Lymphoproliferative disease: an unusual complication

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Abstract

The authors present a case of Waldenstrom disease, diagnosed during the study of a hemorrhagic disorder. The appearance of a pneumoperitoneum led to the diagnosis of a second disease, rarely found in a medical ward.

Key words: hemorrhagic diathesis, pneumoperitoneum, pneumatosis.

Introduction

Pneumatosis intestinalis is a rare entity characterized by the presence of multiple air-filled cysts dispersed within the bowel and/or mesenteric wall.[1,2,3]

It was first described in 1730 by du Vernoi, who found these cysts in a corpse.[1]

The absence of characteristic symptoms, sometimes mimicking conditions of acute abdomen and gastrointestinal bleeding, with radiological aspects of pneumoperitoneum, may lead to unnecessary surgical interventions.[1,2,4]

We present a case of Waldenström's disease in which the finding of a retropneumoperitoneum without any gastrointestinal symptoms alerted us to the possibility that this might be the case of pneumatosis intestinalis.

Case report

The authors present the case of a 78 year old male, Caucasian patient, born and living in Amadora, married and retired (a former navy blacksmith). The patient was admitted to our hospital department to clarify a bleeding diathesis.

About eight months before, the patient had experienced the onset of asthenia, weakness, non-selective anorexia and weight loss of about 10 kg (within one month). Four months later, several episodes of abundant, self-limited bilateral epistaxis occurred, which were not triggered by effort. One month later, the constitutional symptoms worsened and painful bleeding ulcers of about 1 cm in diameter appeared in the buccal mucosa, which gradually increased in size. The patient visited his doctor, who prescribed topical therapy, the name of which the patient did not know. One day before going to the Emergency unit, ecchymosis and petechiae appeared spontaneously on the face and limbs.

It should be noted that the patient denied any other symptoms, including blood loss from other locations, fever, jaundice or sclerotic skin, edemas, or digestive, cardiorespiratory or neurological complaints.

In relation to the patient's personal history, an episode of upper gastrointestinal bleeding (two years before), diagnosed as hiatus hernia, sporadic ingestion of indomethacin for mechanical lower back pain are emphasized. The patient is not a heavy drinker.

On admission, the patient was lucid, oriented in time and space, with a reasonable overall health and nutrition condition, pale skin and mucosa, anicteric, eupnoeic, acyanotic and afebrile. No visible blood loss was observed and enlarged lymph nodes were not palpable in the various ga nglionary chains. The patient had extensive ecchymosis of the cheeks and peri-buccal region, and petechiae throughout the limb surfaces. The right buccal mucosa had a 3-cm diameter ulcer with regular edges and was pink and bleeding. BP 134/76 mmHg (right arm, sitting), pulse 70 ppm (rra), respiratory rate 16 cycles/min. Cardio-

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pulmonary auscultation was normal. The abdomen was painless, without hepatosplenomegaly or abnormal masses; no audible vascular murmurs. Spinal and genito-proctologic examination was normal. The limbs had no deformities or edema, arterial pulses were palpable on the points of election, rhythmic, regular, wide and symmetrical, the venous routes were clear. Neurological examination showed no change, and fundoscopy revealed signs of angiosclerosis.

To clarify the condition, the patient underwent several diagnostic exams:

Blood count: Blood red cells 1 800 000/mm3; Hg 6.0/dL, Ht 18.7%, MCV 99.8 m3; MCHC 32.1/dL; MCH 32.1/dL; reticulocytes 0.3%; platelets 9000/mm3; WBC 3700/mm3 (N 42.6%, L 54.9%, M 2.5%, E 0.7%, B 0.2%); PT 87%; PTT 24 s; urea 46 mg/dL; creatinine 1.1 mg/dL; glucose 109 mg/dL; total and direct bilirubin 1.1/0.2 mg/dL; SGOT/SGPT 34/7 mg/dL; Na 137 mg/dL; K 3.8 mg/dL; Ca 9.2 mg/dL; uric acid 8.3 mg/dL; total protein 7.8 g/dL; electrophoresis: albumin 53.8%, a1 3.9%; a2 8.9 %; b 8.3%; g 25.1%).

Protein immunoelectrophoresis with determination of immunoglobulins: IgG 1770mg/dL (normal from 82 to 1700); IgA 146mg/dL (normal from 100 to 490); IgM 3834 (normal from 50 to 320); Ig IK 540 mg/dL (normal from 200 to 400); IgL l 197 IgM monoclonal gammopathy.

ECG, echocardiogram and abdominal ultrasound showed no changes; skeletal X-ray revealed diffuse osteopenia, and signs of severe osteoarthritis of the lumbosacral spine. Chest X-ray revealed no changes. Myelogram - very abundant marrow, increased cellularity, decreased fat; 0% granulocytes without megakaryocytes, 7% of mast cells, 27% of mature lymphocytes, 21% of lymphoid cells with more loose nuclear chromatin and visible nucleoli. Bone biopsy - hypercellular bone marrow with the medullar space replaced with a diffuse infiltrate of cells with morphology of small lymphocytes. Decreased haematopoiesis (<10%).

Given the clinical symptoms and additional diagnostic exams, we concluded this was a case of Waldenström's disease. The patient started therapy with Prednisolone 1 mg/kg/day and Chlorambucil 6 mg/day. There was a progressive improvement of the bleeding diathesis, with a natural improvement in platelet count.

On day 16 of the corticosteroid therapy and day 12 of the chlorambucil therapy, CAT scan of the chest and abdomen revealed an extensive pneumoperitoneum and retropneumoperitoneum. At that time, the patient underwent a simple X-ray of the abdomen, which confirmed the CAT scan images and also revealed multiple air-filled cystic formations dispersed throughout the walls of the colon, as well as Chilaiditi's sign. Opaque enema reaffirmed these data.

Given that the patient was completely asymptomatic and the abdomen was totally painless (only showing generalized tympanism), we concluded that this was a case of pneumatosis intestinalis. After surgery, the patient remained on absolute bed rest and was treated with Metronidazole (500 mg po 8/8h). New imaging tests were carried out periodically. A gradual reabsorption of the intra-abdominal air and the disappearance of the cysts were observed within about a month. There were no further similar episodes.

Comments

The presence of multiple air-filled cysts on the mucosa and/or subserous membranes of the bowel and/or mesenteric wall, possibly involving the entire digestive tract, is the characteristic feature of pneumatosis intestinalis.[1,2] This entity has three different forms:[1]

• Primary, in which no underlying gastrointestinal pathology is present, and only the submucous membranes are involved; this form is usually segmental and occurs predominantly within the left hemicolon

• Secondary, in which there are predisposing pathological processes, inflammatory bowel, peritoneum or appendix diseases, obstructive conditions of the digestive tract, infectious intestinal diseases (such as tuberculosis, parasitic infections, barotrauma), iatrogenic illness (endoscopy), intra-abdominal ischemia or other pathologies such as cystic fibrosis, lymphosarcomas, peritoneal carcinomatosis, treatment with steroids or chemotherapy, scleroderma and other connective tissue diseases,[3] heart failure and COPD. This type affects particularly the subserous membranes in a continuous or segmental manner, involving mostly the small intestine, right colon or stomach;

• Pediatric, with aspects of necrotizing colitis in children,[4] usually with fulminating evolution.

The cause of this pathology is unknown.[1,4] There are several theories regarding its pathogenesis:[1] mechanical (increased intra-abdominal pressure), bacterial (overproliferation of the anaerobic intestinal flora), pulmonary (alveolar gas breaks through the mediastinum) or diet-related (non-characterized

absence of enzymes that would lead to over-fermentation).

The diagnosis of this rare entity is based fundamentally on X-ray findings,[1] as it will be addressed later. The symptoms may be totally absent or identical to those of irritable colon.[1,5] In the case of lesions of the colon, diarrhoea (56%) and/or hematochezia (50%) may be also occur; in the lesions of the small intestine, complaints of vomiting (59%), abdominal distension (59%), weight loss (55%) or nonspecific abdominal pain (53%)[1] may be reported. However, there are no typical clinical symptoms exist for this entity.

On objective examination, tympanism can be observed, which is usually predominant in the upper quadrants of the abdomen, and sometimes small intra-abdominal crackling masses can be felt on palpation.[1]

In the imaging tests, which are essential for the diagnosis,[1] characteristic aspects can be observed:[2]
1) in a simple X-ray of the abdomen (diagnosis in 2/3 of cases), air-filled cysts and/or pneumo- or retropneumoperitoneum can be observed, as well as compartmentalized collections of air, which may be more, or less extensive. Chilaiditi's sign may also occur (in 15% of cases), consisting of the interposition of an intestinal loop between the liver and hemidiaphragm.
2) In opaque enema, well-defined filling defects or polypoid lesions of the intestinal walls of various sizes are evident, which do not alter the mucosal folds; air-filled contours may also be observed around the abdominal walls.
3) In gastrointestinal endoscopy, transparent masses that are depressible to the touch can be observed; these are not seen in the biopsy.
4) CAT scan, with its high resolution, confirms the aspects revealed by other imaging tests; however, it is not essential for the diagnosis.

Although this disease usually has different clinical presentations, it is important to make a differential diagnosis with other pathologies that can result in similar x-ray findings,[1] such as emphysematous gastritis, acute enteritis, lymphangioma of the peritoneum, sclerosing lipogranulomatosis, Whipple's disease or cystic colitis.

The prognosis of pneumatosis intestinalis is unpredictable,[1,4,5] and chronic cases with periods of remission/exacerbation have been reported; spontaneous cure was seen in about 29% of cases. In the secondary form, the prognosis depends essentially on the underlying disease.[6]

The complications (3%)[1] reported include cases of intestinal obstruction, perforation and gastrointestinal hemorrhage, sometimes requiring surgery.

The medical therapy[1,6] consists of the use of antibiotics (ampicillin or metronidazole) and rest. Some authors advocate the use of oxygen therapy at high concentrations, but the results are not conclusive.

In clinical case presented here, the causes that triggered the disease may have been either the underlying neoplastic process - Waldenström's disease, or hiatus hernia, or the therapy given (Prednisolone and Chlorambucil). With the administration of antibiotics and rest, complete regression of the aspects of pneumatosis was observed, and the patient had suffered no relapse after about one and half years of follow-up. ■

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