IMAGE OF THE QUARTER

IMAGING FINDINGS IN SCLEROSING ENCAPSULATING PERITONITIS

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INTRODUCTION
Sclerosing encapsulating peritonitis (SEP) is the most advanced stage of the peritoneal fibrosing syndromes, which may have a primary or secondary aetiology but are seen most frequently as a complication of peritoneal dialysis (PD). Sclerosing encapsulating peritonitis affects up to 1% of patients on PD, causing gastrointestinal symptoms, weight loss, ascites and deteriorating peritoneal dialysis function. We report two cases of SEP and the imaging findings.

CASE HISTORY 1
A female aged 43 presented with weight loss, diarrhoea, nausea, vomiting and malaise. Clinical examination revealed a malnourished patient with characteristic features of chronic renal disease and ascites. End-stage renal failure had resulted from pregnancy-induced hypertension ten years previously. Peritoneal dialysis, using hypertonic dextrose, was commenced that year and continued almost continuously for nine years. Renal transplantation failed. The adequacy of her PD deteriorated and she was transferred to intermittent haemodialysis. Relevant past medical history included recurrent episodes of peritonitis and long-term beta blocker treatment (metoprolol).

Investigations
Abdominal X-ray revealed medullary nephrocalcinosis, pancreatic calcification and a generalised abdominal haziness consistent with ascites. Ultrasound confirmed multi-loculated fluid collections and a thickened bowel wall (Figure 1). Three computerised tomography (CT) examinations were performed over a three-month period. These demonstrated most of the typical features of SEP, including loculated ascites, thickened bowel wall and a peritoneal membrane with adherent small bowel loops, rapidly progressive peritoneal and bowel wall calcification and clouding of the mesenteric fat (Figure 2 A–C). No CT evidence of bowel obstruction was observed. The patient proceeded to laparotomy for removal of the PD catheter. It was documented that ‘all intra-abdominal organs were encased in a thick pseudomembrane suggestive of SEP’. Peritoneal biopsy showed sub-peritoneal fibrosis and organised inflammatory exudate signifying an active, chronic inflammatory reaction with consequential fibrosis of the peritoneum.

CASE HISTORY 2
A 26-year-old male on PD presented with post-prandial vomiting leading to episodes of hypotension. He also complained of recurring left upper quadrant abdominal pain and persistent abdominal distension. Clinical examination confirmed ascites, features of chronic renal failure and signs of malnutrition. End-stage renal failure had been caused by obstructive uropathy secondary to posterior urethral valves 11 years previously. The patient was treated with PD for 11 years. Haemodialysis was commenced because of ultrafiltration failure. Renal transplantation failed. Past medical history incorporated several episodes of peritonitis, including a serious Staphylococcus aureus infection. The patient was prescribed atenolol for several years and tamoxifen more recently to treat the clinically suspected SEP. Compliance with medication was poor.

Investigations
C-reactive protein was elevated at 25 mg/ml. A plain abdominal film showed renal calcification and bone changes of renal osteodystrophy. Barium follow-through examination illustrated gastric, duodenal and proximal jejunal dilatation and a delayed transit time. Ultrasound revealed non-loculated ascites, and the CT with intra-peritoneal contrast demonstrated gastric dilatation, flecks of peritoneal calcification, bowel wall thickening, mesenteric fat stranding and tethering of the bowel loops to the posterior abdominal wall (Figure 3). A clinical diagnosis of SEP was made and the PD catheter removed.

![Ultrasound of the right upper quadrant showing ascites with multiple septations (s) and thickened small bowel loops (b).](image-url)
IMAGE OF THE QUARTER

DISCUSSION
Aetiology
Sclerosing encapsulating peritonitis (also termed encapsulating peritoneal sclerosis, sclerosing peritonitis and abdominal cocoon) was first described in 1980. It is not specific to PD and may be idiopathic or secondary to a range of causes such as drug therapy (particularly beta blockers) sarcoidosis, systemic lupus erythematosus and other primary abdominal organ disease. The precise aetiology of SEP is poorly understood but is thought to be related to the persistent expression of transforming growth factor β on mesothelial cells causing massive production of extracellular matrix, proliferation of peritoneal fibroblasts and a loss of mesothelial cells. This results in encasement of the intra-peritoneal organs, most notably the small bowel, in a ‘cocoon’ of fibrotic tissue. Stages of peritoneal opacification, tanning and fibrosis are encountered before SEP finally develops, these terms being encompassed under the general title of peritoneal fibrosing syndromes.

Significant contributing factors to the development of PD-induced SEP include duration of treatment with PD, recurrent or prolonged episodes of peritonitis, cumulative exposure to hypertonic glucose-based dialysis solutions and treatment with beta blockers. Theoretically, the factors related to PD solution damage of peritoneum are total glucose exposure, glucose degradation products following heat sterilisation and the acidic pH of PD fluid. Increasing duration of PD causes progressive thickening of the submesothelial zone in the peritoneum over an increasingly large area such that up to 20% of patients on PD for nine years develop SEP.

Clinical findings
Symptoms are usually insidious, although abdominal pain is often present. Bowel wall thickening hinders peristalsis and absorption, causing nausea, vomiting, weight loss and malnutrition. Fibrous bands may cause episodes of sub-acute or acute obstruction. The peritoneal fibrosis blocks lymphatic channels, causing ascites. A decline in ultrafiltration is noted, and dialysate may become blood-stained. Fever and an elevated C-
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FIGURE 3A

FIGURE 3A and B

Axial CT peritoneography demonstrates tethering of the small bowel posteriorly (b), peritoneal calcification (c) and mesenteric fat stranding (f).

reactive protein are seen in some patients.9

Radiology

- Plain abdominal films are often normal and may delay the diagnosis of SEPI. Possible findings include dilated small bowel loops, air-fluid levels and calcific plaques, none of which are diagnostic.10
- Barium studies show delayed transit, a dilated proximal bowel, a thickened bowel wall, fixed and rigid loops or complete obstruction. The mucosal pattern tends to be normal.11 These features are non-specific.
- Ultrasound is the most sensitive modality in detecting SEP. Early changes associated with peritoneal fibrosing syndromes may be identified: typically thickened bowel wall with a trilaminar appearance and adhesion of bowel loops to the anterior abdominal wall is seen. Peristalsis is often increased but may be normal or reduced.11 Early diagnosis allows appropriate treatment when it is most effective, i.e. before ‘full-blown’ SEP develops with associated high mortality.
- Computerised tomography features of SEP are seen later than the ultrasound features. Earliest changes of peritoneal fibrosing syndromes include thickened, adherent bowel loops and small bowel dilatation. Later manifestations include loculated ascites, increased density of mesenteric fat, obstruction, peritoneal and bowel wall calcification and ‘cocooned’ bowel.5,11
- Differential diagnosis includes tuberculosis, peritoneal mesothelioma and pseudomyxoma peritonei.

Prognosis

Survival is much improved if peritoneal fibrosis is detected and treated early; however, the condition often progresses even after removal of the dialysis catheter and with switching the patient to haemodialysis.1 Several treatments have been tried, including immuno-suppression,12 corticosteroids with total parenteral nutrition,5 tamoxifen5 and surgical debridement.14,15 Although various treatments have managed to cure patients, the mortality rate of established SEP remains between 56% and 93%.4

CONCLUSION

The radiological features of SEP have been discussed. In the correct clinical context, imaging findings may be diagnostic of SEP. Radiological investigations have an important role in aiding the diagnosis of peritoneal fibrosing syndromes at the earliest possible stage, when therapy is most effective.

REFERENCES

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All symposia are held at the Royal College of Physicians of Edinburgh unless otherwise stated. Further symposia may be added at a later date.

- Renal medicine 1 June
  
  Joint symposium: RCPE/RCPCH
- Paediatric research – how will it affect you? 16 September
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