Pneumatosis cystoides intestinalis


From The London Hospital

EDITORIAL COMMENT  The association with respiratory disease is further confirmed in this detailed study. The mechanism and its production is still uncertain. The condition tends to run a benign course.

There have been several recent accounts of this unusual condition in which gas-filled cysts are found subserosally and submucosally in the small and large bowel (Koss, 1952; Marshak et al., 1956; Burn and Jones, 1961; Colquhoun, 1965). Patients may present with symptoms ranging from intermittent mild rectal bleeding to intestinal obstruction, though occasionally the condition is symptomless and only revealed by radiological examination. The exact mechanism whereby cysts are formed is uncertain, but they are often found in association with disease elsewhere in the gastrointestinal tract. It has also been pointed out that they can occur in the presence of chronic pulmonary disease (Doub and Shea, 1960; Keyting, McCarver, Kovarik, and Daywitt, 1961; Kenney, 1963) and there has been speculation on their possible relationship to changes in blood gas tensions.

Analysis in vivo of some components of the cyst gas has been carried out (Mujahed and Evans, 1958) and other authors have analysed contents after death with a wide range of results (Sauser-Hall, 1940; Mackenzie, 1951; Bilger, 1956).

This paper presents five cases, four with pulmonary disease; all had simple pulmonary function tests performed and in one an accurate analysis of the cyst gas was made.

MATERIALS AND METHODS

Conventional straight radiographs of chest and abdomen were taken and barium studies of the bowel were carried out using Micropaque. Lung function tests were performed using the techniques normally employed in the Lung Function Laboratory at The London Hospital (Hughes and Lee, 1963). In addition, the maximum expiratory flow rates were determined by the method described by Kory, Callahan, Boren, and Syner (1961). This should exceed 200 l/min. at any age up to 70 years, values below indicating some degree of obstructive Airways disease. In analysing samples of gas from the cysts in case 4 the mass spectrometer was used for estimating nitrogen and hydrogen, an infra-red spectrometer for nitrous oxide and carbon dioxide, and gas chromatography for the remainder and to check the hydrogen value.

CASE REPORTS

CASE 1  B.B. was a 54-year-old man who had suffered from generalized scleroderma for 11 years. Five years previously, when he had complained of dysphagia, a

FIG. 1. Detail of straight radiograph of abdomen showing pneumoperitoneum, basal fibrosis at right lung base, numerous radiolucent cysts in the small bowel, some loops of which are interposed between the liver and diaphragm (case 1).
barium meal and follow-through had demonstrated a dilated and atonic lower oesophagus, normal stomach and duodenum, an atonic jejunum, and a normal colon. He was given prednisone and remained well until 1963 when he complained of abdominal pain, distension, and diarrhoea with flatus. His seven or eight daily stools were semi-watery and contained considerable quantities of mucus but no blood. He had typical sclerodermatous changes of the hands and face, bilateral basal crepitations in the chest, and a distended abdomen, in which, however, no masses were felt and rectal examination was normal. Haemoglobin, leucocyte count, and E.S.R. were normal, but the faecal fat excretion was 7-6 g. per day. Sigmoidoscopy was normal. Radiographs of the chest showed bilateral basal fibrosis. A straight film of the abdomen demonstrated obvious pneumoperitoneum with loops of gut interposed between the liver and diaphragm (Chilaiditi sign) (Fig. 1). The small gut was dilated with numerous radiolucent cysts, apparently within its wall. A barium swallow and meal gave the same results as before and a barium follow-through showed extensive involvement of the small bowel with pneumatosi, (Hughes and Lee, 1963).

Case 2 A.C. was a tailor (weight 14 st.) of 59 years, whose mother, father, and two brothers had died from 'chest trouble'. He was a heavy smoker (30-40 cigarettes daily all his life) who had been discharged from the Army with bronchitis. This disease had occurred in the winter for the past 15 to 20 years with varying severity. During the last four years he had been in hospital several times with exacerbations of bronchitis associated with heart failure, and on one occasion possible myocardial infarction. Twice during the war he had had bleeding haemorrhoids injected and there was a further recurrence in 1956. In January 1963 he again developed rectal bleeding with

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Vital Capacity (ml.)</th>
<th>Percentage of Predicted Vital Capacity</th>
<th>FEVI as Percentage of Vital Capacity (normal &gt; 70%)</th>
<th>MEFR (normal &gt; 200 l/min.)</th>
<th>RV/TLC Ratio (normal &lt; 40%)</th>
<th>Diffusion and Percentage Predicted</th>
<th>PCO₂ (normal 37-45 mm.Hg)</th>
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<tr>
<td>1</td>
<td>2,100</td>
<td>60</td>
<td>75</td>
<td>240</td>
<td>30</td>
<td>9 (42)</td>
<td>43</td>
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<tr>
<td>2</td>
<td>1,600</td>
<td>40</td>
<td>43</td>
<td>60</td>
<td>74</td>
<td>—</td>
<td>73</td>
</tr>
<tr>
<td>3</td>
<td>1,800</td>
<td>69</td>
<td>71</td>
<td>100</td>
<td>61</td>
<td>—</td>
<td>37</td>
</tr>
<tr>
<td>4</td>
<td>2,500</td>
<td>93</td>
<td>68</td>
<td>175</td>
<td>50</td>
<td>23 (110)</td>
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<td>3,450</td>
<td>94</td>
<td>78</td>
<td>200</td>
<td>22</td>
<td>26 (116)</td>
<td>44</td>
</tr>
</tbody>
</table>
Pneumatosis cystoides intestinalis

the passage of loose stools two or three times daily with excessive flatus. On examination, he was an obese, plethoric man with a barrel-shaped chest and loud expiratory rhonchi heard throughout the lungs. The abdomen was a little distended, but no masses were felt and rectal examination was normal Haemoglobin was 110%, haematocrit 56%, and the leucocyte count and E.S.R. were normal. The faecal fat was 6.5 g. per day, and sigmoidoscopy showed first-degree piles and numerous submucous cysts, partly obliterating the lumen of the bowel from 12 cm. upwards.

A chest radiograph demonstrated emphysematous lungs with prominent pulmonary vessels and a barium enema showed gross pneumatoses cystoides of the whole sigmoid and descending colon (Fig. 3). Lung function tests were typical of emphysema with carbon dioxide retention (Table I).

case 3 J.W. was a man of 65 years, who smoked 20 cigarettes a day and suffered from recurrent winter bronchitis. Because of diarrhoea and weight loss, he was admitted to hospital in June 1963, when he was passing up to three watery stools daily with mucus, excessive flatus, but no blood. There were no outstanding features on physical examination, but later sigmoidoscopy showed multiple glistening blue-purple submucous cysts in the rectum and up to 30 cm.; several of these were ruptured with the release of gas. Microscopic examination of biopsy specimens showed cysts lying below the muscularis mucosae, lined by large pink cells, with a minimal and localized inflammatory reaction with macrophages and polymorphs. A chest radiograph and barium meal were normal but in a straight film of the abdomen numerous gas-filled cysts were seen in the left upper abdomen, and on barium enema there were changes of pneumatoses cystoides involving the descending and sigmoid colon. Lung function tests indicated obstructive airways disease (Table I).

case 4 A.L. was a retired case-maker of 73 years, with weight loss (weight 9 st.), marked diarrhoea, associated with considerable flatus and mucus but no macroscopic blood. He had benefited from mist. kaolin et morph., and four years previously a barium meal, cholecystogram, and barium enema had all been normal, save for a small solitary gall-stone. Apart from a palpable descending colon there was no abnormality on examination, but sigmoidoscopy revealed submucosal cysts projecting into the lumen from 14 to 25 cm., and biopsies were reported as normal colonic mucosa. A barium enema demonstrated pneumatoses cystoides intestinalis involving the sigmoid, descending, and left half of the transverse colon. Lung function tests indicated only a mild degree of obstructive airways disease (Table I).

As three specimens of stool contained occult blood, carcinoma of the colon was suspected and laparotomy performed. The colon from the mid portion of its transverse course to the sigmoid was covered by several hundred cysts projecting from the serosal surface. The remainder of the gastrointestinal tract was normal and no carcinoma was found. Gas was obtained from the cysts by aspiration through liquid paraffin into sealed glass syringes, and analysis is shown in Table II.

case 5 V.D. was a 47-year-old man with a six-month history of abdominal pain, distension, diarrhoea, and excessive flatus. His stools were loose, watery with mucus but no blood. Apart from rheumatic fever at the age of 5, there was no significant illness in his past history, but he had smoked 40 cigarettes a day for many years and suffered from mild winter bronchitis. He appeared fit but had a mitral diastolic murmur, slight epigastric tenderness, and distension of the descending colon. Rectal examination was normal and at sigmoidoscopy multiple glistening translucent cysts were seen up to 30 cm. Haemoglobin was 109% with a normal E.S.R. and leucocyte count. A straight radiograph of the abdomen displayed multiple air-filled cysts in the transverse and descending colon and a barium enema confirmed the presence of pneumatoses cystoides. Because of the epigastric pain a barium meal was carried out and a small ulcer on the lesser curve was seen. There were, however, no cysts in the small intestine, and a chest radiograph and lung function tests were normal.

FOLLOW-UP

These five cases have now been followed-up for periods ranging from nine months to two and a half years. Their symptoms have remained static in most cases, though the diarrhoea seems a little less in case 2. There has been no significant weight loss in any. The appearance of the barium enema repeated four months after initial diagnosis in case 4 was unchanged.

DISCUSSION

Although this condition has been reported on several occasions in the past, the present series is larger than most, and from the study of the five patients a number of interesting facts emerge. In all five patients the characteristic mode of presentation was

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**TABLE II**

Analysis of gas removed from cysts at laparotomy in case 4

<table>
<thead>
<tr>
<th>Gas</th>
<th>Percentage</th>
<th>Method of analysis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Nitrogen</td>
<td>72.5%</td>
<td>Mass spectrometer</td>
</tr>
<tr>
<td>Hydrogen</td>
<td>10.0%</td>
<td>Mass spectrometer</td>
</tr>
<tr>
<td>Oxygen</td>
<td>7.0%</td>
<td>Gas chromatography</td>
</tr>
<tr>
<td>Nitrous oxide</td>
<td>4.5%</td>
<td>Infra-red spectrometer</td>
</tr>
<tr>
<td>N-butane</td>
<td>0.3%</td>
<td>Gas chromatography</td>
</tr>
<tr>
<td>Iso-butane</td>
<td>0.15%</td>
<td>Gas chromatography</td>
</tr>
<tr>
<td>Propane</td>
<td>0.075%</td>
<td>Gas chromatography</td>
</tr>
<tr>
<td>Methane</td>
<td>0.038%</td>
<td>Gas chromatography</td>
</tr>
<tr>
<td>Ethane</td>
<td>0.01%</td>
<td>Gas chromatography</td>
</tr>
<tr>
<td>Argon</td>
<td>1.40%</td>
<td>Infra-red spectrometer</td>
</tr>
</tbody>
</table>

*Value checked by two methods.*

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that of diarrhoea and excessive flatus, whilst four had considerable amounts of mucus in their stools. The passage of blood in one patient was thought to have been associated with his haemorrhoids. Such symptoms may suggest a malabsorption state, and in fact two of the present series had a raised faecal fat excretion. Malabsorption secondary to pneumatosis of the small bowel has previously been described by Yunich and Fradkin (1958). Alternatively such altered bowel habit might lead to a suspicion of large bowel carcinoma as in case 4. There is in fact no evidence that pneumatosis is pre-carcinomatous, though it has been reported in association with carcinoma of the stomach (Dale and Pearse, 1950) and widespread abdominal carcinomatosis (Thorpe, 1965). It has also been described in association with lymphosarcoma (Williams, Sutherland, and Clark, 1963) and in this instance there was also malabsorption, though this appeared to be secondary to the lymphosarcoma rather than the pneumatosis. Radiological examination may show the cysts on a straight film of the abdomen but even better contrast is obtained by barium studies. Sigmoidoscopy can be of considerable value as the appearance of intraluminal cysts is characteristic, though according to some reports, the large bowel is only infrequently involved. In this series the commonest site of the lesion was in the large bowel and this is at variance with the distribution in the largest series of cases reported so far (Koss, 1952). This included many cases in children in which the lesion is probably of different aetiology. In a recent article Colquhoun (1965) has distinguished true pneumatosis cystoides intestinalis from other conditions in which gas is found in the walls of hollow viscera. He states that *Pneumatosis cystoides* is most common in the small intestine whilst Koss in his less strictly defined series reported only 13 cases in the large bowel. Sigmoidoscopic diagnosis has been made in few cases. Marshak, Blum, and Eliasoph (1956) reported four cases, and Pemberton, Smith, and Holman (1957), in describing a further case, stated that until their report they could only find seven previous descriptions of this diagnostic success.

Abnormalities of pulmonary function were found in four of the five patients, but there was no constant pattern, obstructive airways disease being present in three cases, and a restrictive pattern with diffusion block in the fourth. Arterial carbon dioxide tension was elevated in one case only.

AETIOLOGY

The aetiology remains obscure and several mechanisms have been postulated:—

**BACTERIAL INFECTION** This was suggested by Naeslund (1924) and others who found that in animals small intestinal infection by certain bacteria produced a similar condition. Many of the cases involving the small gut in infants (Mackenzie, 1951) have been associated with enteritis. However, nothing to incriminate infection has been described in adults.

**DIETARY DEFICIENCY** There is little evidence of this, although some infants with the condition were considered undernourished.

**NEOPLASTIC AND MECHANICAL FACTORS** Allusion has already been made to this (Thorpe, 1965). A number of cases with pneumatosis have been associated with lesions involving loss of integrity of the intestinal mucosa (this was so in some 89% of Koss's cases), and in two of our series there were lesions (scleroderma and gastric ulcer) that could have led to a breach of epithelial continuity. It has been postulated that gas enters through these spaces and permeates submucosally.

**PULMONARY FACTORS** The relationship of pneumatosis to pulmonary disease is complicated. A purely mechanical explanation has been suggested. Keyting et al. (1961) described three patients with pneumatosis occurring in the presence of severe obstructive pulmonary disease in exacerbation who had previously been entirely free from gastrointestinal symptoms. These writers considered that severe coughing bouts might lead to alveolar rupture forcing air into the mediastinum whence it might track through the diaphragm and retroperitoneal tissue, finally emerging along the intestinal arteries in the subserosal plane. Their injection experiments in animals partially supported this thesis. Doub and Shea (1960) reported on 16 patients with pneumatosis, 15 of whom had asthma and another pulmonary fibrosis. They commented on this association with chest disease but suggested no mechanism. Kenney (1963) put forward an intriguing hypothesis, pointing out that some observers had found that the cyst gas contained up to 15% carbon dioxide. The presence of such a soluble and easily diffusible gas in high concentration would only seem feasible if it were present in excess in the body. He further indicated that some diseases described in association with pneumatosis were complicated by excess bicarbonate ion (pyloric obstruction with vomiting and carbon dioxide retention in chronic obstructive lung disease with respiratory failure) and suggested that the excess carbon dioxide led to cyst formation.

The present series confirms the condition to be associated with pulmonary disease but fails to provide an explanation. Kenney's hypothesis receives
no support, as carbon dioxide retention was elevated in one case only and the cyst gas in case 4 had a lower carbon dioxide content than that in alveolar air. The concentrations found were such as to suggest an admixture of gases from blood and intestinal lumen, those of respiratory origin being less than in alveolar air, or arterial blood, or even the anaesthetic, nitrous oxide, assuming a 20 to 25% mixture.

Among the several intestinal gases present there was a surprisingly high concentration of hydrogen, possibly produced by intraluminal bacterial activity. Methane has previously been reported in cyst gas (Urban, 1910) while a more recent analysis by Mujahed and Evans (1958) showed the presence of 89.96% N, 27.62% CO₂ and 2.42% oxygen. There is obviously a need for further accurate analyses of gas from the cysts in this condition. Though the aetiology remains obscure the prognosis in this condition appears good, and patients should be reassured that they have a benign condition.

SUMMARY

Five cases of pneumatosis cystoides intestinalis are presented and their clinical and pathological features are discussed. Diarrhoea with excessive flatus may suggest malabsorption or altered bowel habit a carcinoma of the large bowel. The condition, however, appears benign. Pulmonary function studies were carried out on these patients and confirmed the presence of pulmonary disease in four cases. Gas was withdrawn from the cysts at laparotomy in one case and was accurately analysed. The association of this unusual condition with pulmonary disease was therefore again confirmed although the exact mechanism remains uncertain.

We wish to thank members of the Analytical Section, British Oxygen Company Scientific Sector, under the supervision of Mr. J. H. Glover, and Dr. D. W. Hill, of the Anaesthetic Department, Royal College of Surgeons, who analysed the samples of gas from cysts in Case 4.

REFERENCES

Pneumatosis cystoides intestinalis.

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