

Spontaneous pneumoperitoneum in the absence of visceral perforation in a patient with systemic sclerosis: a case report

Neumoperitoneo espontáneo en ausencia de perforación visceral en un paciente con esclerosis sistémica: reporte de caso

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Abstract

We present the case of a 44 year old woman with systemic sclerosis who presented with intense abdominal pain without signs of peritonitis. An abdominal computed tomography showed generalized intestinal dilation, intestinal pneumatosis and an extensive pneumoperitoneum. A diagnostic laparoscopy was performed but no perforation nor gastrointestinal leakage were found. Spontaneous pneumoperitoneum in patients with systemic sclerosis without visceral perforation is an extremely rare complication. Physicians must have a low threshold of suspicion for this entity when a patient with systemic sclerosis presents with spontaneous pneumoperitoneum in the absence of peritoneal signs.

Keywords: Pneumoperitoneum. Spontaneous pneumoperitoneum. Systemic sclerosis. Scleroderma.

Resumen

Presentamos el caso de una mujer de 44 años con diagnóstico de esclerosis sistémica, quien presentó dolor abdominal intenso sin datos de irritación peritoneal. Una tomografía computarizada de abdomen mostró dilatación generalizada de asas intestinales, neumatosis intestinal y neumoperitoneo extenso, por lo cual se realizó una laparoscopia diagnóstica, sin encontrar sitio de perforación. El neumoperitoneo espontáneo en pacientes con esclerodermia sin evidencia de perforación visceral es una complicación extremadamente rara. El médico deberá mantener un alto índice de sospecha para esta condición ante un paciente con esclerosis sistémica que se presente con un neumoperitoneo espontáneo sin datos de irritación peritoneal.

Palabras clave: Neumoperitoneo. Neumoperitoneo espontáneo. Esclerosis sistémica. Esclerodermia.

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Introduction

Pneumoperitoneum is defined as the presence of free extraluminal air in the peritoneal cavity¹, which in 90% of cases results from a perforation of a hollow viscus typically associated with signs of peritoneal irritation, requiring prompt surgical intervention². However, in the remaining 10% of cases, pneumoperitoneum may exist without perforation of a hollow viscus³. Some causes of pneumoperitoneum in the absence of perforation or peritonitis include ruptured hepatic abscesses, emphysematous cholecystitis, spontaneous bacterial peritonitis, pneumothorax with leakage into the abdominal cavity, and peritoneal dialysis⁴. In the context of systemic sclerosis, spontaneous pneumoperitoneum (in the absence of perforation) has been exceptionally described and is hypothesized to result from connective tissue changes in the gastrointestinal tract occurring in this disease. Recognizing spontaneous pneumoperitoneum in a patient with systemic sclerosis, in the absence of other signs (persistent pain, lack of resolution, signs of peritoneal irritation, abdominal compartment syndrome, etc.), can prevent unnecessary surgical interventions. We present the case of a 44-year-old woman with intestinal pneumatosis and spontaneous pneumoperitoneum in the absence of visceral perforation, due to systemic sclerosis.

Case report

A 44-year-old woman diagnosed with systemic lupus erythematosus 10 years prior to admission, on sulfasalazine, deflazacort, and methotrexate, and also diagnosed with scleroderma with CREST syndrome (calcinosis, Raynaud's phenomenon, esophageal dysmotility, sclerodactyly, telangiectasias) under treatment with baricitinib. Her current condition started 6 months prior to admission with colicky, intermittent, generalized abdominal pain of 6/10 intensity. Physical examination revealed the presence of a soft, depressible abdomen, tender on medium and deep palpation in the mesogastrium, without signs of peritoneal irritation. An abdominal X-ray suggested intestinal perforation, and the patient was referred to the ER, where a contrast-enhanced abdominal CT scan (Fig. 1) showed a partially distended stomach, generalized dilation of intestinal loops, intestinal pneumatosis mainly in the jejunum and ileum, and abundant free intra-abdominal air consistent with extensive pneumoperitoneum,

without a clear site of leakage for the oral contrast medium. Lab test results were within normal parameters, and the patient was admitted for observation. During hospitalization, she presented with increased abdominal distension and pain progression, leading to an emergency diagnostic laparoscopy. Findings included intra-abdominal hypertension (opening intra-abdominal pressure of 12 mmHg), pneumoperitoneum, intestinal, diaphragmatic, and mesenteric pneumatosis, generalized serositis, and free intra-abdominal fluid (Fig.2). An intraoperative panendoscopy showed no perforation site or anatomical abnormalities, reporting only erosive gastritis without any other significant findings. Therefore, a complete intestinal count with cavity lavage and drainage was performed. No perforation or GI leak was ever found, so continued observation and medical management in hospitalization with prokinetics, parenteral antibiotics, and bowel rest was decided. Postoperative abdominal CT scan performed at 24 hours revealed the presence of a significant decrease in pneumoperitoneum and intestinal pneumatosis (Fig.3), leading to hospital discharge for outpatient management.

Discussion

Systemic sclerosis or scleroderma is a rheumatologic connective tissue disease characterized by fibrosis of certain organs and systems⁵. Classification of this disease is based on dermal involvement: limited systemic sclerosis (formerly called CREST syndrome) is characterized by skin thickening distal to the elbows and knees without trunk involvement, while diffuse systemic sclerosis involves proximal limb segments, face, and trunk; both forms are associated with systemic manifestations. In 70% of cases, there is positivity for anticentromere antibodies, anti-Scl 70, or anti-RNA polymerase 3. The most widely affected organs in systemic sclerosis are the skin, the GI tract, lungs, pericardium, kidneys, and musculoskeletal system⁶.

The classic GI changes in systemic sclerosis include atrophy of the muscularis propria layer and its replacement by collagen-based connective tissue, leading to decreased peristaltic activity and stasis¹. Additionally, esophageal dysmotility, delayed gastric emptying, jejunal diverticulosis, malabsorption, and intestinal pneumatosis may occur⁷. Intestinal pneumatosis, present in only 8% of cases, is a rare intestinal disease characterized by the presence of gas within the intestinal wall, generally in the mucosa and submucosa, with exceptionally rare extraintestinal locations⁸. The passage of

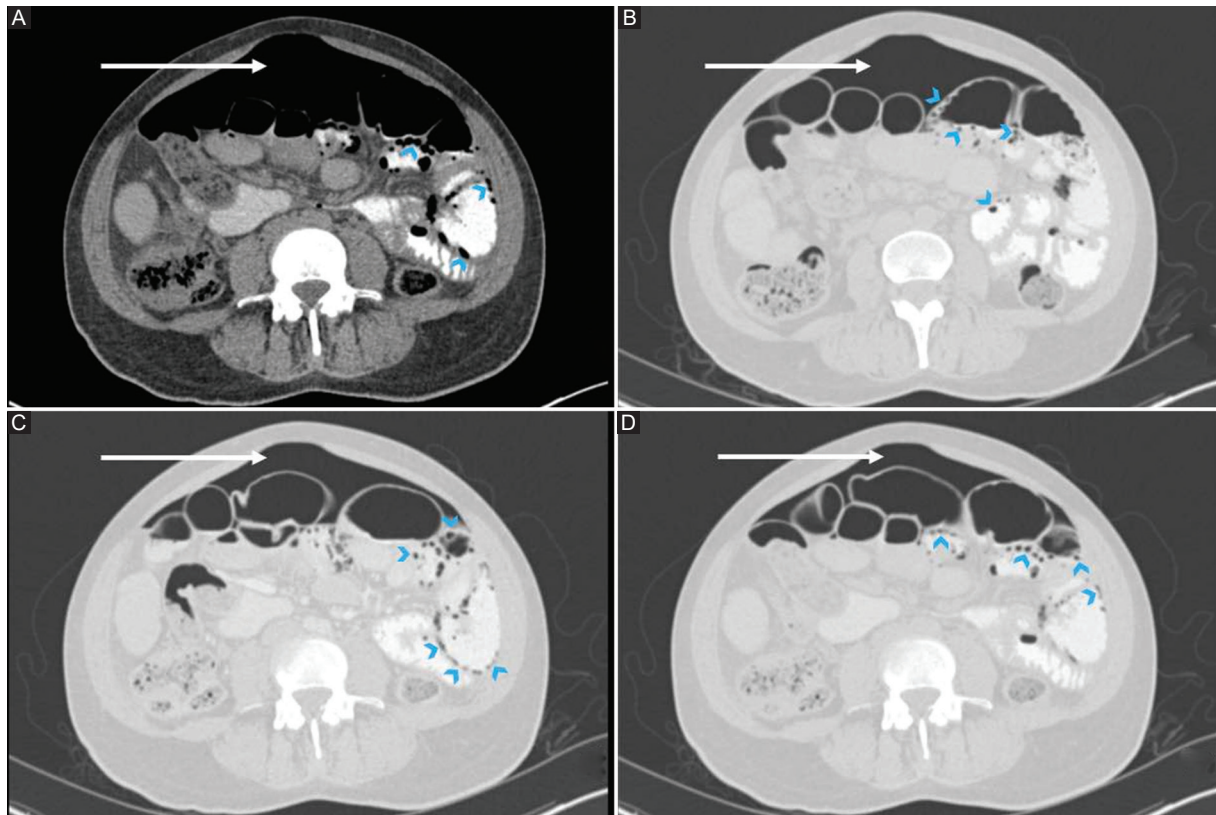


Figure 1. Double-contrast CT scan of the abdomen. **A:** abdominal window. Pneumoperitoneum (white arrow) and intestinal pneumatosis (blue arrowheads). **B, C, and D:** lung window for better visualization. Pneumoperitoneum (white arrows). Pneumatosis in the intestinal loop walls (blue arrowheads) and generalized dilation of intestinal loops. No site of contrast medium leakage.

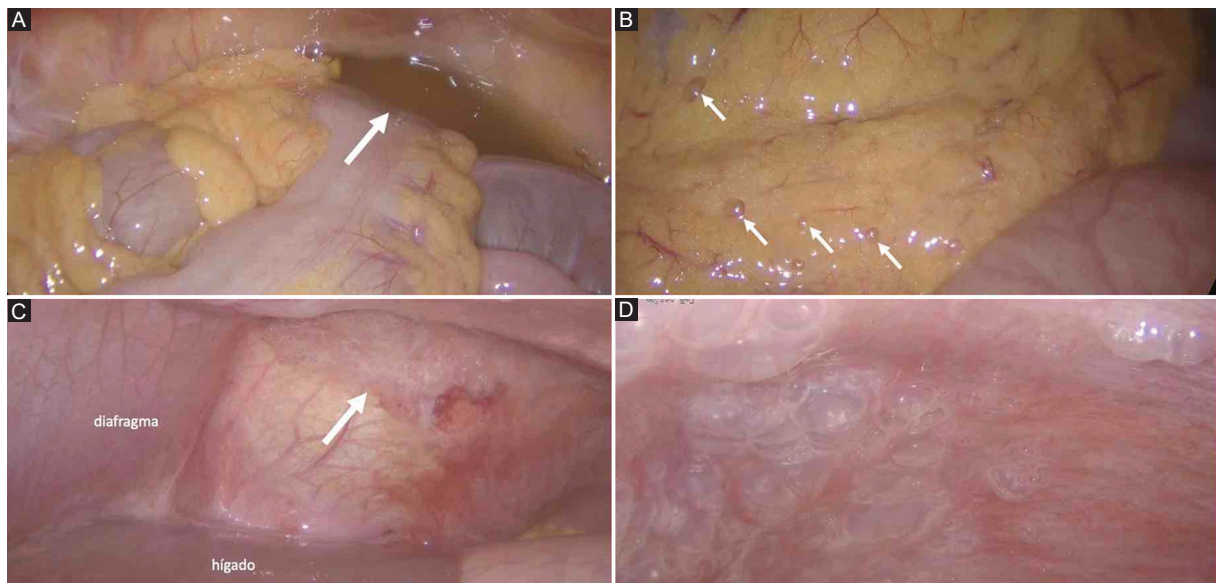


Figure 2. Laparoscopic examination of the abdominal cavity. **A:** free fluid in the abdominal cavity and dilated but normal-appearing intestine. **B:** mesenteric pneumatosis (white arrows). **C:** diaphragmatic pneumatosis (white arrow). **D:** close-up of diaphragmatic pneumatosis.

intraluminal gas to the submucosa requires damage to the mucosal muscular layer, which can occur due to increased intraluminal pressure, inflammatory or

autoimmune processes (such as systemic sclerosis), *Clostridioides difficile* infection, chronic obstructive pulmonary disease, cystic fibrosis, or immunosuppressive

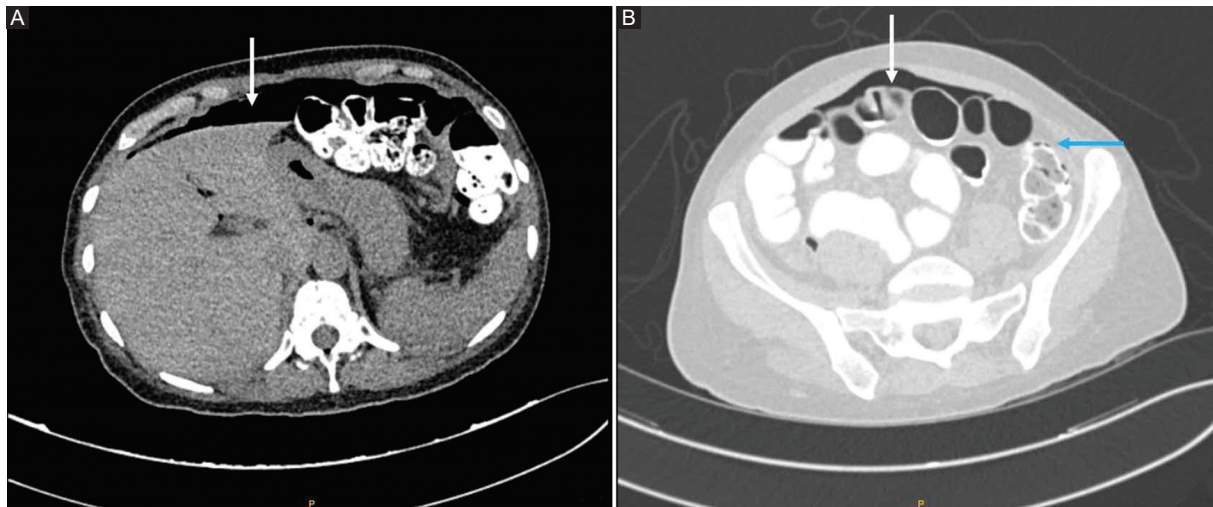


Figure 3. Simple CT scan of the abdomen, one day after surgery. Partial resolution of pneumoperitoneum (white arrow) and intestinal pneumatosis (blue arrow). **A:** abdominal window. **B:** lung window for better visualization.

agents^{9,10}. Corticosteroids and other immunosuppressive agents have been hypothesized as causative factors because they induce intestinal mucosal atrophy, depletion of intestinal lymphoid tissue, and loss of mucosal barrier integrity, allowing intraluminal air dissection through the submucosa and subserosa¹¹. In the context of systemic sclerosis, intestinal pneumatosis appears to be a late complication with a worse 6-month prognosis vs patients without this condition¹². In our patient, various factors, such as systemic sclerosis and the use of corticosteroids and immunosuppressive agents, could have predisposed to extensive intestinal pneumatosis for the described reasons. Another condition related to systemic sclerosis and systemic lupus erythematosus is cystic intestinal pneumatosis, characterized by cysts within the intestinal wall that sometimes associate with pneumoperitoneum when these cysts rupture^{13,14}; however, in cystic intestinal pneumatosis, the cysts are visible macroscopically or by imaging studies^{15,16}.

Spontaneous pneumoperitoneum in patients with scleroderma without evidence of visceral perforation or cystic intestinal pneumatosis is an extremely rare complication of systemic sclerosis. As far as the authors know, it has only been reported six times in the global literature, with no cases published in Mexico^{7,17-19}. In this scenario, it has been postulated that in the absence of clear visceral perforation signs such as free fluid, extravasation of enteral contrast, or peritonitis, the pneumoperitoneum originates from intramural gas collections passing through microscopic defects in a weakened mucosa, which, with increased

intraluminal pressure from any cause (stasis, decreased motility, etc.), reaches the peritoneal cavity, thus forming the pneumoperitoneum²⁰. The clinical presentation in these cases is variable, ranging from asymptomatic to severe abdominal pain, prominent distension, hematochezia, or others^{14,21}. The reference method for diagnosis is a double-contrast CT scan of the abdomen, usually showing findings such as interloop edema, thickening of the intestinal wall with contrast enhancement, and intramural gas^{14,22}. A characteristic and unique aspect of the presented clinical case is that the findings included not only intestinal pneumatosis and overt pneumoperitoneum but also mesenteric pneumatosis and pneumatosis of the omentum and parietal peritoneum at the diaphragm level during diagnostic laparoscopy (Fig. 2). A probable explanation for the findings of air in the parietal peritoneum and mesentery could be autoimmune serositis developing in the context of systemic lupus erythematosus^{9,23}. Despite the lack of an established consensus, most authors recommend that in a patient with pneumoperitoneum and systemic sclerosis, in the absence of alarming signs and peritoneal irritation, expectant management with observation and medical treatment should be established. This includes bowel rest, parenteral nutrition, oxygen therapy, and antibiotics¹⁷. Parenteral nutrition reduces the availability of nutrients in the intestine, thereby reducing gas production by colonic flora, while antibiotics and oxygen therapy theoretically reduce the number of gas-producing anaerobic bacteria²⁴. Any clinical deterioration or suspicion of perforation warrants prompt surgical exploration, while diagnostic uncertainty

prefers laparoscopic exploration due to the risk of missing life-threatening surgical conditions^{3,7}.

Conclusions

Awareness of various conditions and their presentations, though infrequent, allows surgeons to maintain a high index of suspicion in patients diagnosed with spontaneous pneumoperitoneum without intestinal perforation in the context of systemic sclerosis or other rheumatologic and immunological diseases. This enables appropriate therapeutic management even when the case appears surgical based on imaging studies. Understanding this condition and its management will avoid unnecessary procedures, such as intestinal resections, which could compromise intestinal absorption function and the patient's life.

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Conflicts of interest

None declared.

Ethical disclosures

Protection of human and animal subjects. The authors declare that no experiments were performed on humans or animals for this study.

Right to privacy and informed consent. The authors have obtained approval from the Ethics Committee for analysis and publication of routinely acquired clinical data and informed consent was not required for this retrospective observational study.

Right to privacy and informed consent. The authors have obtained the written informed consent of the patients or subjects mentioned in the article. The corresponding author is in possession of this document.

Use of artificial intelligence for generating text. The authors declare that they have not used any type

of generative artificial intelligence for the writing of this manuscript, nor for the creation of images, graphics, tables, or their corresponding captions.

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