PNEUMATOSIS INTESTINALIS - A RARE CLINICAL FINDING REQUIRING MULTIMODALITY TREATMENT

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Received: 11 August 2018 / Accepted 26 September 2018

Abstract

This is a case report of Pneumatosis intestinalis (PI), which has traditionally been associated with immediate operative intervention and a high mortality rate. We present a case of ulcerative colitis that developed Pneumatosis intestinalis and was managed at our hospital. A 29 year old male with known history of ulcerative colitis presented with an acute exacerbation. A week into his treatment, PI developed. Emergency total colectomy with end ileostomy was performed. Postoperatively, he remained critically ill and expired during same hospitalization after 45 days.

Key Words: Pneumatosis intestinalis, Ulcerative Colitis, Adalimumab

Introduction:

Pneumatosis intestinalis (PI), defined as gas within the bowel wall, is an uncommon radiographic sign which represents a wide spectrum of diseases and a variety of underlying diagnoses [1]. Because its etiology can vary greatly, management of PI ranges from surgical intervention to outpatient observation [2-4]. Since PI is infrequently encountered, clinicians may be unfamiliar with its diagnosis and management. We present a case of ulcerative colitis that developed PI and was managed at our hospital.

Case Report:

A 29 year old male presented to the gastrointestinal clinic with 10 weeks history of diarrhoea, weight loss and fever. During his workup to establish diagnosis endoscopic biopsies were taken from all parts of colon which revealed histopathological diagnosis of inflammatory bowel disease. He was started on Prednisolone 45 mg and Mesalazine 1600 mg BD. Three months later, he presented with acute exacerbation of ulcerative colitis with history of 20 to 22 episodes of watery stools per daily, intermittent bloody diarrhea and significant weight loss. He was started on medical management including intravenous fluids, antibiotics and total parenteral nutrition. His medications were switched to sulphasalazine, Azathioprine, and Adalimumab.

After one week of conservative treatment, his condition started deteriorating and he developed hypotension, tachycardia, major electrolyte abnormalities along with altered state of consciousness. He also developed extrapyramidal involuntary movements and his Glasgow coma scale score declined to 8/15. The patient was transferred to the intensive care unit and required endotracheal intubation for his deteriorating condition.

A Magnetic resonance imaging (MRI) scan of the brain reported non communicating moderate hydrocephalus with possible obstruction at the level of aqueduct. This
was managed by urgent placement of an external ventricular drain.

Patient remained critically ill requiring ventilatory and vasopressor support. Four days later, He was noted to have developed subcutaneous emphysema along the chest and anterior abdominal wall. A contrast enhanced computed tomography (CT) scan of the abdomen was performed that showed extensive subcutaneous surgical emphysema along the anterior and posterior chest walls extending up to neck and bilateral upper limbs superiorly and lateral abdominal walls inferiorly. There was gross pneumomediastinum with air extending down to retroperitoneum with free air in retro pancreatic area. In addition, small bilateral pneumothoraces were noted along with bilateral cavitatory lesions with internal mural nodule concerning for fungal infection. This was managed with bilateral chest drain insertion and empiric antifungal therapy with Variconazole.

He showed some clinical improvement but could not be weaned off the ventilatory support due to persistent respiratory distress. A Ventriculo-Peritoneal (VP) shunt was performed for his persistent hydrocephalus. His blood cultures grew gram negative bacteria.

A repeat contrast enhanced CT scan of the abdomen demonstrated new development of pneumatosus intestinalis involving the ascending colon, extending to the hepatic flexure (Figure1). Immediate emergency total colectomy with end ileostomy was performed. He remained on ventilator support post operatively. Histopathology report of the specimen was consistent with ulcerative colitis.

Patient stayed in ICU for 45 days post operatively. He was started on enteral feed initially but had to be stopped because of minimal stoma output and high Nasogastric output. His GCS dropped to 2/10(T). There was no radiological evidence of definite mechanical obstruction. Patient remained on ventilator for 77 days and expired on 100th day of admission due to a cardiac arrest. Autopsy was refused by the family and was not performed.

**DISCUSSION:**

Pneumatosis’ refers to presence of gas or air at an abnormal location in the body[1]. PI also refers to presence of air or gas in the wall of the small or large intestine [1-5]. It is also known as Intramural gas, Pneumatosis Cystoides Intestinalis, etc. PI is a clinical sign and not a disease, usually first diagnosed on radiographic study. Interpretation of this sign must be made relative to the clinical condition of the patient and other associated signs and symptoms as well as laboratory findings [6] in order to decipher a benign disease from a life threatening one.

PI is a relatively rare finding having an overall incidence of 0.03% among the general population based on autopsy findings³. However, recently the overall
incidence has increased to 0.3% due to extensive use of CT scan in recent years. In past literature, PI has been described in association with a number of GIT disorders, including infection, obstruction, or ischemia. Other than GIT related issues, it has also been reported with chronic obstructive pulmonary disease, connective tissue disorders, organ transplantation, leukemia and various states of immunodeficiency[5-7].

PI has been classified into primary and secondary types. Primary PI is when the cause of PI is not known, i.e. idiopathic; whereas secondary PI is termed when there is an underlying disease that may have led to this condition. Primary PI accounts for only 15% of the cases whereas, secondary PI, being more common, accounts for 85% of cases [6]. Secondary PI can be associated with a number of diseases, including Inflammatory Bowel Disease, however the incidence is low.

In our case the cause of PI could be multifactorial. PI was preceded by an episode of bilateral pneumothorax and subcutaneous emphysema which could make it a likely cause. fungal infection is one possibility also the baseline pathology was ulcerative colitis which could still be the primary cause. Also a few cases have been reported in literature associated with UC.[9] But to say with certainty what caused the PI in this patient is not an easy task.

Our patient presented with 10 weeks history of diarrhea, weight loss and fever. If the literature review is taken Patients with PI mostly present with a history of diarrhea, abdominal pain and discomfort, abdominal distention, bloody stools, constipation, loss of weight and tenesmus. In some cases, PI may be an incidental finding as is the case in benign causes of PI like PI associated with pulmonary disease where no particular treatment is needed [7]. However, some patients may present in a critical condition and these patients are mostly amongst those who have PI due to a life threatening underlying disease, such as mesenteric ischemia.

On CT scan our patient was diagnosed as having PI. Amongst the cases reported in literature CT scan has remained a sensitive modality in diagnosing the PI [6,10]

Another possible cause of PI could be adalimumab. Patient was on this drug for a week when he developed a hydrocephalus followed by pneumatosis intestinalis. Although not reported with adalimumab but other molecular targeted therapy has been shown to be associated with PI [10]. We can’t for sure establish a link between the two but its role cannot be ruled out.

CONCLUSION:

PI is a rare entity. It hasn’t been labelled as a disease but a sign. Careful observation followed by aggressive intervention for a patient who deteriorates is the main management strategy. A multidisciplinary approach with careful patient monitoring and a low threshold for surgery can help reduce mortality from this complication.

References:


