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Pneumatosis cystoides intestinalis

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Summary

A brief outline of the aetiology and pathogenesis of pneumatosis cystoides intestinalis is given followed by a review of four cases which have occurred in the Glasgow area recently. The radiological appearances of the condition are considered in detail.

Pneumatosis cystoides intestinalis (PCI) is a rare but well-documented condition in which intramural gas appears throughout the intestine. The radiological appearances may be characteristic and four cases which have occurred in the Glasgow area recently are presented to illustrate these appearances.

Pathology

The disease is characterized by gas-filled cysts lying submucosally and subserosally which can occur

along the whole length of the bowel although the stomach and the duodenum are very rarely affected. The cysts are lined by a single layer of cuboidal or flattened endothelial cells and separated by hyaline tissue which is infiltrated with lymphocytes or histiocytes and occasional foreign-body giant-cells. The wall of the cyst retains its normal capillary blood supply. Analysis of the gas has varied in different reports from normal atmospheric air to a gas containing a high concentration of carbon dioxide (15%).

Site

Although the condition was originally thought to be commoner in the small bowel (Koss, 1952) the disease is being more often reported in the large bowel but this may be due to increasing radiological awareness of the disease in this site (Colquhoun, 1965; Meikle, 1965; Dendy Moore, 1968).

Incidence

The condition is three and a half times more common in men and occurs generally between the ages of 30 and 50 years. In women, the range is between 50 and 60 years.

Etiology

A number of theories have been put forward to explain the condition. The two most widely held are those which postulate a mechanical basis and those in which there is an upset in the acid-base balance.

Mechanical theory

These cases fall into two groups, firstly those in which gas has been forced through a breach in the mucosa of the intestinal tract such as an ulcer particularly when there is a degree of obstruction. It was found that 58% of cases were associated with peptic ulcer and pyloric stenosis (Koss, 1952). In support of this theory a case has been reported where there was successful treatment of PCI by pyloroplasty (Dhall, Mahaffy & Matheson, 1968).

In the second group the patients suffer from long-standing obstructive respiratory disease and it is thought that the gas has been forced down from the thorax and into the bowel via the aortic sheath and the mesentery.

Acid-base theory

Kenney (1963) postulated that certain patients who were suffering from an acid base disturbance in which there was a rise in the carbon dioxide, e.g. pyloric stenosis and chronic respiratory disease, were unable to excrete it in the normal way and that the bowel provided an additional means. The above theories do not explain those cases at present classified as 'primary', i.e. not existing concomitantly with any other disease. It has also to be explained why more patients do not develop pneumatosis in view of the frequency of both respiratory disease and peptic ulceration.

Symptomatology

The disease may be symptomless or may present with any of the more usual bowel symptoms.

Case reports

Case 1

A male patient aged 65 presented with a 6-week history of painless diarrhoea eight times a day containing copious mucus and occasional fresh blood. He also complained of loss of appetite and weight and had clinical and radiological evidence of bronchitis and emphysema and was on steroids for asthma. On examination a tender mass was palpable in the left iliac fossa. A barium enema revealed the

presence of PCI, the condition affecting the whole of the large bowel from the caecum to the recto-sigmoid junction. (Fig. 1). There was no evidence of

FIG. 1. (Case 1) Barium enema. The submucosal cysts are seen as translucent filling defects indenting and narrowing the lumen and the subserosal cysts as translucent lines along the margin of the bowel.

carcinoma. Sigmoidoscopy was done and showed the presence of a large number of submucosal cysts. A biopsy was taken and showed oedematous but otherwise normal rectal mucosa. Following this, a barium meal and follow-through demonstrated no evidence of peptic ulceration or obstruction or small bowel involvement.

One month later a second barium enema showed no change in the condition. No specific treatment was instituted and the patient was discharged. Two weeks later he was admitted with perforation of an acute greater curve gastric ulcer. Immediately post-operatively he made good progress but subsequently went downhill and died. At necropsy no obvious cause for his death was found and it was postulated that he died from biochemical abnormalities. PCI was found involving the large bowel from the ascending colon to the recto-sigmoid junction. The submucosal cysts almost obliterated the lumen of the large bowel. The serosal surface was also affected as were many of the fatty appendages.

Case 2

Male, aged 58. Admitted with a 6-week history of painless diarrhoea two to three times a day containing mucus but no blood and the passage of considerable flatus. He had a negative chest X-ray but had been taking one tablet of betamethasone at night over the previous 3 years for bronchospasm. On examination the abdomen and rectum were normal. Sigmoidoscopy revealed multiple 'polyps' starting at 15 cm. Biopsy showed mild inflammatory changes of the mucosa in the specimen received. There was no evidence of polyps or malignancy. A barium enema showed PCI of the sigmoid colon. The bowel symptoms were treated symptomatically and six months later the patient was doing well. A follow-up plain film after 1 year showed PCI unchanged (Fig. 2).

FIG. 2. (Case 2) Plain film of abdomen showing large bowel submucosal cysts in a redundant loop of sigmoid.

Case 3

Female, aged 75, admitted for investigation of fresh blood passed per rectum and constipation. There was no history of any respiratory disease and her chest X-ray was normal. Physical examination was normal except for the presence of haemorrhoids

which were confirmed by proctoscopy and found to be the cause of the bleeding. Sigmoidoscopy was performed and gas-filled cysts were noted which were biopsied. At the time of the biopsy an escape of gas was noted. The histology showed non-specific chronic inflammatory changes. A barium enema showed hold-up in the sigmoid due to the narrowing of the lumen by cysts (Fig. 3). No barium

FIG. 3. (Case 3) Barium enema. The film shows hold-up of the barium in the sigmoid by submucosal cysts. The narrowed lumen proximally is represented by a central white stripe.

passed beyond the descending colon. The appearances were characteristic of PCI. A barium meal and follow-through showed the disease to involve the distal transverse colon, splenic flexure, descending colon and sigmoid colon. The patient was treated symptomatically and after 7 months a plain film showed disease still present.

Case 4

Male aged 56. Admitted with pyloric stenosis following a 20-year history of ulcer dyspepsia. Barium meal confirmed marked pyloric stenosis.

There was interposition of colon between the liver and the right dome of the diaphragm and cystic areas were noted under both domes of the diaphragm. A diagnosis of PCI was made.

The patient underwent vagotomy and pyloroplasty and at operation the diagnosis was confirmed involving the small bowel only. There was also a partial volvulus of the proximal small bowel. Six days later the abdomen was re-opened after dehiscence of the wound and no cysts were present.

Radiology

Plain film appearances

Kenney in 1963 first drew attention to the significance of the plain film appearances. The cysts in the small bowel are usually subserosal and tend to assume a spherical shape which will cast a circular shadow and lie along the line of the small bowel. The submucosal cysts in the large bowel have a flat base and are convex into the lumen. They vary in size but the larger ones will tend to obliterate the bowel giving rise to a characteristic central stripe. At first glance, the plain film suggests extensive gaseous distension of the large bowel but in fact the gas does not lie within the lumen (Fig. 4).

FIG. 4. (Case 1) Plain film appearances. Whole of large bowel affected by submucosal and subserosal cysts.

The subserosal cysts give rise to a linear translucency lying along the base of the submucosal cysts.

Both large and small bowel can rise and become interposed between the liver and the diaphragm. If

multiple small cysts are projected over each other they give rise to a 'foamy' appearance, an effect best seen in conjunction with hepato-diaphragmatic interposition of colon.

Pneumoperitoneum is a not unusual plain film finding in PCI.

Barium examination

In the small bowel the cysts give rise to translucent polypoidal type filling defects along the walls of the bowel. The smaller submucosal cysts of the large bowel have a similar appearance. The larger ones gradually obliterate the lumen which is reduced to a central white stripe. This is best shown on the post-evacuation film (Figs. 1 and 5). During barium

FIG. 5. (Case 1) Post evacuation film. The central white stripe representing the partially obliterated lumen is well seen.

enema examination the cysts will tend to be flattened by the pressure of the barium but on the post-evacuation film will resume their normal shape. Polyps on the other hand will not be altered by the pressure of barium. Subserosal cysts best seen on the barium film will give rise to a translucent stripe lying along the bases of the submucosal cysts (Fig. 1).

Complications

In summary these are:

1. **Obstruction.** PCI is thought to give rise to obstruction in one of four ways. The submucosal cysts

may physically obliterate the lumen (Meikle, 1955. Fig. 3). The cysts may induce a volvulus; the cysts may exert extrinsic pressure on other loops of bowel. They may give rise to inflammatory reactions and adhesions (Jones & Cole, 1967).

2. *Intussusception*. A submucosal cyst may act as the apex of an intussusceptum.

3. *Pneumoperitoneum*. In rare cases, tension pneumoperitoneum may occur.

4. *Interposition of the bowel between the liver and the diaphragm* (Case 4, Dhall *et al.*, 1968).

Discussion

PCI is a rare condition the diagnosis of which can usually be made with certainty if the possibility is considered. The disease may be discovered as an incidental finding on plain films of the chest and abdomen. The chest X-ray, which may show evidence of chronic respiratory disease can demonstrate interposition or a pneumoperitoneum with cysts and a 'foamy' appearance below the diaphragm. PCI should always be considered when a pneumoperitoneum occurs without other clinical evidence of perforation of a hollow viscus (Somerville, 1951). These appearances may also be found on the plain film of the abdomen and in addition cysts may be seen along the line of the bowel, particularly the colon. The plain film appearances are unmistakable once it is realized that the gas is not intraluminal.

A barium enema will confirm the findings in the large bowel. The differential diagnosis includes

faeces, polyposis, the pseudo-polypoidal form of ulcerative colitis, thumb-printing in mesenteric vascular disease and carcinoma but these conditions will not give rise to *translucent* filling defects.

The diagnosis is less easy if the disease is confined to the small bowel and will be best demonstrated in the follow-through examination. The preceding meal will also reveal any peptic ulceration or stenosis.

Once the diagnosis is made and alimentary tract malignancy has been excluded there is no specific treatment and spontaneous remission can be expected if complications do not supervene.

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Hypercalcaemia in thyrotoxicosis with and without hyperparathyroidism

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Summary

Two cases of thyrotoxicosis with hypercalcaemia are reported. The hypercalcaemia in one of the patients was a manifestation of hyperthyroidism and in the other was due to co-existent hyperparathyroidism.

The literature is reviewed and the effect of thyroid hormone on calcium metabolism is discussed. Routine plasma calcium estimations should be performed in all cases of thyrotoxicosis.

Introduction

Hypercalcaemia in thyroid disease occurs infrequently. Significant hypercalcaemia in thyrotoxicosis may occur with or without accompanying

hyperparathyroidism, and an example of each is reported.

Method

Plasma calcium was measured by the EDTA/calcon method (Bowden & Patston, 1963) and all values were corrected for total serum protein, as suggested by Dent (1962). The normal range of plasma calcium in this laboratory is 8.4–10.5 mg/100 ml.

Case 1: Thyrotoxic hypercalcaemia

A 25-year-old unmarried woman was admitted to Westminster Hospital in February 1969. She gave a