CASE REPORT

Abdominal cocoon

Christian B S Katz,¹ Robert T Diggory,² Abdus Samee³¹Department of General Surgery, Shrewsbury and Telford NHS Trust, Telford, UK²Department of Surgery, Princess Royal Hospital, Telford, UK³Department of General Surgery, University of North Staffordshire, Stoke on Trent, UK

Correspondence to

Christian Bernard Katz,
christiankatz@hotmail.com

Accepted 25 February 2014

SUMMARY

Intestinal obstruction secondary to cocoon formation is not common. We report a case of a patient who had presented with abdominal pain and distension accompanied by vomiting. Investigations, laparotomy and histology together revealed primary peritoneal carcinoma as the cause of the patient's symptoms.

BACKGROUND

The differential diagnosis on discovering an abdominal cocoon is broad. It is an unfamiliar condition to most clinicians. Primary peritoneal carcinoma is rare in itself but with up to 10% being misdiagnosed as ovarian cancer. It is a condition that clinicians should consider in female patients presenting with obstructive or vague abdominal symptoms. The purpose of this case report is to familiarise general practitioners and surgeons with this disease.

CASE PRESENTATION

A 69-year-old woman was referred with worsening dull abdominal pain, distension and vomiting for a week. She had reported reduced appetite and a weight loss of approximately 13 kg along with the dull abdominal pain and gradual distension for a year. There was no change in bowel habit and she had been previously fit and well with no significant morbidities. She was an ex-smoker, having smoked 15 cigarettes daily for 40 years. Her surgical history included a previous hysterectomy for menorrhagia.

Vital parameters were normal. Examination revealed a large distended, soft yet diffusely tender abdomen with oedema of the lower abdominal wall. The abdomen was dull to percussion and bowel sounds were normal. Rectal examination was unremarkable.

INVESTIGATIONS

Blood results showed a white cell count of $10.9 \times 10^9/\text{litre}$ with 90% neutrophilia. Haemoglobin was 12.9 g/dL. C reactive protein was elevated at greater than 90 mg/L. Liver function tests were unremarkable but her albumin was low measuring 30 g/L. Carcinoembryonic antigen measured 1 ug/L and α -fetoprotein was mildly elevated at 2 ku/L. CA125 was mildly elevated at 148 u/mL.

Erect chest X-ray was normal. Abdominal X-ray revealed a non-specific gaseous pattern; however, on closer analysis, revealed a diffuse sac-like structure arising from within the pelvis (figure 1).

A CT scan of the abdomen demonstrated a massive fluid-filled sac arising from the small bowel

Figure 1 Plain abdominal film showing sac-like structure arising from pelvis (arrows).

mesentery that engulfed or 'cocooned' the entire small bowel (figure 2). Jejunal loops appeared thickened and dilated with no obvious transition point to suggest obstruction. The large bowel was

Figure 2 Contrast CT showing abdominal cocoon (the cocoon had engulfed the entire small bowel and mesentery—arrows).

CrossMark

To cite: Katz CBS, Diggory RT, Samee A. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-203102

found to be spared and unremarkable. The pelvic organs, including ovaries, were reported to be normal. A small left-pleural effusion was noted however it was felt less relevant in establishing the diagnosis and in providing the initial treatment.

DIFFERENTIAL DIAGNOSIS

The working differential diagnosis was sclerosing encapsulating peritonitis (SEP) or an ovarian malignancy. It was debated at this time whether to perform fine-needle aspiration of the fluid but with fear of causing seeding, it was decided to perform an exploratory laparotomy.

TREATMENT

Laparotomy revealed a thick fluid bag containing the entire small bowel, consistent with the CT findings. There was no evidence of complete acute or subacute obstruction. The large bowel was spared. Pelvic organs could not be visualised but were felt to be normal.

A tissue biopsy of the sac along with the fluid was sent for microbiological and histological analysis. The sac was debulked as much as possible. Postoperative recovery was unremarkable and the patient was shortly discharged.

Gross pathological examination confirmed fibrofatty peritoneum and white fibrous tissue of the cyst wall itself. Histology revealed a moderately differentiated adenocarcinoma with an intestinal/endometroid pattern. Immunohistochemical tests suggested the primary site being that of peritoneum or ovary.

OUTCOME AND FOLLOW-UP

The findings were discussed in a multidisciplinary meeting. Palliative management was decided and appropriate chemotherapy was administered. The patient was followed up jointly by gynaecologists and oncologists. Following discharge, she was re-admitted with similar symptoms and was managed conservatively.

DISCUSSION

A definitive diagnosis as to the cause of abdominal cocoon is difficult to achieve in the preoperative period. Symptoms and signs are often non-specific and the condition is largely unfamiliar to clinicians. The differential diagnosis is broad on discovering a cocoon. The condition can be seen in SEP, abdominal tuberculosis or in primary peritoneal carcinoma (PPC).

PPC is a rare form of peritoneal cancer that arises from mesothelial cells making up this serous tissue, however non-serous tumours have also been described.¹ These cells derive from the mesoderm during embryological development. The germinal epithelium of the ovary derives from the same origin and thus the behaviour of PPC mimics that of epithelial ovarian carcinoma (EOC). First described in 1959 by Swerdlow,² it is often misdiagnosed as an ovarian cancer and in fact PPC is found in 10% of postmortems on individuals with a diagnosis of ovarian cancer.

Studies into the genetic basis and molecular changes giving rise to this cancer have suggested a possible link to p53 gene. A study performed showed 83% of 29 patients with PPC with overexpression of this tumour suppressor gene.³ However, the same authors also suggest that PPC may be multifocal in origin.^{1 3}

The risk factors for PPC are unknown.¹ An epidemiological study comparing PPC with ovarian cancer demonstrated the former to be associated with older women with late menarche.⁴

Clinical presentation is similar to that of advanced epithelial ovarian cancer. Most reported cases demonstrate a median age

of 56–66 years and predominant occurrence in Caucasians. Symptoms include abdominal distension and pain with associated gastrointestinal symptoms. Ascites is found in approximately 85% of cases.¹ Studies have shown elevated serum CA125 levels in patients with PPC.⁵

PPC can be treated both surgically and using chemotherapy, with similar treatment regimes used with epithelial ovarian cancer—a reflection of the similar natures of PPC and EOC. However, the use of debulking or cytoreduction in PPC may be limited, due to its potential anatomical multifocal origin. There is debate within several studies to date as to the impact on optimal and suboptimal debulking of tumour to the median survival.^{6–8} However, there is agreement on the positive effects on median survival following cytoreductive surgery. Currently, according to the Gynaecology Oncology Group criteria, the first-line chemotherapeutic regimen of ovarian cancer is also recommended for patients with PPC postoperatively.⁴ Platin-based and taxane-based agents are commonly used in current regimens.

Prognosis is poor with median survival of 12–24 months and a 5-year survival rate of 10–20%.¹

Another condition giving rise to abdominal cocoon is SEP. This condition can be classified as either idiopathic or secondary to a host of other conditions. The idiopathic form has been classically described in adolescent females from tropical or subtropical countries. Common secondary causes are seen in peritoneal dialysis patients, individuals with tuberculosis and historically those treated with β -blocker practolol.⁹ Clinically, it presents with recurrent episodes of acute or subacute small bowel obstruction, weight loss, nausea and anorexia and at times with a palpable abdominal mass caused by the formation of a fibro-collagenous membrane that wraps around the loops of the intestine, producing obstruction.

Learning points

- ▶ Abdominal cocoon is a rare cause of small bowel obstruction associated with numerous different aetiologies such as the primary peritoneal carcinoma (PPC), abdominal tuberculosis and peritoneal dialysis.
- ▶ PPC is a rare cancer that behaves similar to epithelial ovarian cancer pathophysiologically, histologically, clinically and therapeutically.
- ▶ PPC is a condition that general practitioners and surgeons should be aware of, particularly in female patients presenting with vague, obstructive abdominal symptoms and associated weight loss.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Eltabbakh GH, Piver MS. Extraovarian primary peritoneal carcinoma. *Oncology* 1998;12:813–19. discussion 820, 825–6.
- 2 Swerdlow M. Mesothelioma of the pelvic peritoneum resembling papillary cystadenocarcinoma of the ovary; case report. *Am J Obstet Gynecol* 1959;77:197–200.
- 3 Moll UM, Valea F, Chumas J. Role of p53 alteration in primary peritoneal carcinoma. *Int J Gynecol Pathol* 1997;16:156–62.

- 4 Eltabbakh GH, Piver MS, Natarajan N, *et al.* Epidemiological differences between women with extra-ovarian primary peritoneal carcinoma and women with epithelial ovarian cancer. *Obstet Gynecol* 1998;91:254–9.
- 5 Kawaguchi R, Yanase Y, Haruta S, *et al.* Paclitaxel plus carboplatin chemotherapy for primary peritoneal carcinoma: a study of 22 cases and comparison with stage III-IV ovarian serous carcinoma. *Case Rep Oncol* 2012;5:173–80.
- 6 Zhang C1, Li XP, Cui H, *et al.* Advanced primary peritoneal carcinoma: clinicopathological and prognostic factor analyses. *J Zhejiang Univ Sci B* 2008;9:435–40.
- 7 Eltabbakh GH, Werness BA, Piver S, *et al.* Prognostic factors in extraovarian primary peritoneal carcinoma. *Gynecol Oncol* 1998;71:230–9.
- 8 Look M, Chang D, Sugarbaker PH. Long-term results of cytoreductive surgery for advanced and recurrent epithelial ovarian cancers and papillary serous carcinoma of the peritoneum. *Int J Gynecol Cancer* 2004;14:35–41.
- 9 Eltringham WK, Espiner HJ, Windsor CWO, *et al.* Sclerosing peritonitis due to practolol: a report on 9 cases and their surgical management. *Br J Surg* 1977;64:229–35.

Copyright 2014 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- Submit as many cases as you like
- Enjoy fast sympathetic peer review and rapid publication of accepted articles
- Access all the published articles
- Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow