CASE REPORT

Pneumatosis cystoides intestinalis confined to the small intestine treated with hyperbaric oxygen

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Paw HGW, Reed PN. *Pneumatosis cystoides intestinalis* confined to the small intestine treated with hyperbaric oxygen. Undersea Hyperbaric Med 1996; 23(2):115–000.—*Pneumatosis cystoides intestinalis* is a rare disease characterized by the presence of multiple intramural gas-filled cysts in the gastrointestinal tract. The etiology remains unknown, but the disease can present with profound disturbances of bowel function. We report the successful management of *pneumatosis cystoides intestinalis* of the small intestine with the use of hyperbaric oxygen.

Intestinal disease; Pneumatosis cystoides intestinalis, *respiratory therapy; hyperbaric oxygen*

Pneumatosis cystoides intestinalis is an uncommon condition of unknown etiology which is characterized by multiple gas-filled cysts in the mucosa of the intestine and also in the lesser and greater omentum, mesentery, lymph nodes, and sporadically in the wall of the gall bladder, bladder, and the vagina (1). It is generally referred to as *Pneumatosis coli* when the cysts are restricted to the colon. It is uncommon for the lesions to be confined to the small intestine and to cause such severe symptoms. It is more prevalent among men, with a ratio of 3:1, and occurs more frequently between the ages of 40 and 60 yr (2). These cysts vary in size from a few millimeters to 1 or 2 cm and may obliterate the bowel lumen. *Pneumatosis cystoides intestinalis* occurs mainly in middle-aged men and presents with a variety of symptoms such as mucous diarrhea and excessive flatus (3). Because these are common symptoms of carcinoma of the rectum and other colonic diseases, the correct diagnosis is only occasionally made in the first instance (4). Accurate diagnosis and knowledge of this treatment are important for avoidance of unnecessary surgery. We describe a patient with incapacitating symptoms from small intestinal pneumatosis who was treated with hyperbaric oxygen (HBO). The use of HBO in the colonic form has been previously described (5) but we had no knowledge of its use in small intestinal pneumatosis and believe this to be the first reported.

A 72-yr-old retired orthopedic surgeon with no known history of pulmonary or bowel disease was referred with a 6-mo. history of epigastric pain and increased frequency of bowel motion with passage of large quantities of flatus and weight loss. These symptoms became increasingly more severe and persistent, usually occurring 20 min after a meal. The epigastric pain was constant and would last for several hours. Hospital admission was required when the pain became intolerable and was accompanied by shoulder-tip pain. An erect chest x-ray revealed gas under the diaphragm. The plain abdominal x-ray taken in the erect position revealed fluid levels in the small intestine and a large amount of gas present. The plain abdominal x-ray taken in the supine position revealed numerous radiolucent cysts in the small intestine and small bowel dilatation. No diagnosis was made at this time. In the absence of peritonitis he was managed conservatively. Radiologic signs and symptoms settled within 2 days of i..v. fluid therapy and no oral intake. Two weeks after discharge, he was readmitted with similar symptoms. A barium meal and follow-through (Fig. 1) demonstrated numerous submucosal air-filled cysts within the wall of the small intestine. At this point, *Pneumatosis cystoides intestinalis* was recognized as the cause of his symptoms. Treatment was total parenteral nutrition with no oral intake. His symptoms improved but did not resolve completely in the next 10 days. In view of the severity and recurrent nature of his symptoms, it was felt that he should have a course of HBO therapy and was referred to the hyperbaric unit in Peterborough District Hospital. The use of HBO in the colonic form has been described previously (5) but we had no knowledge of its use in small intestinal pneumatosis.

He was treated in a Hyox monoplace chamber on an outpatient basis. Each course of treatment consisted of a 60-min period at 100% oxygen concentration at a pressure of 2 atm abs. His treatment regimen was five courses a week for 2 wk

FIG. 1—Barium follow-through examination before treatment showing extensive involvement of the cysts (*arrows*).

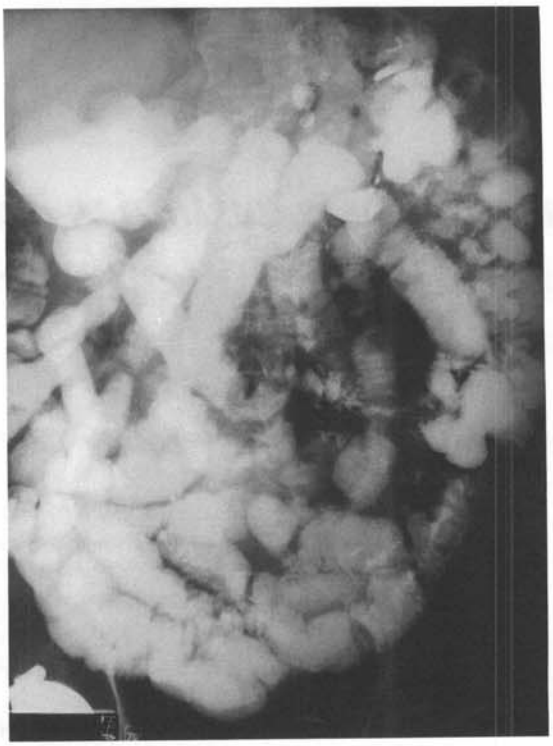

FIG. 2—Barium follow-through 3 mo. after treatment with HBO showing disappearance of the cysts.

reduced to two courses a week for another 2 wk and finally on a once weekly basis for a further 4 wk. He dramatically became symptom-free on the evening after his first treatment and continued to remain in remission to date (first treatment on 20 January 1995). Ten days after starting treatment his bowel habit returned to normal and he was able to resume his normal diet. At follow-up examination 3 mo. after treatment, repeat barium follow-through (Fig. 2) showed no recurrence of cysts. He did not suffer any pulmonary or neurologic sequelae of O_2 therapy. In fact, his only complaint was that of boredom.

DISCUSSION

For many years the etiology of *Pneumatosis cystoides intestinalis* has been a matter for debate (6). There are three hypotheses concerning the pathogenesis. The mechanical theory suggests that gastrointestinal gas, under abnormal pressure because of obstruction, is forced through tiny mucosal defects and enters submucosal layer. According to the bacterial theory, bacterial fermentation and gas build-up lead to cyst formation. Finally, the pulmonary hypothesis (7) postulated that severe coughing produces alveolar rupture and pneumomediastinum, with gas dissecting downward to the retroperitoneum and then along perivascular spaces to the bowel wall.

The introduction of oxygen as a successful form of therapy led some authors (8) to suggest that the cysts are created and maintained by a fastidious anaerobic gas-forming organism which produces gas at a rate that exceeded the rate of absorption until an equilibrium is reached. The high tissue PaO_2 achieved with O_2 therapy kills these organisms and the gas is then reabsorbed in the same way as gas contained within any natural or artificially created space in the body. This hypothesis has not been substantiated.

FIG

Oxygen therapy remains the cornerstone in the treatment of *Pneumatosis cystoides intestinalis*, but the most satisfactory method of its administration is not known. The use of HBO for the treatment of *Pneumatosis cystoides intestinalis* is not new (5) but there is no experience of its use in cases confined to the small intestine. Treatment with normobaric O_2 is well described in the literature (9–11) but is not without disadvantages. The main problem of normobaric O_2 is that it requires a high concentration to be maintained for long periods, thus requiring in-patient treatment. It also exposes the patient to pulmonary O_2 toxicity (11). Hyperbaric oxygen has the advantage that it requires a short duration of treatment and can be conducted on an out-patient basis.

The rationale for treatment of *Pneumatosis cystoides intestinalis* with O_2 in a high concentration is that other gases,

especially nitrogen, are washed out of the venous blood. This lowers the total pressure of gases in the surrounding tissues, thereby increasing the pressure gradient between the cysts and these tissues. The balance of pressure then favors diffusion, and the cysts will deflate. It is important for the surgeon to recognize the condition to prevent unnecessary surgical intervention. If pneumoperitoneum occurs in the benign form of *Pneumatosis cystoides intestinalis* without evidence of peritonitis (as was the case here), it should be managed conservatively.

It is hoped that this aggressive HBO therapy regimen will incur a long-term remission.

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REFERENCES

1. Jamart J. *Pneumatosis cystoides intestinalis*: a statistical study of 919 cases. Acta Hepato-Gastroenterol 1979; 26 419–422.
2. Shallal JA, Van Heerden JA, Batholomew LG, Cain JC. *Pneumatosis cystoides intestinalis.* Mayo Clinic Proc 1974; 49:180–184.
3. Hughes DTD, Gordon KCD, Swann JC, Bolt GL. *Pneumatosis cystoides intestinalis*. GUT 1966; 7: 553–557.
4. Smith WG, Anderson MJ, Pemberton HW. *Pneumatosis cystoides intestinalis* involving left portion of colon. Gastroenterology 1958; 35:528–533.
5. Masterson JST, FratkinN LB, OslerTR, Trapp WG. Treatment of *Pneumatosis cystoides intestinalis* with hyperbaric oxygen. Ann Surg 1978; 187:245–247.
6. Reyna R, Soper RT, Condon RE. *Pneumatosis intestinalis.* Am J Surg 1973; 125: 667–671.
7. Keyting WS, McCarver RR, Kovarik JL, Daywitt AL. *Pneumatosis intestinalis*: a new concept. Radiology 1961; 76:733–741.
8. Down RHL, Castleden WM. Oxygen therapy for *Pneumatosis coli*. Br Med J 1975; 1:493–494.
9. Forgacs P, Wright PH, Wyatt AP. Treatment of intestinal gas cysts by oxygen breathing. Lancet 1973; 17: 579–581
10. Elberg JJ. Oxygen therapy for *Pneumatosis coli*. Acta Chir Scand 1985; 151:399–400
11. Simon NM, Nyman KE, Divertie MB, et al. *Pneumatosis cystoides intestinalis*: treatment with oxygen via a close-fitting mask. JAMA 1975; 231:1354–1356