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Pneumatosis intestini and pneumoperitoneum in acute lymphoblastic leukaemia

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Pneumatosis intestini and pneumoperitoneum can originate after infection or gastrointestinal obstruction. Very rarely this condition develops as a random event in patients with collagen disorders and vasculitis, acute and chronic graft versus host disease and various immunodeficiency with no clinical impact.

A patient with acute lymphoblastic leukaemia who developed pneumatosis intestini and pneumoperitoneum with no clinical evidence is reported on. According to the clinical data we assume that the etiology of this condition in the patient is multifactorial: leukaemia, chemotherapy, corticosteroids, sepsis and/or fungal infection.

Correlation between patient clinical status and radiological evidence of pneumatosis peritonei is crucial to obtaining the right picture of clinical situation.

Key words: leukaemia, pneumoperitoneum; pneumatosis intestinalis

Introduction

Pneumatosis intestini is a common radiological finding in patients with intestinal necrosis. Intestinal perforation is closely accompained with "surgical" pneumoperitoneum and peritonitis. This serious condition has typical clinical finding of acute abdominal pain which leads to urgent abdominal operation.

In contrast to this situation "benign" pneumatosis[1] or "internistical" pneumoperitoneum[2] usually has no clinical signs and can be diagnosed only by plain abdominal film. This "benign" pneumatosis is caused by other extraintestinal diseases or conditions. Such patients are treated by conservative therapy and very rarely by surgical operation.[3,4]

The main purpose of this case report is to point out specific radiologic signs of "benign" pneumatosis and pneumoperitoneum.

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Case report

A forty-seven-year-old male was admitted to the hospital because of weakness, high number of WBC, anemia and a low number of platelets. Bone marrow aspiration showed immature cells

compatible with acute lymphoblastic leukaemia.

Chemotherapy by daunorubicine, cyclophosphamide, vincristine and 6-methylprednisolone was introduced (day +1). Ten days later, mucositis grade IV was observed. At the same time the patient became febrile and blood culture revealed infection with Streptococcus viridans. Antibiotics regimen, consisting of netylmicin and penicillin, was started. At the same time chest X-ray showed multiple infiltrates in both upper lung fields compatible with fungal infection. Amphotericin B and 5-flucitosine was introduced. Unfortunately, bone marrow aspiration on day +23 showed 90% of blast, and salvage chemotherapy of intermediate doses consisting of arabinoside and m-amsacrine over 6 days was started.

During all this period patient did not have signs of abdominal pain or surgical abdomen.

On day +39 routine chest X-ray was performed because of previously diagnosed multiple lung infiltrates. Examination of this X-ray showed regression of lung infiltrates but with air shaped like a sickle under both sides of diaphragm. Patient was subfebrile, 37.5°C, but in a good condition without any pain or other symptoms. Karnofsky score was 70%. Clinically we found gentle abdomen with enlargement of liver (3 cm bellow right costal margin). There was an evidence of intestine movement.

Plain-films of abdomen showed evidence of pneumoperitoneum, with air in wall of small and large intestine, especially in transverse colon and mesocolon (Figure 1 and 2). Air collections, were oval or rounded but in some parts irregular shaped. This air mass parallely followed intestinal contour. In mesocolon, it was radially placed but parallely followed the mesenterium vessels. There was no evidence of significantly distinct intestine meteorism.

Patient position had no influence on the above mentioned radiologic finding.

Radiologic evidence of pneumoperitoneum and pneumatosis intestini slowly decreased over 11 days without any clinical impairment.

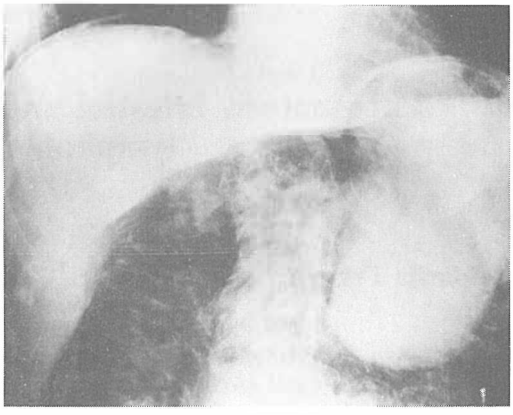

Figure 1. Pneumoperitoneum: gas shaped like a sickle show subdiaphragmatic and subhepatic.

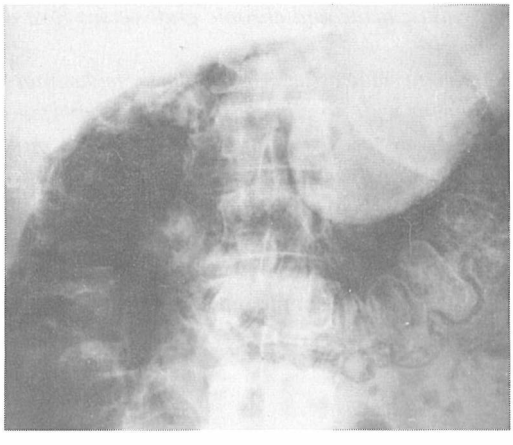

Figure 2. Pneumatosis intestinalis: gas in the wall of the small and large bowel, especially in transverse colon and mesocolon.

On day +50 there was no evidence of air in abdominal cavity. During all this time the patient was maitained on fluid replacement, intestinal decontamination, antibiotics, antifungal and sterile diet. Bone marrow aspiration on day +65 showed presence of blasts. Because of that patient received high doses of cytosine arabinosie over 6 days.

This period was complicated with heart failure and patient died on day +75 after starting of initial chemotherapy course.

Discussion

Intestinal pneumatosis is collection of air in intestinal wall especially in submucosa and subserosa.

"Benign intestinal pneumatosis" was first described in 1973.[1]

It is a common finding in newborns with severe necrotic enterocolitis.[4–10] There were reports on the same condition in patients with leukaemia and lymphoma,[11–14] with collagen disorders,[15, 16] with severe respiratory distress syndrome,[2, 17, 18] in acquired and hereditary immunodeficiency disorders,[7, 12, 19–22] and after immunosupressive therapy.[19, 21]

Etiologycal explonation of intestinal pneumatosis is multifactorial.[5, 23, 24]

Conditions such are disease (leukaemia, lymphoma, malignant diseases, immunodeficiency and collagen disorders) chemo-radio therapy, administration of corticosteroides, bacterial, viral or fungal infection could cause mucosal damage and intestine wall necrosis.

Severe anaemia in patients with malignant disease might cause ischaemic intestine wall necrosis.[5, 14]

Administrations of corticosteroides, especially in infancy, might develop atrophia of lymphoid tissue in small and large intestine which leads to submucosal or subserous air disection.[12, 21]

A patient receiving high doses of chemoradiotherapy, as a condition regimen prior to marrow transplatation, may experience this condition due to the toxic effect on gastrointestinal tract. Although, gastrointestinal tract is a target tissue for acute and chronic graft versus host disease.

According to Keats[13] and Yeager[20] mucosal damage appeares 2–3 weeks after finishing chemotherapy.

One of the major factor in developing such condition is presence of bacteria, especially anaerobes, in the bowel.[5, 6, 23]

In some patients with intestinal pneumatosis infections with rotaviruses and adenoviruses were documented.[25]

Viral enteritis in patients receiving marrow graft is a very rare and there are no available literature data on this condition.

Clinical features of pneumatosis intestini usually consist of abdominal pain.

In some cases, so called "silent" or "indolent" cases pneumatosis intestini and pneumoperitoneum is a random finding on routine chest X-ray.[11, 12, 24]

The main site of this air mass is terminal ileum, large colon, especially colon transversum and ascendent,[8, 13, 20] rarely in aboral part of colon.[26]

In our patient chemotherapy was started 39 days before routine chest X-ray were performed.

The patient experienced no abdominal symptomatology. Plain abdominal film showed air shaped like a sickle under both sides of the diaphragm in diameter of 1 cm. Pneumatosis was extended to the terminal ileum, mesocolon and large intestine up to the rectosigmoidal border.

Finding with such extension as in our patient, without clinical impairment, is very rare.[12, 19, 26]

Usually, evidence of air in abdominal cavity disappears within two weeks.[13]

In our patient 11 days after first X-ray there was no evidence of air collection in the wall of small and large intestine and mesocolon.

The characteristics of "benign" asymptomatic pneumatosis are oval or rounded air cystic collections in the serosa and subserosa. This distinguishes "benign" pneumatosis from symptomatic pneumatosis in which air collection is linear and narrowly located in the submucosa.[3, 27]

"Benign" pneumatosis is also easily distinguished from pneumatosis cystoides intestinalis in which air collections accumulate in the large oval cysts like spaces along the intestine wall.

Moreover, in "benign" pneumatosis patient's position (standing vs lying) and time (24 hour period between X-rays) have no influence on the position of air in abdominal cavity.[8]

In our opinion, such differentiation of the intestinal pneumatosis, despite the fact that it emphasises the importance of radiological findings obtained by standard examinations, which

is the aim of this paper, is not accurate unless clinical symptomatology of a patient is not build in the diagnosis as a whole.

Similar attitudes are also presented by Thomas VI.[28]

Some recent studies report usefulness of ultrasound and computerized tomography in determining location and progression of intestinal pneumatosis,[9, 27–31], but the main diagnostic method is still plain abdominal film.

In conclusion, we agree with studies of Keats[13] and Yeager[20] who emphasize that a well informed radiologist and carefully examined series of plain abdominal films may give a very useful information on the development of pneumatosis intestini and "benign" pneumoperitoneum.

Correlation between patient's clinical status and radiological evidence of pneumatosis intestini and pneumoperitoneum is crucial in obtaining the right diagnosis.

References

1. Mc Sweeny WJ and Stempel DA. Noniatrogenic pneumomediastinum in infancy and childhood. *Pediatr Radiol* 1973; **1**: 139–44.
2. Zerella JT, Mc Cullough JY. Pneumoperitoneum in infants without gastrointestinal perforation. *Surgery* 1981; **89**: 163–7.
3. Knechtle SM, Davidoff AM, Rice RP. Pneumatosis intestinalis: Surgical menagement and clinical outcome. *Ann Surg* 1990; **212**: 160–5.
4. Sibbald WJ, Sweeney JP, Inwood MJ. Portal venous gas (PVG) as an indication for heparinization. *Am J Surg* 1972; **124**: 690–3.
5. Sidney WN. Ekstraluminal gas colections due to diseases of the gastrointestinal tract. *AJR* 1972; **115**: 225–48.
6. Chabot VH, Slovis ThL and Cullen M. Recurrent pneumatosis intestinalis in young infants. *Pediatr Radiol* 1992; **22**: 120–2.
7. Sivit CJ, Josephs ShH, Taylor GA and Kushner DC. Pneumatosis intestinalis in children with AIDS. *AJR* 1990; **155**: 133–4.
8. Rosenquist CJ. An unusual pattern of intramural gas in small bowel infarction. *Radiology* 1971; **99**: 337–8.
9. Vijayaraghavan SB. Sonographic features of pneumatosis of the small bowel. *J Clin Ultrasound* 1990; **18**: 579–81.
10. Dudgeon DL, Coran AG, Lauppe FA, Hodgman JE and Rosenkrantz JG. Surgical management of acute necrotizing enterocolitis in infancy. *J Pediatr Surg* 1973; **8**: 607–14.
11. Galal O, Osse G, Weigel W, Gahr M. Pneumatosis intestinalis in children with leukaemia: report of three cases. *Eur J Pediatr* 1981; **137**: 91–3.
12. Borns PF, Johnston TA. Indolent pneumatosis of the bowel wall associated with immune suppressive therapy. *Ann Radiol* 1973; **16**: 163–5.
13. Keats TE, Smith TH. Benign pneumatosis intestinalis in childhood leukaemia. *AJR* 1974; **122**: 150–2.
14. O'Connell DJ and Thompson AJ. Pneumatosis coli in non-Hodgkins lymphoma. *BJR* 1978; **51**: 203-5.
15. Mueller ChF, Morehead R, Michener W. Pneumatosis intestinalis in collagen disorders. *AJR* 1972; **115**: 300–5.
16. Wang CL, Wang F, Wong KC, Jeyamalar R. Benign persistent pneumoperitoneum in systemic sclerosis. *Singapore* MJ 1993; **34** (6): 563–4.
17. Kiryu T, Kobayashi H, Kawaguchi S, Kanou S, Uwabe Y, Sakai M, Matsuoka T, Nagata N. A case of status asthmaticus complicated by with pneumoperitoneum during mechanical ventilation therapy. *Japanese Journal of Thoracic Diseases*. 1993; **31** (6): 771–4.
18. Kandylakis S, Makrigiannakis A, Mirra N, Perpyrakis G, Kontakis K, Apalakis A. Pneumoperitoneum without perforation. *Minerva Chirurgica* 1993; **48** (12): 717–20.
19. King S, Shuckett B. Sonographic diagnosis of portal venous gas in two pediatric liver transpalant patients with benign pneumatosis intestinalis. *Pediatr Radiol* 1992; **22**: 577–8.
20. Yeager AM, Kanof ME, Kramer SS, Jones B, Saral R, Lake AM and Santos GW. Pneumatosis intestinalis in children after allogenic bone marrow transplantation. *Pediatr Radiol* 1987; **17**: 18–22.
21. Day LD, Ramsay KCN and Letourneau JG. Pneumatosis intestinalis after bone marrow transplantation. *AJR* 1988; **151**: 85–7.
22. Lagundoye SB and Itayemi SO. Tension pneumoperitoneum. *Br J Surg* 1970; **57**: 576–80.
23. Yale CE, Balish E, Wu JP. The bacterial etiology of pneumatosis cystoides intestinalis. *Arch Surg* 1974; **109**: 89–94.
24. Pfister J, Riedtmann-Klee HJ. Idiopathic spontaneous pneumoperitoneum. Case discussion based on two cases, assessment procedure and therapy and review of the literature. *Helvetica Chirurgica Acta* 1993; **60** (1–2): 49–56.

25. Capitanio MA and Greenberg SB. Pneumatosis intestinalis in two infants with rotavirus gastroenteritis. *Pediatr Radiol* 1991; **21**: 361–2.

26. Chippindale AJ and Desai S. Two unusual cases of pneumatosis coli. *Clin Radiol* 1991; **43**: 180–2.

27. Candill JL, Rose BS. The role of computed tomography in the evaluation of pneumatosis intestinalis. *J Clin Castroenterol* 1987; **9**: 223–6.

28. Thomas VI L, Cohen AJ, Omiya B, McKenzie R and Tominaga G. Pneumatosis intestinalis associated with needle catheter jejunostomy tubes: CT findings and implications. *J Comput Assist Tomogr* 1992; **16**: 418–9.

29. Connor R, Jones B, Fishman EK, Sigelman SS. Pneumatosis intestinalis: role of computed tomography in diagnosis and management. *J Comput Assist Tomogr* 1984; **8**: 269–71.

30. Kelvin FM, Korobkin M, Rauch RF, Rice RP, Silverman PM. Computed tomography of pneumatosis intestinalis. *J Comput Assist Tomogr* 1984; **8**: 276–8.

31. Chadba D. Kedar RP, Malde HM. Sonographic detection of pneumoperitoneum: an experimental and clinical study. *Australasian Radiology* 1993; **37** (2): 182-5.