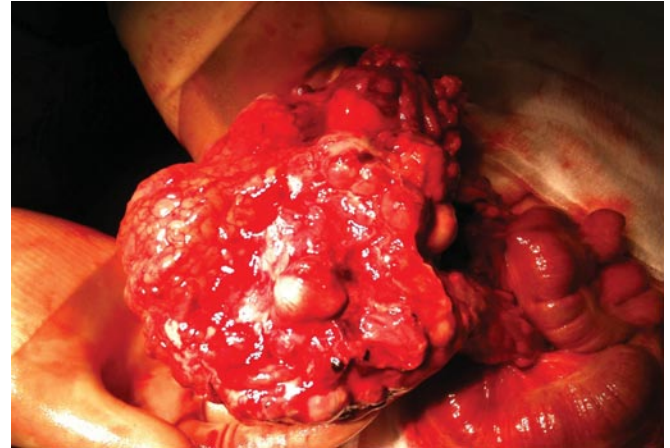


FIGURE 2: Macroscopic appearance of the resected mass during laparotomy.

were involved (Figure 3b). Following discussion at a multidisciplinary conference involving the pathology, surgical and oncology teams, the consensus was that because of the absence of ovarian pathology, the tumour should be considered as a primary peritoneal micro-papillary serous carcinoma. Following the diagnosis, an abdominal and pelvic ultrasound showed endometrial fundus thickening greater than 10mm, but no cystic or solid lesions were noted in the ovaries. A repeat CT abdomen identified a few small lymph nodes in the retroperitoneal space with no mesenteric deposits. In addition, there were no suspicious metastatic lesions or lymph nodes involved.

The tumour marker cancer antigen 12-5 (CA12-5) was shown to be elevated at 207kU/L (0-35kU/L). Serologies for tumour markers CA19-9 and carcinoembryonic antigen (CEA) were negative. The patient was subsequently referred to a medical oncologist for six courses of chemotherapy, specifically carboplatin 770mg/m² and paclitaxel 276mg/m².

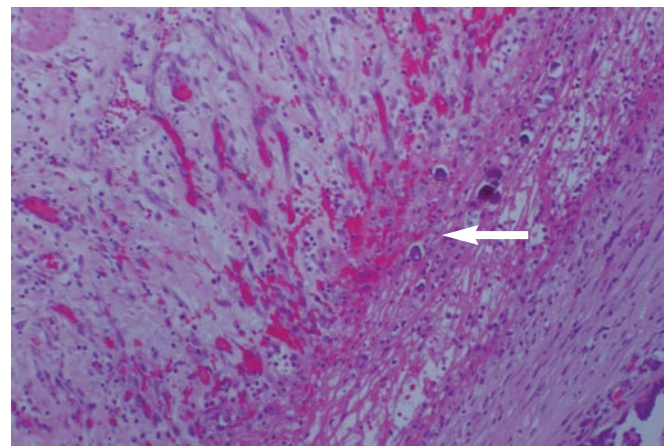


FIGURE 3b: Microscopic image showing tumour infiltration of the small bowel serosal surface (arrow) (H&E staining, magnification x100).

Discussion

About 1.9% of acute abdomen cases are associated with intra-abdominal malignancy, and this figure increases to 10% in patients over 50 years.⁵ PPCa is best considered as a diagnosis of exclusion; the more evident diagnosis of metastatic carcinoma or primary lymphoma must be considered first. Presenting complaints of patients with PPCa are similar to that of primary ovarian cancer, which include abdominal pain and distension, back pain, irregular period and dyspareunia.⁶ Ascites is the most frequent presenting clinical sign.⁶

In this case, there was no clinical, radiological or operative suspicion of a PPCa. The patient presented with abdominal and back pain associated with abdominal distension and constipation, but ascites was not present during physical examination and laparotomy. The patient's presenting symptoms are often associated with benign conditions and warrant thorough investigation. The differential diagnosis for left iliac fossa pain includes diverticulitis, ischaemic bowel, inflammatory bowel disease, bowel obstruction, colorectal carcinoma and ovarian carcinoma. In this case, an abdominal aortic aneurysm was also suspected given the patient's complaint of back pain. However, relevant negative symptoms, including blood in the stool, absent bowel sounds, ascites, consistent family or social history, chest pain, bruits or an abdominal pulsating mass, helped to rule out these differential diagnoses.

Imaging techniques aided in the diagnosis and management of the patient. In fact, the abdominal CT scan revealed a large inflammatory mass in the left iliac fossa that was not detected on chest and abdominal x-ray. Such a mass suggests intra-abdominal malignancy; however, a definitive diagnosis cannot be reached without histological confirmation. A laparotomy was performed with curative intent. However, the resected mass was

subsequently evaluated, which established the diagnosis of PPCa. PPCa and ovarian cancer have similar responses to chemotherapy and are treated with the same chemotherapeutic agents.⁷ The standard first-line chemotherapeutic agents in ovarian carcinoma or PPCa are carboplatin and paclitaxel.⁸ Intraperitoneal chemotherapy was reported to improve overall survival in patients with ovarian carcinoma or PPCa by 5.5 months when compared to intravenous administration. This was attributed to a greater bioavailability of chemotherapeutic agents when delivered directly into the peritoneal cavity.⁹ The patient underwent a primary cytoreductive surgery and six cycles of chemotherapy. Recurrence of the malignancy has yet to be detected.

The prognosis of PPCa is generally poor. A recent systematic review reported a median survival for PPCa of two to six months less than survival rates for ovarian cancer.¹⁰ However, the largest published UK series of patients with PPCa suggested similar prognosis and survival in both groups – 21 months and 19 months for PPCa and ovarian carcinoma, respectively.⁴ In summary, tissue diagnosis must first be established to diagnose a PPCa. Since the presenting complaints and initial clinical investigations from patients with PPCa are similar to those with primary ovarian cancer, intra-operative assessment of the ovaries with corresponding ovarian imaging is essential to exclude a primary ovarian epithelial tumour. PPCa should be considered in patients who present with abdominal pain, particularly in the presence of a mass on imaging and absence of ovarian pathology.

Acknowledgements

We would like to thank the Department of Radiology at Beaumont Hospital for providing the radiological image.

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