BENIGN PERSISTENT PNEUMOPERITONEUM IN SYSTEMIC SCLEROSIS

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ABSTRACT

We describe a 50-year-old Chinese woman who had severe gastrointestinal manifestations from systemic sclerosis complicated by spontaneous pneumoperitoneum in the absence of either visceral perforation or pneumatosis cystoides intestinalis. This is a rare complication of systemic sclerosis; only four other cases have been reported. Recognition of this condition is important so as to avoid unnecessary surgery.

Keywords: *spontaneous pneumoperitoneum, systemic sclerosis, gastrointestinal manifestations*

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INTRODUCTION

Systemic sclerosis is a connective tissue disorder with multisystem involvement. Typical changes of the gastrointestinal tract are characterised by atrophy of muscularis propria and its replacement by collagen tissues; this reduces the peristaltic activity. Wide-mouthed diverticula of the colon also occur[1]. Pneumatosis cystoides intestinalis occur ocassionally and rupture of these cysts result in pneumoperitoneum and this has been associated with chronic benign pneumoperitoneum[2].

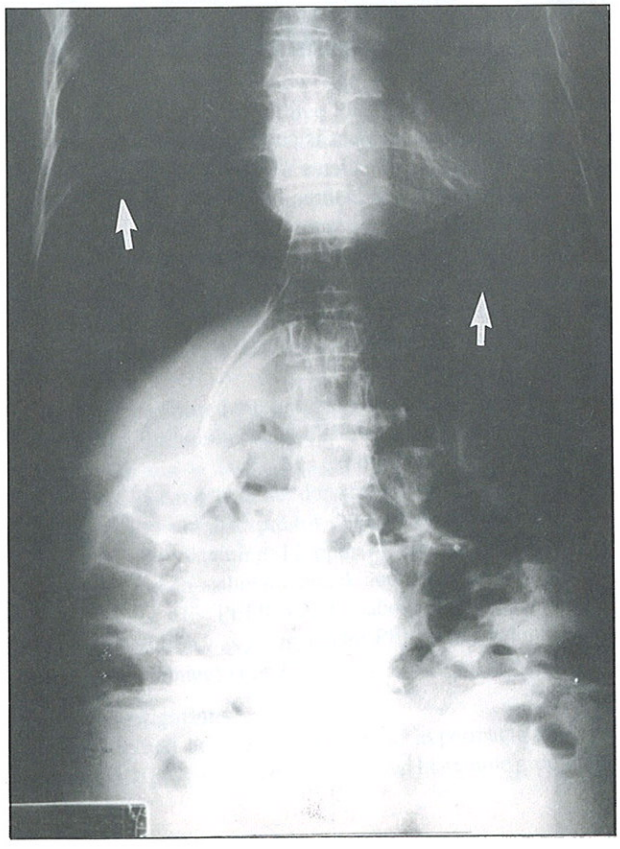
CASE REPORT

A 50-year-old Chinese woman was diagnosed to have systemic sclerosis 7 years ago when she presented with polyarthralgia, Raynaud's phenomenon and sclerodermatous facies. She was treated intermittently with non-steroidal anti-inflammatory analgesia for joint pain by her general practitioner. She presented to the University Hospital Kuala Lumpur with a 5-year history of intermittent and increasing abdominal distension associated with upper abdominal discomfort, significant weight loss and passing of loose stools at least twice a day. She also had mild dysphagia and complained of food sticking occasionally.

On examination, she was thin with mild sclerodermatous facies, moderate sclerodactyly and Raynaud's phenomenon of the fingers. Blood pressure was 120/70 mm Hg and pulse was 72/min. Cardiac and chest examination was normal. Her abdomen was moderately distended, soft, nontender with loss of hepatic dullness. There were no ascites or palpable masses. Bowel sounds were reduced and rectal examination revealed no abnormality.

Laboratory investigation revealed mild anaemia Hb 11.2g/ml with normal indices; serum urea, creatinine and electrolytes were normal; serum albumin was 32g/L, globulin was elevated at 66 g/L, liver enzymes included alkaline phosphatase were within normal limits. Anti-nuclear antibody was detected at 1:1024 (speckled pattern), anti-RNP was positive, anti–DNA antibody was negative, complement levels (C3, C4) were normal. Stool butterfat test was consistent with fat malabsorption. Serum iron was 10.2 umol/L (normal 11-29), serum folate was 20 nmol/l (normal 6.78-34), vitamin B_{12} was 234 pmol/L (normal 147-721). Serum calcium and phosphate levels were normal at 2.25 mmol/L and 1.0 mmol/L respectively. Radiographs of the abdomen revealed large amount of free air under the diaphragm (Fig 1). Contrast studies of the gastrointestinal tract included barium meal and follow-through showed dilated loops of the small bowel with increased mucosal folds, no other abnormality was noted. Gastrografin enema was normal. Pulmonary function tests showed mild restrictive defect, CO diffusion coefficient (DLCO) at 19.08 (90% of predicted value). As the patient was well with no evidence of peritonitis or visceral perforation, she was treated conservatively with tetracycline for stagnant loops and consequent bacterial overgrowth.

Fig 1 – Radiographs of the abdomen revealed large amount of free air under the diaphragm



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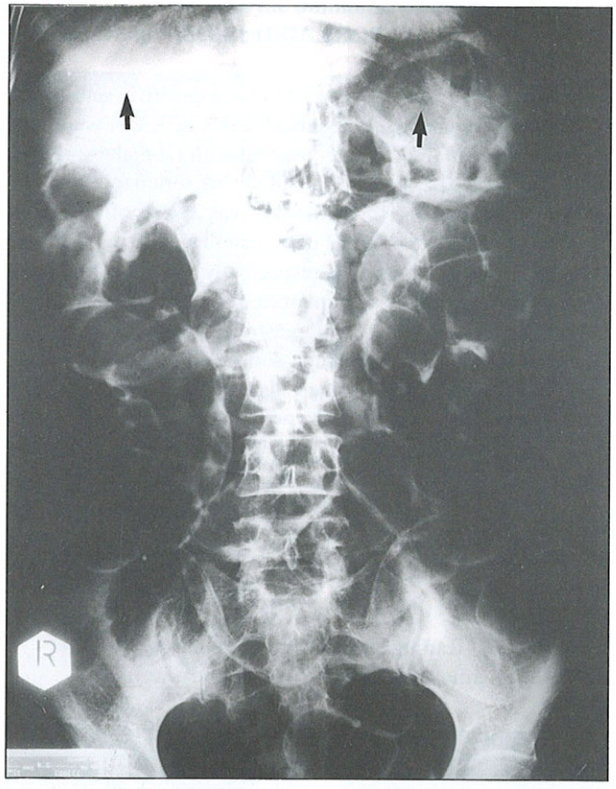
Her symptoms improved over the subsequent 2 weeks and she was allowed home. On review 2 months later, she remained well. Her abdominal distension was still present but much reduced. A repeat abdominal radiograph confirmed persistent pneumoperitoneum. At a further follow-up 6 months later, her symptoms of diarrhoea was noted to return, she had mild abdominal distension, a repeat abdominal radiograph showed complete resolution of pneumoperitoneum (Fig 2).

Fig 2 – Abdominal radiograph showing complete resolution of pneumoperitoneum



DISCUSSION

Gastrointestinal involvement is common in systemic sclerosis occurring in over 50% of patients[1]. Oesophageal hypomotility is very prominent occurring in three-fourths of patients[3]. Involvement of stomach, small or large intestine is less frequent, the major sign is intestinal dilatation. Large mouthed diverticula located along the anti-mesenteric border of the colon are characteristic[1].

Malabsorption is the commonest complication of intestinal systemic sclerosis, consequent upon hypomotility and bacterial overgrowth. The other complication is pneumatosis cystoides intestinalis which are air-filled cysts within the submucosa or the subserosa of the small or large intestine. The aetiology of these cysts is uncertain although a number of mechanism had been postulated[2]. These cysts occasionally rupture producing pneumoperitoneum; this was first reported by Fallen[4]. Since then a number of cases had been reported in association with various connective tissue disorder such as mixed connective tissue disease[5] and systemic lupus erythematosus[6].

Spontaneous pneumoperitoneum in the absence of detectable cause such as bowel perforation, ruptured diverticula in the small or large intestine or more rarely in association with pneumatosis cystoides intestinalis is very unusual. There have been only 4 other cases reported.

The pathogenesis of this phenomenon is uncertain. It has been suggested that recurrent microperforations in the colonic diverticula or elsewhere along the gastrointestinal tract caused air leakage from the bowel into the peritoneal cavity or that pneumatosis cystoides intestinalis was too small to be detected by conventional contrast studies[7]. Rupture of these cysts could give rise to persistent pneumoperitoneum.

An interesting feature demonstrated by this patient is the severity of gastrointestinal involvement manifested by pneumoperitoneum and malabsorption, whilst other organs remain relatively intact. She had extensive pneumoperitoneum for which no cause was found. Decision as to whether exploratory laparotomy is indicated can be difficult in some cases where patients are treated with steroid which may mask signs of peritonitis. In our cases, the pneumoperitoneum resolved with conservative management. Recognition of this condition is important as unnecessary surgical intervention can be avoided in this group of patients.

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