International Journal of Research in Medical Sciences
Agale SV et al. Int J Res Med Sci. 2015 Sep;3(9):xxx-xxx
www.msjonline.org

pISSN 2320-6071 | eISSN 2320-6012

DOI: http://dx.doi.org/10.18203/2320-6012.ijrms20150001

Case Report

Pneumatosis intestinalis in an adult: a report of an unusual case

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Received: 15 July 2015
Accepted: 11 August 2015

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ABSTRACT

Pneumatosis intestinalis (PI) is a rare condition affecting 0.03-0.37% of general population. This entity has varied presentation which ranges from asymptomatic to surgical emergency in the form of acute abdomen. The acute abdomen presentation can be bowel necrosis, perforation or persistent bowel obstruction. We present a case of PI of small intestine in an adult patient admitted in emergency department who presented with acute abdomen and pneumo-peritoneum.

Keywords: Pneumatosis (cystoides) intestinalis, Pneumatosis coli, Emphysematous gastroenteritis

INTRODUCTION

Pneumatosis intestinalis (PI) of the small bowel is a rare disease characterized by presence of gaseous cysts in the bowel wall.[1-5] PI was first described by Du Vermnoy in 1730 in autopsy findings.[3] The incidence of 0.03% has gone up to 0.37% due to accurate diagnosis on computerized tomography scan.[3,4] PI can be classified as primary (idiopathic) and secondary based on the absence or presence of etiological factors.[3-5]

Primary PI accounts for 15% of cases and commonly involves the sub-mucosal layer of colon, whereas secondary PI accounts for 85% of cases which involves the sub-serosal layer of small intestines, and it is seen in association with necrotic, non-necrotic gastro-intestinal or pulmonary diseases.[3,4] The secondary PI was described by Koss in 1952 after analyzing 213 pathological specimens.[5]

We present a case of pneumatosis intestinalis of small intestine with acute abdomen and pneumo-peritoneum in an adult patient admitted in emergency department.

CASE REPORT

A 45-year-old male who was a non-smoker and non-alcoholic presented in the emergency with acute abdomen. He also had complaints of recurrent abdominal pain and vomiting since last seven months. The abdominal pain was crampy and diffuses with no clear localization in the abdomen and had no clear relationship with meals or evacuation. Bowel habit was not characterized by diarrhoea but by constipation since five months. There was no history of bleeding per rectum. There was no history of diabetes mellitus, tuberculosis and bronchial asthma.

General physical examination did not reveal pallor, edema, lymphadenopathy or icterus. Blood pressure was-100/70 mm Hg, pulse rate-78 rpm, respiratory rate-13 cycles per minute. The baseline investigations revealed: Hb-15.7 g/dl, Total white-blood cell count-13,800 /cu mm, Differential leukocyte count: N-78%, L-20%, E-1%, M-1%, Random blood sugar level-109 mg/dl, Amylase-66 U/l, Serum electrolytes: sodium-137 mEq/l, potassium-1.4 mEq/l, Serum urea-29 mg%, Serum creatinine- 0.8 mg%, HIV-I & II and HBsAg-non reactive. On per abdominal examination, the abdomen was distended with increased bowel sounds with

tenderness over the umbilical area and no palpable mass. Plain X-ray abdomen showed features of intestinal obstruction. CT scan of abdomen showed multiple extra-luminal intra-peritoneal free air foci suggestive of pneumo-peritoneum. However no lead point or abnormal dilatation of proximal bowel loops was seen suggestive of transient intussusception (Figu[illegible]A and 1B). As the clinical diagnosis was acute intestinal obstruction, the patient underwent exploratory laparotomy.

On gross examination numerous grape-like variably-sized cysts filled with air (gas) were seen involving the serosa of the resected part of ileum. Cysts ranged in size from 0.5 to 1.5 cms in diameter (Figu[illegible]A). Crepitus could also be elicited while handling the specimen. Cut section showed no such cysts in the mucosa and submucosa but attenuated mucosa 3 cm and 6 cm away from one end of the resected ileal segment. The intestinal obstruction was seen due to reduced intestinal lumen secondary to thickened wall and sub-serosal cysts (Figu[illegible]B).

On microscopic examination, the sections studied revealed mucosa, sub-mucosa, muscle coat and serosa (Figure 3a).The lamina propria showed mixed inflammatory infiltrate comprised of lymphocytes, plasma cells, neutrophils and eosinophils extending up to the serosa. The muscularis propria showed vacuolar degeneration with gas dissecting through the muscularis propria separating the smooth muscle bundles. Serosa showed variable sized gas-filled cystic spaces lined by foreign-body type giant cells and mixed inflammatory infiltrate comprised of lymphocytes, plasma cells, neutrophils and eosinophils (Figure 3b). The diagnosis of pneumatosis intestinalis was rendered based on the gross and microscopic findings.

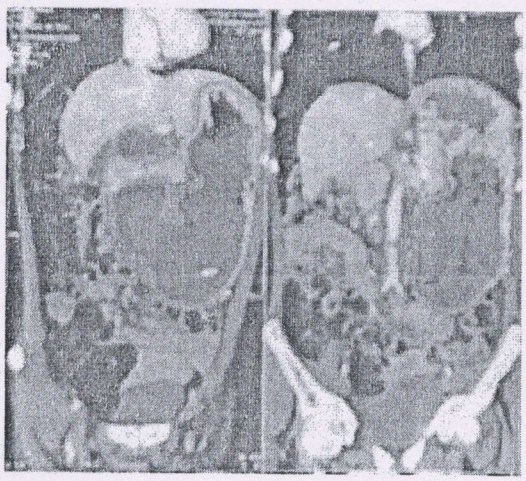

Fig 1a Fig 1b

Figu[illegible]A & 1B: CT scan of abdomen showing multiple extra-luminal intra-peritoneal free air foci suggestive of pneumo-peritoneum.

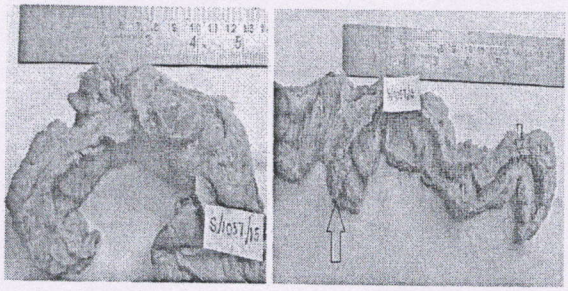

Fig 2a Fig 2b

Figure 2: (2a) Gross examination showed cysts ranging in size from 0.5 to 1.5 cm in diameter filled with air (gas) involving the serosa of the resected part of ileum. (2b) Cut section showed reduced intestinal lumen secondary to thickened wall and sub-serosal cysts (arrows).

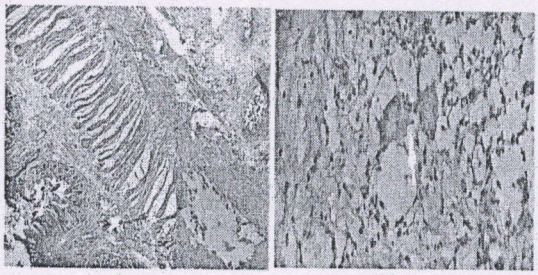

Fig 3a Fig 3b

Figure 3: (3a) Microscopic examination showed lamina propr[illegible], sub-serosal gas-filled cyst is seen (H&E, X40). Inset shows gas-filled cyst with arrow showing foreign-body type giant cells (H&E, X100) (3b) Sub serosa showed gas-filled cystic space lined by foreign-body type giant cells (H&E, X 400).

Figure 3[illegible]ows microscopic examination showed lamina propria infiltrated by mixed inflammatory infiltrate comprised of lymphocytes, plasma cells, neutrophils and eosinophils extending upto the serosa. The muscularis propria showed vacuolar degeneration with gas dissecting through the muscularis propria separating the smooth muscle bundles. A sub-serosal gas-filled cyst is seen (H&E, X40). Inset shows gas-filled cyst with arrow showing foreign-body type giant cells (H&E, X100). Figure 3b shows sub serosa showed gas-filled cystic space lined by foreign-body type giant cells (arrow) and mixed inflammatory infiltrate comprised of lymphocytes, plasma cells, neutrophils and eosinophils (H&E, X 400).

DISSCUSION

Pneumatosis intestinalis (PI) is an unusual condition which may involve any portion of the gastro-intestinal tract from stomach to rectum. Most reports of PI describe continuous portions of diseased bowel but there are few case reports of segmental involvement. The various studies have reported PI involving small intestine in 20%

to 51.6%, colon in 36% to 78% and both in 2 to 22%.[5-7] The synonyms for PI include pneumatosis intestinalis coli, pneumatosis cystoides intestinalis, pneumatosis intestinorium cystoided hominus and a simple term 'benign ~~intra~~-intramural gas.' In PI presence of intra-peritoneal free air and/ or retro-peritoneal air is very uncommon with few reported cases.[8]

The etiopathogenesis of PI is uncertain and three main theories have been suggested [4]:

1. *Mechanical theory:* The gas in the intestine is pushed to the mucosal defect into lymphatic channels and is then distributed distally by peristalsis. This may happen due to trauma, surgery, bowel obstruction or colonoscopy.
2. *Bacterial theory:* The sub-mucosal localization of *Clostridia* and *E. coli* leads to production of gas which is retained by sub-mucosa and lymphatic channels.
3. *Pulmonary theory:* The gas freed by the rupture of alveoli travels through the mediastinum into the retro-peritoneal space and then comes to the peri-vascular spaces into the intestinal wall.

Recently added theories are:[2]

4. Chemical theory / nutritional deficiency theory:
5. PI associated with chemotherapy, hormonal therapy and connective tissue disorders.

The pathological findings in resected specimen of intestine also vary depending upon the location of the cysts. The sub-mucosal cysts which are seen in primary PI have bubble-like appearance and in secondary PI, the sub-serosal cysts are readily recognizable on external examination as band-like continuous lines.[1,9] Microscopically, the lesions of PI are characterized by presence of multiple air filled cysts which are devoid of an endothelial lining. The surrounding tissue usually shows mixed inflammatory infiltrate of lymphocytes, neutrophils, plasma cells, macrophages and foreign-body giant cells. In cases where the giant cells and inflammatory component is predominant, it has to be be differentiated from Crohn's disease, tuberculosis and other granulomatous inflammatory conditions of the intestine. The histological differential diagnoses of PI which should be considered are lymphangiectasia and pseudo-lipomatosis.[4]

The most common presenting symptoms of PI are diarrhea or constipation, bloody stools, abdominal pain, flatus or weight loss. The complications of PI include pneumo-peritoneum, volvulus, intestinal obstruction, intussusception, haemorrhage and perforation.[4]

Pneumo-peritoneum usually represents a ruptured serosal cyst which was seen in this case. Pneumo-peritoneum is evident in up to 10% of cases with small bowel pneumatosis intestinalis and 2% of those with large bowel pneumatosis.[10] Development of pneumo-peritoneum in absence of signs and symptoms of peritoneal irritation is pathognomonic feature of PI.

CONCLUSION

This case highlights a rare and potentially ominous complication of pneumo-peritoneum in pneumatosis intestinalis.

Funding: None
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Agale SV, Warpe BM, Valand A, Kumari G. Pneumatosis intestinalis in an adult: a report of an unusual case. Int J Res Med Sci 2015;3(9):xxx-xx.