

**PNEUMATOSIS CYSTOIDES INTESTINALIS: A CASE REPORT****Dr. Mukesh Kumar<sup>1</sup>, Dr. Pawan Sharma<sup>2</sup> and Dr. Ankit Panwar<sup>3\*</sup>**<sup>1</sup>MS General Surgery, Medical Officer, Regional Hospital Bilaspur (HP).<sup>2</sup>MS General Surgery, Medical Officer, Civil Hospital Theog (HP).<sup>3</sup>MS General Surgery, Medical Officer, Civil Hospital Rajgarh (HP).**\*Corresponding Author: Dr. Ankit Panwar**

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**ABSTRACT**

Pneumatosis cystoides intestinalis (PCI) or pneumatosis intestinalis (PI) is defined as gas within the bowel wall. It is an uncommon condition. The etiology is not clear but it is associated with a variety of underlying diagnoses. PCI is usually found in the large bowel, but can less commonly occur in the small bowel. The clinical picture vary from benign conditions requiring only observation to life threatening conditions requiring surgical intervention. Early appreciation of the overall clinical picture is therefore paramount to enable the practitioners to distinguish between the benign and the fatal cases of PCI and enable precise decision making regarding its management. We present a case of pneumatosis intestinalis in a 90 years old female who posed a challenging diagnostic dilemma.

**INTRODUCTION**

Pneumatosis cystoides Intestinalis (PCI) refers to the presence of gas within the wall of the intestine. It is a rare condition with incidence of 0.03 %.<sup>[1]</sup> It is characterized by the presence of numerous gaseous cysts containing nitrogen, hydrogen and carbon dioxide in the intestinal submucosa and subserosa.<sup>[2]</sup> This is a finding in a number of conditions ranging from benign to life-threatening. The exact etiology of the disease is still unknown however PCI may arise in association with pulmonary disease, inflammatory bowel disease, diabetes, drugs (for example steroids and chemotherapeutic agents), collagen vascular diseases such as systemic sclerosis, colonoscopy, ileal surgery, ingestion of sorbitol or lactulose.

In 1998, Pear classified PI pathogenically into four types: bowel necrosis, mucosal disruption, increased mucosal permeability and pulmonary disease.<sup>[3]</sup> Although PI can be seen on abdominal radiographs, CT is the most sensitive imaging.<sup>[4]</sup> The management of PI is not well documented. Operative intervention had been considered the mainstay of management.<sup>[5]</sup> Recently there has been a trend to a more conservative approach, as patients will often recover with non-surgical management.<sup>[6]</sup>

As there is a variety of presentations of PCI, the clinicians should interpret the clinical picture and the radiographic findings in order to ensure a correct diagnosis and to guide suitable management. The early recognition of the overall clinical picture is most important for decision making, with focus placed on key clinical features to efficiently distinguish between life threatening and non urgent causes of PCI.

**CASE REPORT**

75 years of old female from Kinnaur district of Himachal Pradesh presented with complaints of pain in her upper abdomen and distension of abdomen for three days. There was no history of loose stools, vomiting, constipation, fever, trauma, abdominal surgery, endoscopy or malignancy.

General physical examination was normal. The respiratory and cardiovascular systems were also within normal limits. On examination of the abdomen, there was abdominal distension. There was tenderness on palpation and slight rigidity, however there was no guarding or rebound tenderness. The tympanic note was present all over. On investigation routine blood investigations were normal. Ultrasonography revealed pneumoperitoneum. Chest x-ray (Fig 1) revealed gas under the right dome of diaphragm. Abdominal x-ray erect and supine (Fig 2&3) revealed no air fluid levels but gas under diaphragm on right side. CT scan (Fig 4) revealed air foci in perihepatic, perisplenic space and in the vicinity of duodenum suggestive of pneumoperitoneum.

Diagnosis of peritonitis due to hollow viscus perforation was kept. Exploratory laparotomy with assesment of intrabdominal organs was done. On opening the abdomen there was no gush of air or free fluid. Small intestine was dilated and there were multiple diverticulae present in antimesentric border of small intestine suggestive of pneumatosis cystoides intestinalis (Fig 5&6). Biopsy was taken from small intestine and abdomen was closed in layers. Patient was managed in the post operative period with bowel rest, intravenous

fluids and antibiotics. Post operative period was uneventful. Patient was discharged on 7<sup>th</sup> post operative

day and was advised regular follow-up.

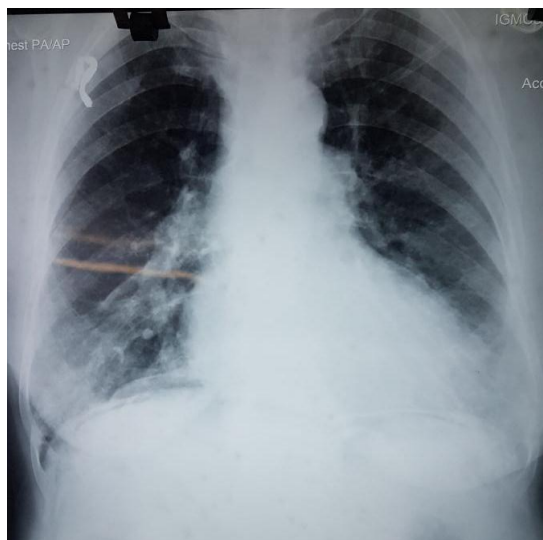


Fig 1.

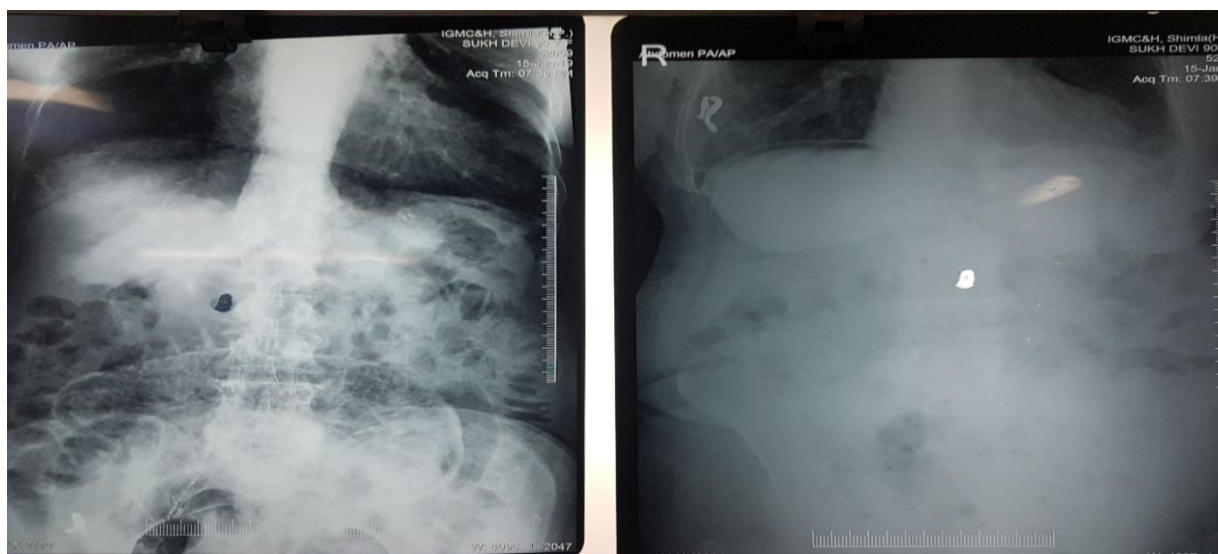


Fig 2 & 3

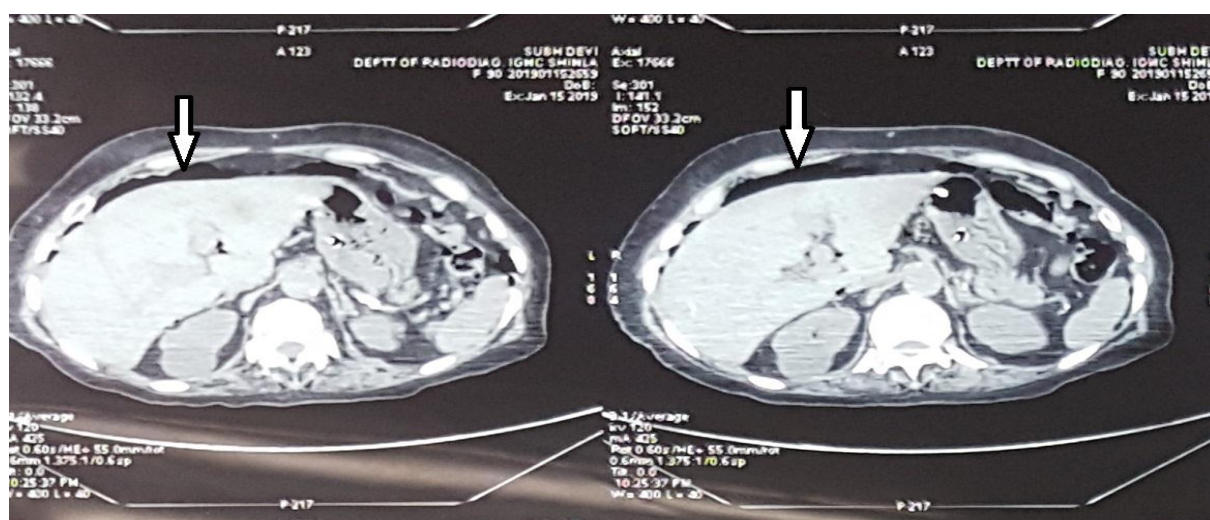


Fig 4

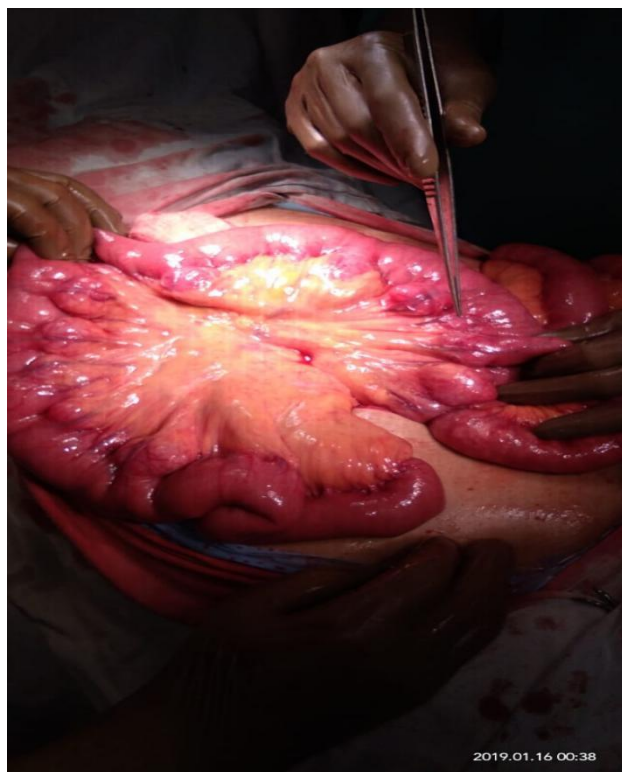


Fig. 5.

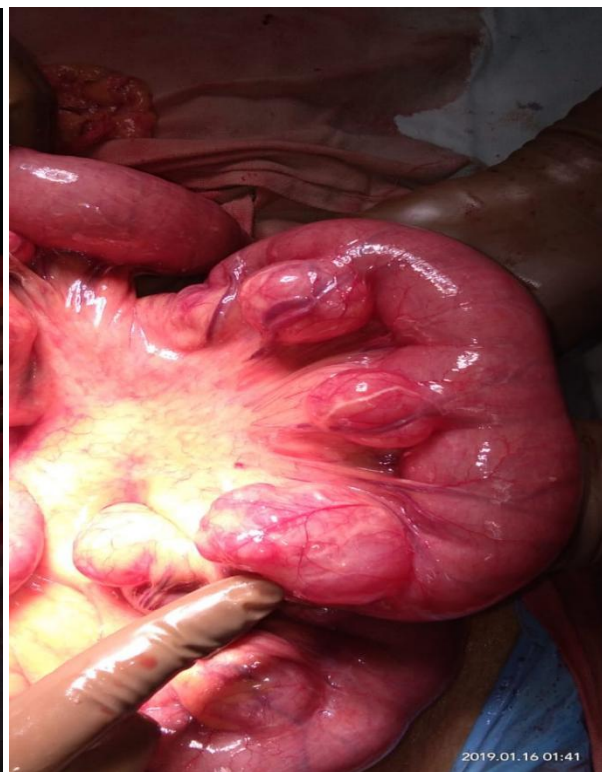


Fig. 6.

## DISCUSSION

PI is an unusual condition caused by a wide range of underlying pathophysiological processes varying from benign to life-threatening conditions. It affects the large intestine in 46% of cases and small intestine in 27% of cases. The incidence of both the colon and small intestine combined is only 7%.<sup>[7]</sup> There are two theories of development of PI. The first is the mechanical theory according to which there is diffusion of gas across the mural portion of the bowel, precipitated by inflammation or necrosis or direct gas diffusion across an intact mucosal membrane due to increased transabdominal pressure. The second theory is the origin of the gas from bacterial overgrowth and invasion of the bowel wall leading to PI.<sup>[4,8]</sup> The mechanical theory explains the association of PI with trauma, surgery, endoscopy and bowel obstruction. The infectious theory postulates that PI results from cysts formed by gas producing organisms. The infectious theory is supported by the evidence of elevated hydrogen content in the intraluminal cysts which is a byproduct of bacterial metabolism.<sup>[4,8]</sup>

It is hard to delineate the aetiology of PCI but is important to establish because the overall mortality is estimated to be 20% to 25%.<sup>[4]</sup> PI does not have a definite clinical presentation, patients may be asymptomatic or sometimes the patient may be sick depending on the underlying cause, with a mortality rate that may reach 75%.

Two third of the patients of PI may have characteristic radiological changes on X-ray. However, one-third of the

patients do not have a suggestive X-ray and require a CT scan or magnetic resonance imaging.<sup>[8]</sup> The patterns of pneumatosis in a CT scan such as linear, bubble, curvilinear gas collections localised or diffuse are useful to differentiate benign and clinically serious case of PI.<sup>[8]</sup> CT scan also helps to delineate the associated findings like bowel wall thickening, altered contrast mucosal enhancement, dilated bowel, soft tissue stranding, ascites, and the presence of portal air.

Treatment options include bowel rest, antibiotics and surgery. Antibiotics including metronidazole and quinolones can inhibit intestinal bacterial infection. Now recently hyperbaric oxygen therapy has emerged as an important aspect of the management. The rationale of oxygen therapy is that oxygen therapy increases the partial pressure of oxygen in the blood which increases the pressure gradient of the gas in the cysts. So the cysts release gases contained within them and refill with oxygen which is then metabolised leading to resolution. Oxygen therapy can be made through humidified oxygen administered by Venturi mask (6 L/min) or nasal cannula (4 L/min).<sup>[8]</sup> Endoscopic fine needle aspiration contributes to the diagnosis and treatment of PCI, by puncturing the cyst to exhaust gas.<sup>[9]</sup> PCI induced intestinal obstruction can be treated by the high-frequency endoscopic resection of the cyst wall, and cyst collapse after gas discharge.<sup>[9]</sup>

PI is a challenging condition to identify and manage. Although this lady initially presented with symptoms suggestive of bowel pathology, she was clinically stable. However, her CT scan was indicative of bowel



perforation, which heavily influenced the decision to operate. A laparotomy was performed which could have been avoided. Many authors advocate the use of laparoscopic approach over laparotomy when the patient is stable.<sup>[10]</sup> This is a more diagnostic and less invasive procedure. As the patients present in extremis and have multiple co-morbidities, there is an extremely high risk of peri-operative mortality. So the treatment should be tailored to the clinical symptoms, radiologic and endoscopic manifestations to avoid unnecessary surgery.

## CONCLUSION

Pneumatosis intestinalis is uncommon condition manifesting as multiple gas filled cyst of GIT. Overall incidence of pneumatosis intestinalis in general population is 0.03%. Cyst can occur anywhere in GIT from esophagus to rectum. Spontaneous rupture give rise to pneumoperitoneum. It should be kept in mind to avoid negative laparotomy.

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