Things are not always what they seem

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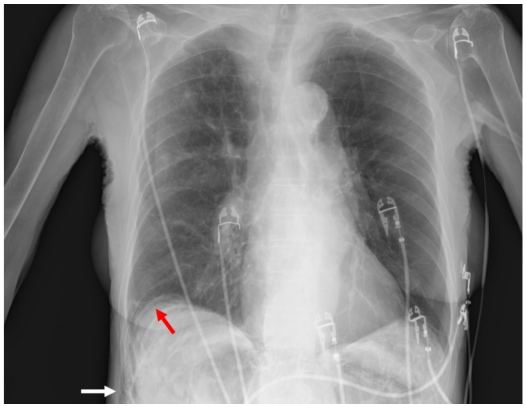

Figure 1. Chest X-ray.

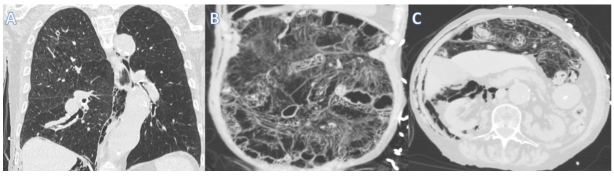

Figure 2. Coronal CT scan of the chest showing severe pulmonary emphysema and large amount of free air in the mediastinum (**A**). Coronal (**B**) and axial (**C**) CT scan of the abdomen showing abundant and widespread accumulation of air in the intestinal walls and large amount of free air in the abdomen.

A 80-year-old woman, heavy ex-smoker (60 pack-years) affected by chronic obstructive pulmonary disease (COPD), presented to the emergency department with worsening dyspnea in the previous three hours, metabolic alkalosis (pH 7.55, pO2 33 mmHg, pCO2 34 mmHg, HCO3 30 mmol/L, lactates 3.3 mmol/L) and persistent epigastric pain for one week. She was under treatment with low-dose aspirin, metoprolol, atorvastatin, pantoprazole, ramipril, salmeterol and fluticasone propionate, tiotropium bromide, allopurinol, benserazide, diazepam, furosemide. On physical examination, she presented with tachypnea (30 beats/min) and reduced peripheral oxygen saturation on room air (83%); her remaining vital signs were normal and there were no signs of peritonism. Blood tests revealed an increase of white blood cell (11.07 x 10^9/l) and neutrophil (9.73 x 10^9/l) count, glycemia (217 mg/dl), C-reactive protein (2.49 mg/dl; normal values < 0.5 mg/dl) and B-type natriuretic peptide (259 pg/mL; normal values <100 pg/mL) levels. Estimated glomerular filtration rate was moderately to severely decreased (36 mL/min/1.73 m^2). Remaining blood exams (including hepatic function and coagulation time) were normal, in particular lactate dehydrogenase levels (200 U/L; normal values < 248 U/L). Chest X-ray showed a small amount of abdominal free air below the right hemidiaphragm (Figure 1, red arrow) and subcutaneous emphysema in the right lateral abdominal wall (Figure 1, white arrow). A subsequent thoraco-abdominal comupted tomography (CT) scan documented severe pulmonary emphysema (Figure 2 A), abundant and widespread accumulation of air in the intestinal walls (Figure 2 B and C), large amount of free air in the mediastinum (Figure 2 A) and abdomen (Figure 2 B and C).

Question

Given the patient's history and CT scan result, which is the most likely diagnosis?

1. Acute bowel ischemia with intestinal perforation and pneumomediastinum
2. Bowel perforation with pneumomediastinum
3. Pneumatosis cystoides intestinalis with benign pneumoperitoneum and pneumomediastinum
4. Bowel inflammation with intestinal perforation and pneumomediastinum

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Informed consent: the patient provided consent for the access to medical records at the time of admission.

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Answer

Pneumatosis cystoides intestinalis with benign pneumoperitoneum and pneumomediastinum is the correct answer. After two weeks of conservative management (ceftriaxone 2 g iv daily and metronidazole 500 mg Q8H for 7 days, intestinal rest with total parenteral nutrition and oxygen), a CT scan documented a complete resolution of abdominal findings and pneumomediastinum. The patient was discharged after 16 days of recovery in good clinical condition. Pneumatosis cystoides intestinalis is characterized by the presence of multiple gas-filled cysts within the submucosa or subserosa of the gastrointestinal tract.[1,2] It is a rare disease affecting approximately 0.03% of the adult population.[3,4] It can be idiopathic (15%) or secondary (85%) to various predisposing factors, including pulmonary conditions (COPD, asthma), gastrointestinal pathologies (inflammatory bowel diseases, intestinal occlusion), scleroderma, mechanic ventilation and drugs (steroids, immunosuppressants, chemotherapy agents).[5] Patients with COPD may experience increased intraalveolar pressure (for example caused by coughing), which can led to alveolar rupture, air dissection within the mediastinum and the abdomen and air trapping in the bowel wall to form cysts.[1,6] In the absence of signs of any life-threatening conditions (such as peritonitis, sepsis, acidosis and portal venous gas on CT scan images), conservative management (antibiotics, intestinal rest and oxygen) is warranted because in these cases pneumatosis cystoides intestinalis is a self-limited process.[3,7,8]

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