PNEUMATOSIS INTESTINALIS:
A case report with a brief review of literature

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A 49-YEAR-OLD Kadazan male was seen at the Queen Elizabeth Hospital, Sabah, on July 18th 1969 with complaints of severe abdominal pain and vomiting of 24-hour duration. He also gave a history of recurrent attacks of epigastric pain after meals for the past 10 years, often associated with vomiting of food. A clinical diagnosis of acute intestinal obstruction was made supported by pre-operative portable X-rays of the abdomen which showed evidence of intestinal obstruction. At laparotamy, no abnormality except "a mass 12 inches long in the lower ileum" was found. A post-operative diagnosis of ? pneumatic cysts was made and the specimen was sent for histopathological examination. The affected segment was resected and an end-to-end anastomosis was performed. Post-operatively, the patient lapsed into irreversible shock and died the following day.

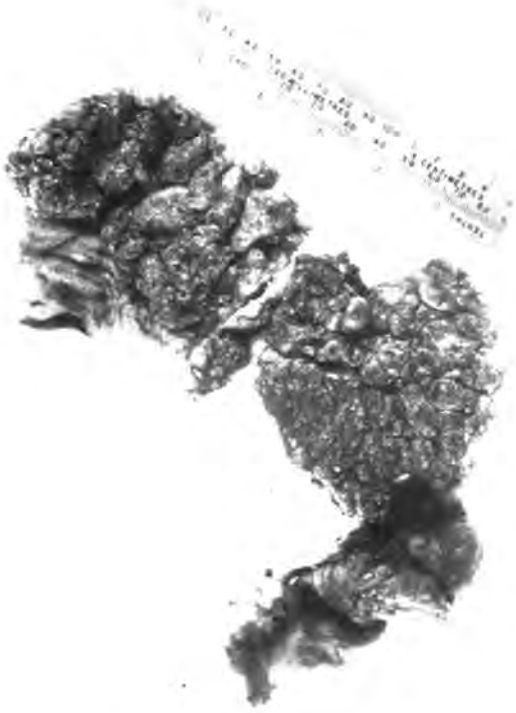
The specimen of lower ileum, measuring about 12 inches long, was covered by grape-like clusters of gas cysts of varying sizes with thin fairly transparent walls. and on cutting, they appeared smooth-walled and empty. (Photographs 1 & 2). Histologically, there were numerous subserosal cysts lined by flattened cells and some of the cysts were communicating with one another. The mucosa and submucosa of the ileum were normal. Thus histology confirmed the gross diagnosis of Pneumatosis Intestinalis.

DISCUSSION

Incidence

The occurrence of gas in linear or cyst-like forms within the subserosal, the submucosal or both layers of the intestinal wall, is termed Pneumatosis Intestinalis. Since Duverney's (1648) original description of this condition, many reports have appeared from time to time in the literature. Hunter (1728), Bang (1876), Lerner and Gazin (1946), Burt (1949), Marshak et al (1952), Greese (1954), Sedgwick and Ruddell (1954), Mathews (1955), Witowski et al (1955), Kutty & Unni (1965), Moore (1968), Dodd (1968). Though the exact incidence of this condition has not yet been ascertained, it appears that by 1965 more than 289 cases had been reported in the world literature. It is likely that in many patients with vague gastro-intestinal symptoms, the diagnosis is often overlooked, and in the event of secondary obstructive complications, the condition is often missed. This condition being exceptionally rare, this case is deemed worthy of record. And this will stress that Pneumatosis Intestinalis should be entertained in the differential diagnosis of chronic gastro-intestinal diseases and acute abdominal conditions.

Majority of the reported cases have been in adults; however, the condition has also been described in infants, but this is to be considered acquired rather



Photograph 1

Photograph 2

than congenital. Paris (1955), in a review of a series of affected infants, noted that the gas cysts were invariably associated with acute gastro-enteritis.

Pathology and Aetiopathogenesis

It is known that there are two types of Pneumatosis Intestinalis. (1) Primary Pneumatosis Intestinalis which develops spontaneously, and (2) Secondary Pneumatosis Intestinalis related to pathologic processes, such as peptic ulceration, neoplasms and emphysema. The gross appearance is that of gas cysts in the submucous or subserous layers of the intestine and though the site of occurrence is commonly the colon and the small intestine, rectal involvement has been reported (Griffiths 1955). In the colon cysts are located predominantly in the serosa, but the two may be combined in the same patient. The cysts may occur singly or in clusters not communicating with the intestinal lumen.

Though no conclusive evidence has so far been adduced regarding the etiology and pathogenesis of this condition, many theories, some factual and some conjectural, have been postulated, from which it appears that Pneumatosis Intestinalis is not *per se* a disease entity but the result of a variety of conditions. The mechanical theory postulates a breach such as in peptic ulceration, enteritis and neoplasm in the mucosal surface of the intestine, allowing for abnormal entrance of gases and air which permeate the nearby lymphatics. Increased intra-luminal pressure of doubtful origin augments the gaseous influx with resultant distension of the gas-filled lymphatics. Occurrence after such procedures like sigmoidoscopy and polypectomy, as suggested by Marshak et al (1956) should be reflected in a far greater incidence of the condition than that actually observed. This not being the case, such procedures cannot be incriminated so readily. The mechanical theory finds support in the frequent association of the condition with peptic ulcer. Nitch (1924), in a review of 85 cases, reported peptic ulceration in 50% of cases and of these, 80% had stenosis, while many of the remainder had mucosal lesions. Koss (1952) reported 58% associated with stenotic lesions of the pylorus.

Microphotograph 1

Ischemic necrosis of the mucosa as a consequence of chronic distention is thought to be another cause of intramural gas (Rigler & Pogue 1965). Altered permeability of the mucosa as a result of nutritional deficiency or nearby intestinal disease, is suggested as another mechanism in the occurrence of intramural gas. Pneumatosis Intestinalis of the swine has been produced experimentally by feeding them purely on polished rice. It appears more reasonable to combine the mechanical and altered mucosal permeability theories to explain the presence of abnormal intramural gas. Though Sauser-Hall (1940) suggested that gas cysts arose secondary to intestinal obstruction, the reverse is generally accepted. Masson (1920) regarded intra-mural gas a derivative of chemical interaction of chyle with the acid products of intestinal fermentation in the lymphatics. Alford (1956) attributed the symptoms of diarrhoea, flatulence and abdominal distention to excessive intestinal fermentation resulting in increased lactic acid and gas-forming bacteria. A defective absorption of these gases by the blood stream was thought to result in retention of gases within the intestinal wall, leading to the formation of giant cells and connective tissue which encapsulates the gas to form cysts. The neoplastic theory, like the infective theory, has been discarded for want of substantiative evidence.

It has recently been postulated that intra-mural gas cysts are produced as a result of rupture of emphysematous bullae and conditions of raised intra-thoracic pressure causing retrograde diffusion and distension of lymphatics of the gut-wall. The significant occurrence of allergic respiratory complaints in 15 out of 16 cases reported by Doub & Shea (1960) and others, lists chronic respiratory diseases as another possible etiological factor in the development of intestinal gas cysts. It is of interest that Pneumatosis Intestinalis has been described in patients with intestinal scleroderma Seaman et al (1966), Meihoff et al (1968) and Atlas (1968), but whether this association is merely fortuitous or has a causal relationship is not known.

Pre-operative diagnosis

Since there are no symptoms peculiar to this condition and when present are referable to any number of intestinal conditions, the diagnosis is difficult. Somerville (1951), while noting aerophagy in cases of Pneumatosis Intestinalis, conceded that symptoms were often those of the underlying disease. Many cases, however, are asymptomatic if unassociated with complications. Some are discovered incidentally at laparotomy for some other condition, or at post mortem. While sometimes sigmoidoscopy is sufficient to diagnose cysts of the descending colon, radiographic studies are the best diagnostic aid available. Plain X-rays of the chest and abdomen reveal lesions of the hepatic and splenic flexure as small gas shadows adjacent to or superimposed on the air in the colon; sometimes in abdominal films in the erect position, the cysts appear as grape-like clusters of air shadows, the walls of the intestine being delineated by a double-contoured line. (Paris 1955). Evidence of pneumoperitoneum, in the absence of bowel perforation may be pathognomonic of gas cysts. In the absence of obstruction, films after a barium enema reveal gas cysts as discrete cystic translucent areas indenting the intestinal wall as a series of filling defects of varying sizes. Sometimes the barium obliterates the cysts and characteristic findings are seen in post-evacuation films as cysts or as linear radiolucent stripes along the intestinal margin.

Progress and Termination

From the case reports of Jones (1948), Griffiths (1955) and Moore (1968), there is some evidence to suggest that Pneumatosis Intestinalis is insidious in onset. Though spontaneous disappearance of the cysts is possible as suggested by some – Koss (1952), Doub & Shea (1960) however, reported one case where there was complete resolution within 5 years. Untreated, the cysts may increase in number, causing various types of intestinal obstruction and pneumoperitoneum. Griffiths (1955), reported a case with severe obstructive symptoms in whom resection of the affected bowel segment was followed by an asymptomatic and satisfactory follow-up period, indicating that Pneumatosis Intestinalis is probably self-limiting.

SUMMARY

Pneumatosis Intestinalis is a rare condition characterised by the presence of gas-filled cysts in the submucosal, the subserosal or both the layers of the intestine. The paucity of such reports in the literature prompted us to place this on record and it is felt that this should be considered in the differential diagnosis of chronic or acute gastro-intestinal diseases. A brief review of the literature, touching upon certain important aspects such as pathogenesis and pre-operative diagnosis, has been made.

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