Pneumatosis Intestinalis in Children with Leukaemia: Report of Three Cases

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Abstract. Three children with leukaemia (one with acute myeloid, two with acute lymphoblastic leukaemia) developed pneumatosis intestinalis during cytostatic treatment. The aetiology of pneumatosis intestinalis in these children could not be elucidated. Pneumatosis intestinalis may be caused by entry of gas into a bowel wall which is altered by steroid or cytostatic treatment. Otherwise, anaerobic bacteria may produce gas in the intestinal walls, therefore we treated all children with metronidazol.

Key words: Pneumatosis intestinalis - Leukaemia - Cytostatic therapy - Metronidazol

Introduction

In childhood pneumatosis intestinalis is a rare symptom characterized by multiple gas filled cysts in the submucosal or subserosal layer of the gastrointestinal tract. This condition is not necessarily associated with severe disease except for necrotizing enterocolitis in the newborn infant. Though many theories have been published the cause of this disease remains obscure. We wish to present three children all suffering from leukaemia who developed pneumatosis intestinalis during their illness.

Case Reports

1. Patient A.D., female, born 22.6.1965

In October 1971 acute lymphoblastic leukaemia was diagnosed. Treatment was initiated with prednisone and vincristine but without cranial radiation. In October 1972 she developed leukaemic meningitis. The skull was irradiated with 2000 rad. Maintenance therapy with cyclophosphamide, 6-mercaptopyrurine and amethopterine was finished 1977. In January 1979 she developed a left hemiparesis with central paresis of the facial nerve. On cranial computer tomography there were several areas of hyper-density. These areas were suspected of being leukaemic infiltrates and were treated by radiation with 1000 rad. Intensive chemotherapy was then started with prednisone, L-asparaginase, vincristine and daunorubicin for four weeks followed by a high dose amethopterine-therapy (500 mg/m²) with cytosine-arabinoside and 6-mercaptopurine. This therapy was tolerated well. Beginning in January 1980 we treated the child with amethopterine, nitrosourea (CCNU), procarbazine and dexamethasone. The therapy with dexamethasone ended in February 1980. Some days later the girl was admitted to the hospital because of severe maxillary sinusitis. The routine radiograph of the chest showed normal lungs but air under the diaphragm. On the lateral projection the air was in an anterior but also in a posterior position above the left lobe of the liver. The transverse colon was abnormal: It had lost the typical haustral appearance and the clear demarcation of the colonic wall, and it resembled soap bubbles. The abdominal radiograph disclosed the typical features of intestinal pneumatosis with ring shaped radio-lucencies along the ascending and transverse colon. The very clear demarcation of the upper psoas and the crus of the diaphragm led us to suspect that air had come along the retroperitoneal space and not via a gastrointestinal perforation. There were no clinical signs of gastroenteritis or enterocolitis: all tests for blood and pathogenic bacteria in the stools were negative. Nevertheless we started parenteral alimentation for several days. Antibiotic treatment consisted of gentamycin, cefotaxim and metronidazol. The radiological signs of pneumatosis intestinalis slowly disappeared over 14 days and the child has done well since then.


Acute myelocytic leukaemia was diagnosed in December 1979. Treatment consisted of fluocortolone and thioguanine for 10 days. We then added vincristine, daunorubicin and cytosine-arabinoside. Four weeks later she began to vomit and to have loose, bloody, mucus stools with a distended abdomen.

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Fig. 1. Patient L.D. The abdominal film with severe intestinal pneumatosis throughout the entire colon which is significantly distended. The psoas and the left crus of the diaphragm are sharply demarcated (arrows)

Abdominal plain films gave evidence of severe intestinal pneumatosis (Figs. 1 and 2). The left upper abdomen was exceptionally translucent due to a gas bubble which was assumed to be in the gastrocolic ligament. Radiologic signs increased during the next 8 days and gas extended along the left lateral abdominal wall. Again there was never evidence of free air in the abdomen on upright films indicating gastrointestinal perforation. The child was in poor condition and parenteral alimentation was initiated through a central venous catheter. In addition to the antibiotic therapy which she had for two weeks (sulfamethoxazol/trimethoprim (SMX/TMP), colistine, cefotaxim, gentamycin) we treated her with metronidazol because an anaerobic infection could not be excluded. During the following two weeks the condition of the patient slowly impaired. Stools normalized and occult blood could no longer be demonstrated. Four weeks after the diagnosis of intestinal pneumatosis the X-ray picture of the abdomen was normal.


In December 1979 she began to suffer from diffuse arthralgia. Investigation revealed acute lymphoblastic leukaemia. Treatment according to the scheme published by Riehm and Gadner (1974) was initiated in January 1980 (prednisone, vincristine, l-asparaginase and daunorubicin). After four weeks of treatment she developed cough with thoracic tenderness and a distended abdomen. X-ray films of the chest showed nothing abnormal within the thorax but disclosed pneumatosis of the ascending and transverse colon, and in addition gas under the right hemidiaphragm (Fig. 3). We began parenteral alimentation and, in addition to the antibiotic therapy with SMX/TMP, colistine and amphotericin B, we started a treatment with cefotaxim and metronidazol. After seven days the abdominal symptoms had resolved and the X-ray findings were again normal. Further cytostatic therapy (including dexamethasone) was tolerated without complications.

Discussion

The etiology and the pathophysiology of pneumatosis intestinalis are unknown. Several theories have been published (for a review see Ecker et al. 1971). The most convincing is that gas is forced mechanically into the bowel wall. Two major causes seem to predispose to pneumatosis intestinalis: i) A direct lesion of the mucosal layer of the intestinal tract, e.g. ulcer, tumor, volvulus, pyloric stenosis, perforation, infection, chronic enteritis; and also trauma during sigmoidoscopy (Olmstead and Madewell 1976). In all these instances an increased intraluminal pressure allows gas to enter into the bowel wall at the site of a "locus minoris resistentiae." Air can escape the intestinal lumen to enter venous vessels and end in portal veins, or enter lymphatics. A more widespread distribution can be seen when air has perforated into the interstitial...