

Necrotizing Enterocolitis With Hepatic Portal Venous Gas And Pneumatosis Intestinalis In A Patient With AIDS

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Citation

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Abstract

A 43-year-old man was admitted in our hospital because of severe *Pneumocystis jirovecii* pneumonia [formerly called *Pneumocystis Carinii* Pneumonia (PCP)]. During his stay in Intensive care unit, he developed vomiting, abdominal distension and absent bowel sounds. On abdominal radiograph, hepatic portal venous gas (HPVG) and pneumatosis intestinalis of the small and large intestine were present. HPVG caused by necrotizing enterocolitis was diagnosed, and an emergency laparotomy was thus performed that showed necrosis of the small and large bowel.

Bowel resection was done. The patient is alive as of two years after operation. The prognosis of necrotizing enterocolitis accompanied by HPVG and pneumatosis intestinalis is extremely poor. The presence of HPVG suggests the occurrence of a serious lesion in the abdominal cavity. Therefore, appropriate treatment should be performed immediately.

CASE REPORT

A 43 year old healthy Afro-American male presented to the emergency department with dry cough, worsening dyspnea and fever for last one week. The patient denied chest pain, headache, night sweat, weight loss, and had no previous history of gastrointestinal and genitourinary problems. Physical examination was remarkable for heart rate of 120/min, respirations 40/min, and saturating at 90% on room air. Auscultation of the chest revealed bilateral crackles in lung bases. Chest x-ray on admission showed bilateral air space disease. Admission labs were significant for serum lactic acid dehydrogenase of 541U/L and blood gases on room air showed PH 7.42, PaCO₂ 29mm Hg and PaO₂ 71mmHg. Rest of the labs including serum blood count, electrolytes, cardiac enzymes, serum lactate and liver biochemistry were all within normal limits. Patient was started on ceftriaxone and azithromycin intravenously for community acquired pneumonia.

On 2nd day of admission; he got intubated for acute hypoxic respiratory failure and was transferred to Intensive care unit. Diagnostic impression was *Pneumocystis carinii* pneumonia (PCP). Antibiotics were changed to Imipenem, Vancomycin, Ciprofloxacin, Trimethoprim/Sulfamethoxazole and intravenous methylprednisolone was also added. Additional tests including HIV testing, lymphocyte count, serum cryptococcal antigen, hepatitis profile, titers for

toxoplasmosis and cytomegalovirus (CMV), stool for *Clostridium difficile*, *Cryptosporidium* and CMV, blood cultures, tracheal aspirate for Acid fast bacilli, gram stain and cultures were obtained. He tested positive for HIV with CD4 cell counts of 04 only. Rest of the labs came back normal. Azithromycin was added in his regime for *Mycobacterium Avium intracellulare* (MAI) prophylaxis. On 10th day of admission; PCP pneumonia was confirmed by Broncho alveolar lavage growing PCP. He continued to do well but on the 14th day of admission, he developed high grade fever up to 103F, vomiting, and abdominal distension.

Bowel sounds were absent. A provisional diagnosis of bowel obstruction was made; plain chest and abdominal radiographs were obtained. Repeat blood tests revealed significant drop in platelets to 90k/mcL, lactate level of 6 mmol/L, bicarbonate 10 mmol/L, BUN 32 mg/dl and creatinine of 3.2mg/dl. Repeat blood cultures were obtained that came back negative.

Chest x-ray was negative for free air. Abdominal films showed linear lucencies overlying the liver and large amount of intramural gas within the large and small bowel. Findings were consistent with intestinalis pneumatosis with hepatic portal venous gas suggesting bowel ischemia. Patient was taken immediately to Operation room to perform life saving exploratory laparotomy.

Operative findings included necrotic small and large bowel from the mid Jejunum all the way to the ileocaecal valve, Ascending colon and Transverse colon up to splenic flexure. Resection of majority of small bowel and large bowel up to splenic flexure with jejunostomy was done. Pathology of specimen showed acute necrotizing enterocolitis, multifocal, with focal gangrenous bowel. Mesenteric vessels showed no evidence of primary vasculitis. These findings were consistent with an infectious enterocolitis in the absence of a primary vascular/ mesenteric disease, clinical hypovolemia and the extensive involvement of small bowel and colon.

After surgical intervention, gradual clinical and biochemical improvement was noted and patient was discharged home after acute rehabilitation.

DISCUSSION

Necrotizing enterocolitis (NEC) is an acute disease that primarily affects premature neonates of low birth weight. NEC in adults is relatively common in the developing countries and its etiology is multifactorial. Infectious agents, inflammatory mediators and circulatory disturbance have all been implicated in the etiology and pathogenesis of NEC. The common organisms implicated are bacteria like Klebsiella, E. coli, Enterobacter and Pseudomonas and viruses like Corona virus and Entero virus (1). The case described here illustrates the fact that advanced AIDS can be associated with severe necrotizing enterocolitis with out any other identifiable cause.

It is also suggested that reduced mesenteric blood flow leads to ischaemia, which in turn causes hypoxic cell damage and release of inflammatory mediators. The reperfusion of these tissues results in the release of oxygen free radicals that in turn cause loss of cellular integrity. The final common pathway in the development of NEC is believed to involve mediators like platelet-activating factor, endotoxin lipopolysaccharide, tumor necrosis factor and other cytokines, prostaglandins and leukotrienes. (1)

Pneumatosis intestinalis is pathognomonic of NEC. The classical triad of increasing thrombocytopenia, acidosis and hyponatraemia suggests severe NEC, and these are the patients, who are more likely to require surgical intervention, like our patient. Surgical intervention is necessary if there is intestinal necrosis or frank perforation or when there is clinical deterioration over 12-24 hours despite intensive medical support, as evidenced by persistent or worsening metabolic acidosis, increasing ventilatory requirement,

deteriorating hematological parameters and persistent thrombocytopenia (2).

Pneumatosis intestinalis was first described by DuVeroni in 1730 (3). The case described here illustrates the fact that intramural air should be perceived not as a diagnosis, but rather a radiological finding. This case also illustrates that clinical presentation, physical examination and laboratory results -- not radiographic findings alone -- should be considered when determining the cause and significance of intramural bowel gas.

The cystic variant of intramural gas is characterized by macroscopic submucosal cysts, ranging in size from a few millimetres to several centimetres in diameter. These protrude into the bowel lumen, creating characteristic indentations that can be seen radiographically and endoscopically (3, 4). The cystic variant of intramural gas is nonspecific but suggestive of a benign etiology, particularly if the clinical presentation supports this.

Linear pneumatosis carries a more ominous prognosis, as it is thought to represent the tracking of gas through compromised submucosa. This variant is identified by a streaking, linear or bubbly pattern that is oriented parallel to the bowel wall, plicae circulares or haustra. Although it can be associated with benign etiologies, linear intramural air suggests the need to rule out bowel ischemia or infarction, and mandates a search for portal venous gas -- best detected using CT or ultrasound.

Hepatic portal venous gas (HPVG) is an important prognostic feature, and if associated with bowel ischemia, mortality rates are as high as 83% (5). The pathogenesis of HPVG is not fully understood. Two theories, mechanical and bacterial, have been proposed. The mechanical theory proposes that gas dissects into the bowel wall from either the intestinal lumen or the lung (6).

Mucosal damage and bowel distention are important factors in this theory. Mucosal damage allows intraluminal gas to enter the venous system (6). The bacterial theory proposes that gas forming bacilli enter the submucosa through mucosal rents and produce gas within the intestinal wall and then enter into the portal vein (6).

In cases of ischemic bowel distention with mucosal damage, mucosal damage allows gas to enter the mesenteric vessels directly (7, 8). Bowel distention and ischemia can also produce minimal mucosal disruption that allows intraluminal

gas to become intravascular (9).

SUMMARY

Necrotizing enterocolitis in adults associated with hepatic portal vein gas and pneumatosis intestinalis is very rare and carries ominous prognosis. Presence of hepatic portal vein gas warrants immediate and aggressive management.

However, mortality is still very high despite aggressive treatment.

Pneumatosis intestinalis is a radiological or histological finding and is not a diagnosis in itself. As illustrated by the case presented, it could be a manifestation of ominous condition requiring aggressive treatment including

surgery and removal of necrotic tissue. Correct diagnosis and management is based on the results of clinical assessment, laboratory testing and imaging modalities. An understanding of the causes and associations of this finding will help physicians make appropriate and timely decisions about surgical, medical or conservative management.

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