

PNEUMATOSIS CYSTOIDES INTESTINALIS

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Pneumatosis cystoides intestinalis is characterized by serous, subserous and submucous gas cysts occurring anywhere between the stomach and rectum but mainly in the caecum and terminal ileum. The condition is found most frequently in males in the 4th—6th decades. It may exist either in asymptomatic and apparently innocuous form, in which the cysts are of the 'primary' type, or it may be secondary to gastric or duodenal ulceration, stenotic pyloric lesions, intestinal obstruction from various causes, acute and chronic appendicitis or intestinal parasites.

The symptoms are vomiting, abdominal distension, abdominal pains, constipation, diarrhoea and other results of interference with transit of the intestinal contents.

The cysts are mainly confined to the small bowel, terminal ileum and caecum. The complications that may occur are (1) pneumoperitoneum, although Lerner and Gazin³ claim that this is not due to free air but to accumulations of gas lying between the muscular and peritoneal layers of the diaphragm, (2) chronic constipation, (3) intestinal obstruction due to obliteration of bowel lumen by submucous cysts, and (4) volvulus or adhesions due to serous or subserous cysts.

On straight X-ray of the abdomen, multiple scattered rounded translucencies may be seen, which bear some relation to the bowel wall but do not contain fluid levels. A barium meal and enema will demonstrate multiple polyp-

like filling defects of the bowel lumen with the bowel wall outlined by translucent gas-filled cysts producing a scalloped bowel margin.

Many theories have been advanced to explain the origin of the cysts but the one most favoured by Koss² is that, owing to some change in the hydrogen-ion concentration of the bowel contents, gas diffuses into blocked and stagnant lymphatics. Anton *et al.*¹ consider that it is due to the poor nutritional state of the patient combined with chronic obstruction.

Recent literature indicates that the majority of cases are not diagnosed pre-operatively but are found in the course of an operation for an associated abdominal complaint, or during an exploratory laparotomy to exclude multiple polyposis or a malignant polypoid tumour. One case reported by McKay⁵ was diagnosed by barium enema as multiple polyposis. Another case reported by Rounthwaite *et al.*⁷ was operated on for a polypoid type of tumour thought by the radiologist to be malignant. Ramos and Powers⁶ reported a case with increasing constipation and lower abdominal distension which on barium enema was diagnosed as polyposis with obstruction of the sigmoid. A hemicolectomy was performed, and histological examination showed the case to be one of pneumatosis.

Koss,² who in 1952 reviewed 213 cases in American and European literature, found that in his own series of 33 cases only 9 were in the large bowel. According to Villaume,⁸ it is uncommon in the sigmoid, but isolated cases have been reported by McGee *et al.*⁴ and Ramos and Powers.⁶

The following case presents such distinctive clinical and radiological signs, that although there is no histological proof, there can be no doubt about the diagnosis.

CASE REPORT

Mr. G., aged 47, a European male, was seen in the out-patient department in 1941 complaining of attacks of abdominal pain. A barium meal and enema revealed no abnormality. In 1954 he had a coronary thrombosis and in June 1955 a chest X-ray demonstrated gas under the right dome of the diaphragm. The case was investigated with the object of excluding a leaking peptic ulcer. A barium meal revealed some deformity of the duodenal cap but no definite ulcer. Barium meals repeated on 6 November and 23 November 1955 were negative. The patient next reported on 2 December 1958 complaining of abdominal distension and unsatisfactory emptying of the bowel, and also of rectal bleeding which he had been experiencing on and off for many years. Haemorrhoids were found on clinical examination. A barium enema revealed multiple polypoidal filling defects and a differential diagnosis of possible polyposis, faecal masses or granulomatosis was suggested. The barium enema was repeated and multiple translucencies were demonstrated; some were intraluminal but the majority of the gas cysts were shown outlining the bowel margins (Fig. 1). The sigmoid colon presented a redundant loop and it was here that the majority of the translucencies were situated. Some were observed in the descending colon, and a few scattered cysts in the transverse colon.

In the differential diagnosis the distinction from multiple polyposis was based on the following features:

1. The greater radiolucency of the filling defects in the barium column as compared with the soft-tissue density of polyp or other tumours. The cysts have the same radiolucency as the air which was used as a medium for double-contrast enema.
2. The change in the shape and size of the gas cysts, with flattening of the base, when the bowel is distended.
3. Because of the localization of cysts within the bowel wall, a portion of the translucent defect extends well beyond the contour of the barium column (Fig. 1).

DISCUSSION

An accurate diagnosis of this benign condition was important because the patient, owing to his previous cardiac infarct,



Fig. 1. Barium enema demonstrating translucent filling defects in bowel lumen and gas cysts outlining the margins of the sigmoid colon.

was not a suitable candidate for an exploratory abdominal operation. Although many cases have been diagnosed initially as polyposis, the resemblance of pneumatosis to multiple polyposis is very superficial; certainly in this case the X-ray features were unequivocal. The cysts were confined largely to the sigmoid colon, and undoubtedly the patient was already suffering from this condition in 1955, when a pneumoperitoneum was reported.

SUMMARY

A case of pneumatosis cystoides intestinalis is presented. It is unusual in that the gas cysts are mainly confined to the sigmoid colon. Although the patient developed a pneumoperitoneum in 1955, the cause of this only became obvious in 1959 at the barium enema examination.

The symptoms, complications and radiology of the condition are briefly discussed and a reference is made to recent literature.

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