Pneumatosis Cystoids Intestinalis in an Immunosuppressed Patient

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Citation

Abstract
We report the case of a 59 year old woman who presented to the emergency room with a four day history of abdominal pain worsening over the last 24 hours. She had previously been diagnosed with Alport’s syndrome and had undergone an unrelated donor renal transplant in August 2001. She was on several immunosuppressants. There was some evidence of peritoneal signs on physical exam. Plain film X-ray and CT scan revealed marked pneumatosis interstinalis of the right and transverse colon. The patient underwent a laparotomy. The laparotomy showed crackly air throughout the serosa of the right and transverse colon, which was also pale in appearance. A subtotal colectomy and ileostomy was performed to correct the PCI and perforation. The patient had a successful recovery post-surgery. The success of surgical intervention in our case demonstrates the potential role for surgery in a patient with suspected PCI, with evidence of perforation.

INTRODUCTION
Pneumatosis intestinalis or pneumatosis cystoides intestinalis (PCI) is defined by the presence of submucosal and subserosal gas filled cysts within the large or small bowel. This condition often occurs in patients with ischemia or inflammatory disease of the bowel or in conditions such as chronic obstructive pulmonary disease, connective tissue disorders (such as Alport’s syndrome), celiac disease, leukemia, amyloidosis, and acquired immunodeficiency syndrome. It can also be found in association with organ transplantation and immunosuppression. PCI is a relatively rare condition [12] that can often be successfully treated with conservative management including bowel rest, oxygen therapy, parenteral nutrition and broad spectrum antibiotics [23].

Here we report a case of a patient who presented with PCI and peritoneal signs. The patient had a history of Alport’s syndrome and was on several immunosuppressants due to an unrelated renal transplant secondary to Alport’s syndrome. To correct this condition the patient underwent surgical intervention. In the presence of secondary complications such as peritonitis, ischemia or perforation, surgical correction is often indicated; however this is debated and infrequently reported in the literature [386].

CASE REPORT

A 59 year old woman presented with a four day history of abdominal pain worsening over the last 24 hours. She had previously been diagnosed with Alport’s syndrome and had undergone an unrelated donor renal transplant in August 2001. She is currently on several immunosuppressants. There was some evidence of peritoneal signs on physical exam. Plain film X-ray and CT scan revealed marked pneumatosis interstinalis of the right and transverse colon (Figure 1, 2).
Figure 1
Figure 1: CT scan of the transverse colon demonstrating air in the wall of the bowel

Figure 2
Figure 2: CT scan of right colon showing air in the wall of the colon

The patient was then brought for a laparotomy. The laparotomy showed crackly air throughout the serosa and somewhat pale appearing serosa throughout the right and transverse colon. A subtotal colectomy and ileostomy was performed for pneumatosis intestinalis and peritoneal signs.

The gross pathological specimen showed a dilated bowel wall with an edematous and cystic appearance of the cecum and ascending colon.

Figure 3
Figure 3: Gross image of the bowel shows cystic lesions.

Histological sections of the serosa, submucosa and muscularis propria showed cystically dilated spaces lined by unremarkable flattened endothelial like cells, devoid of red blood cells or proteinaceous fluid.
Figure 4
Figure 4: H & E 2 x of bowel wall shows cystic changes mainly in the submucosa.

Figure 5
Figure 5: H & E 10x of bowel wall shows multinucleated giant cells surrounding the cystic spaces.

Figure 6
Figure 6: Factor VIII x 20 shows negative staining of the flattened endothelial like lining of the cystic spaces.

Figure 7
Figure 7: CD31 x20 shows negative staining of the flattened endothelial lining of the cystic spaces.

These findings can be seen in both pneumatosis intestinalis and lymphangiomas. Immunostains factor V111 and CD31 were negative, ruling out lymphangioma as the cause (Figure 6, 7).
This patient is on long-term immunosuppression and therefore at risk for opportunistic infections. There were no fungal forms or CMV inclusion aiding in the exclusion of an opportunistic infection as a cause. A chronic inflammatory cell infiltrate is seen in the submucosa and muscularis propria with eosinophilic predominance. Multinucleated giant cells are seen surrounding the cystic spaces. A section of the serosal surface of the ascending colon shows infiltration with chronic inflammatory cells and prominent vascular congestion. These findings are supportive of the presence of perforation and the findings of peritoneal signs on physical exam. In some areas there is surface mucosal ulceration with preservation of the lower parts of the crypts, this may suggest an element of ischemic necrosis associated with pneumatosis intestinalis. The histological findings are typical of pneumatosis intestinalis. In addition, the presence of perforation explains the clinical decision to remove the bowel surgically instead of the usual conservative medical management.

**DISCUSSION**

Pneumatosis intestinalis or pneumatosis cystoides intestinalis (PCI) is a rare condition defined by intramural gas collection in the intestine. Predisposing factors for developing PCI include immunodeficiency, connective tissue disorders and bowel ischemia. In patients with PCI without evidence of bowel perforation conservative management is often the most successful course of action \cite{11}.

Many factors likely play a role in the etiology of PCI. The bacterial or infectious hypothesis suggests that gas forming bacteria, such as Clostridia, produce PCI by diminishing the integrity of the mucosa promoting the dissection of gas through tissue of the GIT \cite{10}. The mechanical theory suggests that gas would penetrate into the submucosal and subserosal layers of the bowel through mucosal lesions caused by ulcerations or ischemia of the bowel \cite{10}. Immunosuppression with corticosteroids has also been implicated in the causation of PCI, either by submucosal lymphoid depletion or by altering the mucous production \cite{10}. In this case we present a patient with a history of a connective tissue disorder (Alport’s syndrome) and who was on immunosuppressants for a previous renal transplant. When this patient presented, there was evidence of bowel perforation, through the presence of peritoneal signs and CT evidence. PCI can be a difficult diagnosis to make \cite{11}, however, previous papers report that the majority of patients with PCI are correctly diagnosed with clinical investigation, x-ray and CT scan \cite{11}. Given the presentation of PCI with peritonitis it was determined that this patient was in need of surgical intervention to repair the perforation. A subtotal colectomy and ileostomy was performed to correct the PCI and perforation. The patient had a successful recovery post-surgery.

Previous cases report the recovery of patients with PCI and pneumoperitoneum that responded well to conservative treatment. However in these cases peritonitis was not present \cite{12}. The success of surgical intervention in our case demonstrates the potential role for surgery in a patient with suspected PCI and with evidence of perforation on CT scan and peritoneal signs on physical exam.

**References**

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