

Pneumatosis cystoides intestinalis: a case report and review of the literature

Pnömatozis sistoides intestinalis: olgu sunumu ve literatürün gözden geçirilmesi

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ÖZET

Pnömatozis sistoides intestinalis ve portal venöz hava altta yatan hastalıkla ilişki olarak bazı vakalarda acil cerrahi gerekliliğinin radyolojik bir işarettir. Etiyolojisinde ve patogenezinde birçok faktör öne sürülmüştür. Portal venöz hava, klinik olarak daha önemlidir çünkü sıklıkla bağırsak iskemisi veya nekroz ile ilişkilidir ve kötü bir prognozu temsil eder. Bununla birlikte, prognoz, portal venöz hava veya pnömatozis intestinalisin değil, altta yatan hastalığın şiddetinden daha fazla etkilenir. Bağırsak iskemisi veya nekroz birincil neden olduğunda, acil ameliyat hastanın prognozunu iyileştirmek için çok önemlidir. Biz burada portal venöz hava ve pnömatozis sistoides intestinalis ile gelişen bir mezenterik iskemi olgusunu sunuyoruz.

Anahtar kelimeler: Pnömatozis sistoides intestinalis, bilgisayarlı tomografi, portal venöz hava

SUMMARY

Pneumatosis cystoides intestinalis and portal venous gas which is a serious underlying abdominal disease in some cases, is a radiological finding requiring urgent surgery. Many authors suggested various factors in the etiology and pathogenesis. Portal venous gas is more important because it is usually related with intestinal ischemia or necrosis. But prognosis is affected more by the severity of the underlying disease than by hepatic portal venous gas or pneumatosis intestinalis. Intestinal ischemia or necrosis is the first reason, urgent surgery is crucial to improve the prognosis of the patient. In this case we report mesenteric ischemia which is presenting with portal venous gas and pneumatosis intestinalis.

Keywords: Pneumatosis cystoides intestinalis, computed tomography, portal venous gas

INTRODUCTION

Many authors have recommended that pneumatosis cystoides intestinalis (PCI) is an infrequent situation delineated by entity of various cysts in the intestinal wall. PCI which is pathogenesis indeterminate, is a clinical sign not a disease (1). PCI and portal venous gas (PVG) are individualistic uncomfortable visual findings. They are related with severe mesenteric ischaemia and when the diseases occur with abdominal pain symptoms, the death rate is exaggerated to 72%. They can exist without pathology, either independently or together (2,3). We report a rare script where PCI and PVG were presented in chronic renal failure case.

CASE

A 59-year-old male admitted to emergency service with complaints of nausea, severe vomiting, abdominal pain and swelling that had begun 3 days prior. The patient was hemodynamically stable and had mild generalized abdominal pain but with a soft distended abdomen. His past medical history was chronic renal failure. On admission, his temperature was 37.6°C, his heart rate was 52 beats/min, his respiratory rate was 13 breaths/min and his blood pressure was 100/60 mmHg. The laboratory test results on admission were within the normal limits except white cell count was slightly elevated at : 11200/ml, with C-reactive protein raised to 48 mg/l (normal: 0-3mg/l), hemoglobin level (Hb: 10.6/dL) and normal platelet count. The blood biochemical test results were as follows: blood urea nitrogen (BUN): 137 mg/dl, creatinine: 6.52 mg/dl, the others were within normal limits. Chest X-ray was normal. PVG was revealed in ultrasonography (USG). On the computed tomography (CT) imaging of the whole abdomen intramural gas within the bowel was seen logical with PCI (Figure 1a) and with evidence of air in the intrahepatic portal system (Figure 1b).



Figure 1a: Computed tomography demonstrating pneumatosis intestinalis within the walls of the bowel

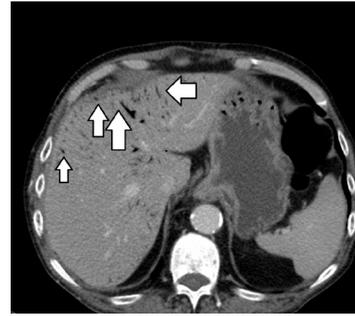


Figure 1b: White arrows show gas filling peripheral portal veins in the liver.

Emergency exploratory laparotomy was performed with a diagnosis of mesenteric ischemia. Perforation was not detected, consequently, an exploration of the abdomen was positive for bowel necrosis. There was a necrosis from splenic flexura to rectum in small intestine and colon. Ileostomy and colostomy was performed. The patient died fourth postoperative day due to sepsis.

DISCUSSION

PCI, also known as intramural gas, is located in the bowel wall. It is seen usually in those aged in males and 30–50 years (4). Many authors reported approximately 0.3% incidence of PCI inspired by progressively use of CT. In our case patient had a similar demographic characteristics. Primary and secondary is two types of disease. The primary type which is not unrelated to other accompanying diseases figure out approximately 15% of cases. 85% of the remaining cases is the secondary type (1,5). The most common form of PCI occurs according as acute gastro-intestinal ischaemia. Others are malignancy, chemotherapy, trauma, liver-failure (2,6). Our case was occurred due to mesenteric ischemia.

The clinical presentation of PCI is various, from asymptomatic to symptoms associated with life threatening complications like ischemia, perforation and peritonitis. Patients may present with abdominal pain and distension, diarrhoea or constipation. Clinically severe symptoms like bleeding or ileus may also present. It is assumed that PCI clinical findings should not be related to intramural gas or its location in the gastrointestinal tract, whereas its related to the underlying irregularity (7). When the PCI is suspected, simple procedures such as direct radiography and advanced diagnostic techniques such as USG and colonoscopy and CT can be desired. Since PCI is manifested by a wide range of clinical signs and symptoms, most cases are incidentally (8). Our patient

had a life threatening complications such as mesenteric ischemia with a symptoms of abdominal pain, and swelling and PVG was revealed in USG.

Many authors have reported that mortality rates vary between 0% and 75%, but mortality is immensely dependent on coexisting conditions such as bowel ischemia or inflammation. The patients who had PVG and PCI on CT scans, mortality rate rised approximately 72%. The presence of hepatic portal venous gas is likely related with a exaggerated mortality rate, because CT findings of hepatic PVG usually show the entity of mesenteric ischemia or infarct (6). The current case was PVG and PCI on CT scans and the patient died due to sepsis.

There is no specific treatment recommended for PCI asymptomatic patients identified radiologically. Treatment of PCI ranges from supportive care including nasogastric decompression, intestinal rest, broad spectrum antibiotics, to laparotomy. Surgery is indicated in patients with severe pain, rectal bleeding, fever, or evidence of ischemic bowel (9,10). "In our case emergency exploratory laparotomy was performed with a diagnosis of mesenteric ischemia.

In conclusion, PCI and PVG are infrequently seen and There was varied factors responsible in the etiology. PCI and PVG is not a surgical indication and the treatment is actually dependent on the underlying disease. The prognosis is related to the pathology itself and is not affected by its presence. Delayed therapy can cause serious morbidity and mortality.

REFERENCES

1. Morris MS, Gee AC, Cho SD, Limbaugh K, Underwood S, Ham B, et al. Management and outcome of pneumatosis intestinalis. *Am J Surg* 2008; 195(5): 679-83.
2. Wayne E, Ough M, Wu A, Liao J, Andresen K, Kuehn D, et al. Management algorithm for pneumatosis intestinalis and portal venous gas: treatment and outcome of 88 consecutive cases. *J Gastrointest Surg* 2010; 14(3): 437-48.
3. Hussain A, Mahmood H, El-Hasani S. Portal vein gas in emergency surgery. *World J Emerg Surg* 2008; 3(1): 21.
4. Bolukbas F. Bolukbas C. Pnomatozis sistoides intestinalis. *Güncel gastroenterol derg* 2004; 8: 182-5.
5. Hanna P, Kassir R, Tarek D, Bassile B, Saint-Eve P, Elias B. Pneumatosis cystoidis intestinalis presenting as bowel perforation, a rare entity. *Int J Surg Case Rep* 2016; 20: 7-9.
6. Wiesner W, Mortelé KJ, Glickman JN, Ji H, Ros PR. Pneumatosis intestinalis and portomesenteric venous gas in intestinal ischemia: correlation of CT findings with severity of ischemia and clinical outcome. *Am J Roentgenol* 2001; 177(6): 1319-23.
7. Vogel Y, Buchner NJ, Szpakowski M, Tannapfel A, Henning BF. Pneumatosis cystoides intestinalis of the ascending colon related to acarbose treatment: a case report. *J Med Case Rep* 2009; 3(1): 9216.
8. Polat C, Tokyol Ç, Sen S, Yazicioğlu B, Türel S. Asit, kronik duodenal ülser ve pilor stenozu ile birlikte pnömatozis sistoides intestinalis. *Turk J Surg* 2008; 24(4): 205-7.
9. Muyembe V. Pneumatosis cystoides intestinalis associated with ascites and pyloric stenosis secondary to a chronic duodenal ulcer: case report. *East Afr Med J* 2002; 79(12): 667-8.
10. Arikanoglu Z, Aygen E, Camci C, Akbulut S, Basbug M, Dogru O, et al. Pneumatosis cystoides intestinalis: a single center experience. *World J Gastroenterol* 2012; 18(5): 453.