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PNEUMATOSIS INTESTINALIS- WHETHER TO OPERATE OR NOT? A RARE CASE REPORT WITH REVIEW OF LITERATURE.

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ABSTRACT

Pneumatosis Intestinalis (PI), also referred to as Pneumatosis Cystoides Intestinalis, pneumatosis coli, and intestinal emphysema, is defined as the presence of extra-luminal bowel gas that is confined within the bowel wall. Patients with a radiographic diagnosis of this condition should have a thorough history and physical examination. Most of the diagnosed cases can be managed conservatively, and only with the knowledge of underlying disease and patient's presentation, surgery should be decided to avoid unnecessary complications. We present a case of Pneumatosis Intestinalis (PI) of the colon in the setting of Aplastic anemia, post bone marrow transplant on immunosuppressants and steroid therapy that was treated with medical management and the relevant review of literature.

KEYWORDS

INTRODUCTION

The incidence of PI is unknown; the frequency is reported to be highest in the sixth decade. Idiopathic and secondary forms of the disease can be distinguished. The primary PI is extremely rare, in most cases PI is due to an underlying disease or condition such as traumatic and mechanical, inflammatory and autoimmune disease, drug induced, immunosuppression, transplantation or neoplasm. [1] In the primary idiopathic form, multiple thin walled cysts develop in the submucosa and subserosa of the gut. This form has no associated symptoms in most cases, and the cysts may be found incidentally through radiography or endoscopy. The secondary form is associated with obstructive pulmonary disease, as well as with obstructive and necrotic gastrointestinal diseases. [2]

PI can involve any part of gastrointestinal tract out of which most commonly small bowel is involved in 42%, the colon in 36% or both in 22% of cases. [3]PI is an alarming radiological finding that usually prompts an emergent surgical consultation for concerns of bowel ischemia and impending bowel rupture. Complications are present in 3% of PI patients and include pneumoperitoneum, bowel obstruction, volvulus, intussusceptions, and haemorrhage. [4] Due to the risk of these emergent complications, suspected PI patients should be carefully evaluated for possible surgery. Some authors have suggested five criteria that will be in favour of bowel necrosis requiring surgery; history of acute abdomen and physical examination findings supporting the history, metabolic acidosis, elevated lactate and serum amylase, and presence of portal venous gas. [5] For symptomatic PI of mild to moderate severity, treatment of the underlying disease with administration of antibiotics, oxygen therapy, and elemental diet may be sufficient for PI resolution. Here we describe a 20 year old patient with PI in the setting of aplastic anemia on immunosuppressants and steroid therapy along with GI infections who was treated conservatively.

CASE REPORT

A 20 year old male presented with chief complaints of fever and loose stool since 3 days. On physical examination, general and systemic examination was normal except for minimal generalized tenderness was present in abdomen. Laboratory investigations of the patient were normal. A computed tomography (CT) scan of the abdomen and thorax showed pneumatosis intestinalis involving the entire large bowel with presence of retroperitoneal and mediastinal air pockets. (fig1) Patient had a past history of aplastic anemia for which he underwent for which he underwent full evaluation leading to bone marrow transplant and was on immunosuppressants and high dose steroids for long time due to transient rejection for the same. Based on the findings and significant history, patient was started on high dose steroids and intravenous antibiotics (metronidazole and azithromycin). Patient became symptomatically better and his fever was resolved. As his symptoms and signs were improving, he was treated with bowel rest, oxygen therapy and IV antibiotics, Steroids tapered. The symptoms of the patient resolved with medical management and he was discharged on oral prednisolone.

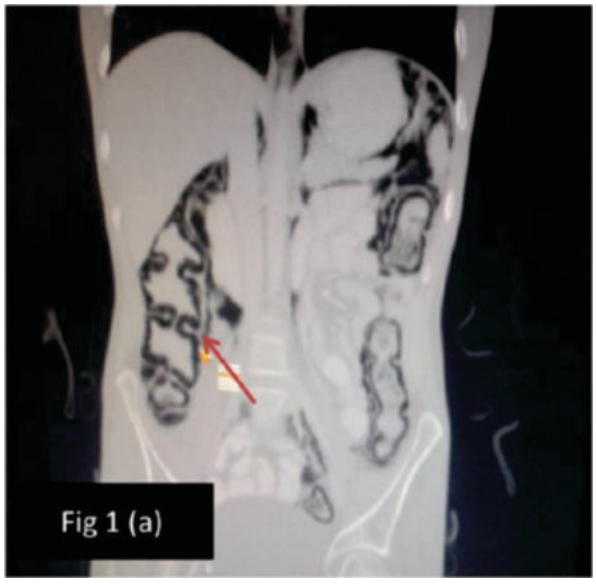

Figure 1(a) - Abdominal computer tomography of patient coronal view: pneumatosis intestinalis in ascending and descending colon

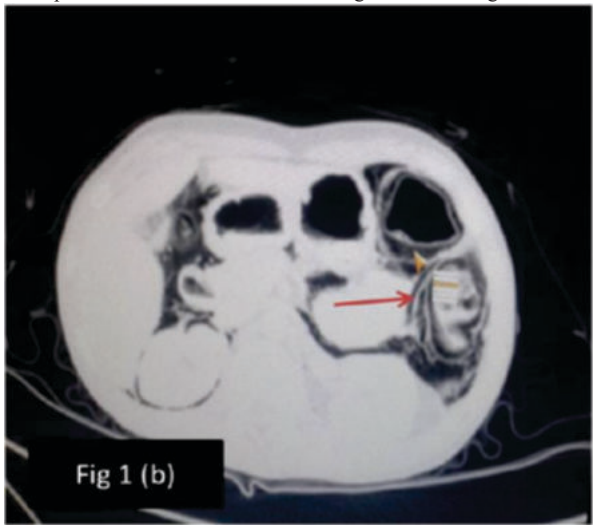

Fig 1 (b) - Also shows air in the walls of large intestine as well as retroperitoneum.

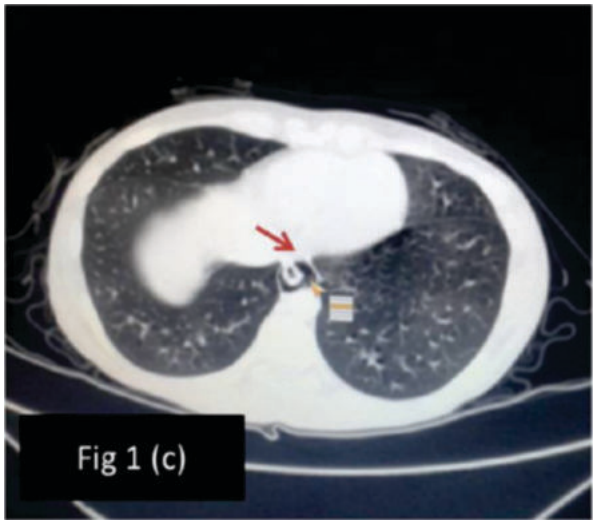

Fig 1(c) - Thoracic Tomography of the patient showing air pockets in mediastinum

DISCUSSION

PI is a rare disease usually caused by an underlying condition. The incidence is unknown due to asymptomatic course of the disease in most of cases. Primary PCI is extremely rare. Authors have divided the underlying conditions for secondary PI into six groups, traumatic and mechanical, inflammatory and autoimmune, infectious, pulmonary, drug induced and other. [6]The most common symptoms of primary PI are diarrhea or constipation, bloody stools, abdominal pain, flatus or weight loss which may be caused by mechanical effects of the cysts, but in many cases even large cysts remain asymptomatic; thus showing no correlation between clinical manifestation and the amount of intramural gas. [7] The location of PI within the gastrointestinal tract can dictate the associated symptoms. Patients with small intestine PI most frequently present with vomiting (60%), abdominal distension (59%), weight loss (55%), and abdominal discomfort (53%). Patients with colonic PI most commonly present with symptoms of diarrhea (56%), hematochezia (50%), abdominal discomfort (32%), and abdominal distension (28%). [8] While the pathophysiology has been debated, it appears to be related to the breakdown of the mucosal and immunological barrier of the intestines, especially in the setting of increased intraluminal pressure. In our patient with aplastic anemia, post bone marrow transplant immunosuppressants and steroid therapy likely contributed to PI etiology. Although the causality of high dose corticosteroids in PI is not established, postulated mechanisms suggests that immunosuppression of antimicrobial defences lead to intramural infection and impairment of the intestinal wall barrier [9] Our patient's prior upper GI endoscopies and prior colonoscopy with biopsy can be a contributing factor to developing PI as well, as these increases the risk of gas dissection into submucosa by compromising colonic mucosal integrity. [10]

Plain radiography, barium enema, ultrasound, abdominal CT and endoscopy can be helpful in diagnosing PCI, CT being the best method with the highest sensitivity. The radiological characteristics of PI on CT are intramural gas tracking parallel to the bowel mucosa as shown in fig1 (a) and fig (b). Many authors describe the value of ultrasound for diagnosing PI and portal venous air. [11]

Patients with radiographic diagnosis of PI should have a thorough history and physical examination and only along with the knowledge of the underlying disease the decision for an exploratory laparotomy can be made. PCI can be managed conservatively in most cases. Usually no treatment is necessary for 85% of patients who are asymptomatic. The resolution of gas collections has been reported after inhalation of 70% of oxygen and hyperbaric oxygen therapy. Surgical treatment may be needed in some cases but conservative treatment gives positive result in most cases. Signs of bowel ischemia, bowel necrosis, and bowel perforation resulting in intraperitoneal air indicate an emergent setting in which surgery may be appropriate. [12] Other complications of PI such as bowel obstruction may indicate surgical intervention or endoscopic puncture and sclerotherapy of the cysts. [13] However as in this case, patients with cardiovascular stability and minimal abdominal signs on abdominal examination may be closely observed and treated with bowel rest and antibiotics.

CONCLUSION

PI is a rare disease and suspicion of this disease process should be based on imaging and clinical finding. PI as a radiographic finding does not routinely require surgery. Because it represents such a wide spectrum of diseases, rapid evaluation of PI may be difficult. Although some general principles are certainly acceptable, there are more clinical scenarios to keep in mind. A thorough history and physical examination are obligatory since PI is mostly a consequence of an underlying disease. PI can be managed conservatively in most cases and surgery is required only in selected conditions.

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