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Pneumatosis intestinalis due to gastrointestinal amyloidosis: A case report & review of literature



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ABSTRACT

INTRODUCTION: Pneumatosis intestinalis (PI) is not a disease but a radiological finding with a poorly understood pathogenesis. It can be divided into primary/idiopathic (15%) or secondary (85%) Kim et al. 2007, based on the factors thought to play a role in its development. Amongst the rare causes of secondary PI is gastrointestinal (GI) amyloidosis.

PRESENTATION OF THE CASE: We report a case of a 46-year-old gentleman who presented with a one month history of acute on chronic abdominal pain, associated with one episode of melena. Upon further investigation, he was found to have pneumoperitoneum. He was taken to the operating theatre, where he was noted to have features of pneumatosis intestinalis of the small bowel with no evidence of bowel perforation. Postoperatively, he underwent an upper GI endoscopy with biopsies that revealed GI amyloidosis.

DISCUSSION: One of the rare causes that can lead to secondary PI is GI amyloidosis as proven in our case. Patients with symptomatic gastrointestinal amyloidosis usually present with one of four syndromes: gastrointestinal bleeding, malabsorption, protein-losing gastroenteropathy, and, less often, gastrointestinal dysmotility.

CONCLUSION: GI amyloidosis is a rare cause of secondary pneumatosis intestinalis. The presentation of the disease varies from patient to patient, therefore, the management should be tailored accordingly.

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1. Introduction

PI is not disease but a radiological finding with a poorly understood pathogenesis. It can be divided into primary/idiopathic (15%) or secondary (85%) [8], based on the factors thought to play a role in its development. Amongst the rare causes of secondary PI is gastrointestinal amyloidosis. One of the rare causes that can lead to secondary PI is G.I amyloidosis as proven in our case. Patients with symptomatic gastrointestinal amyloidosis usually present with one of four syndromes: gastrointestinal bleeding, malabsorption, protein-losing gastroenteropathy, and, less often, gastrointestinal dysmotility [13].

2. Case report

46 years old gentleman from Asian descent presented to the Accident & Emergency department with abdominal pain that started one month ago. The pain is generalized and not radiating.

It increased in intensity during last week prior to admission. He had one episode of melena. No associated vomiting or fever. The patient has noticed weight loss over the past few months. He has a history of hyperacidity since one year, which he took medications for. Otherwise he had no other abdominal complaints.

On examination: he was vitally stable and afebrile. Abdominal examination: Soft abdomen, distended but non-tender. Digital rectal examination: showed black tarry stool.

His hemoglobin was low (9.8 g/dl), D-Dimer was (1.46) and he has high PT and PTT. Other than that all his laboratory tests were within normal limits.

Abdominal X-ray revealed features of small bowel obstruction with pneumoperitoneum (Fig. 1).

CT scan showed extensive pneumoperitoneum with focal dilation of the small bowel loop that is associated with pneumatosis intestinalis (Fig. 2).

In view of the imaging findings, patient underwent diagnostic laparoscopy that was converted to exploratory laparotomy due to extensive bowel dilation.

Intra-operatively diffusely dilated small bowel loops were seen from the Doudenoejunal junction up to the proximal ileum. The terminal ileum was seen collapsed and of normal caliber. We

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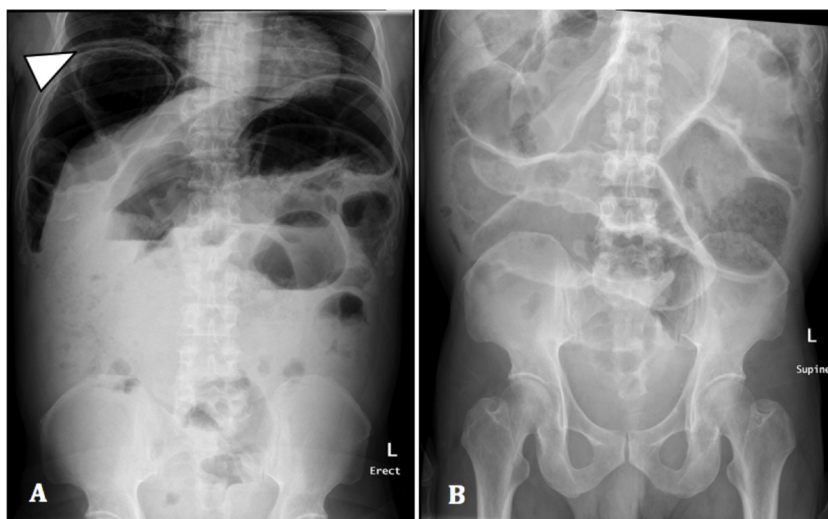


Fig. 1. Abdominal X-ray showing (A) erect view with air fluid levels & free air under right hemidiaphragm (arrow head), (B) supine view with significant dilated bowel loops.

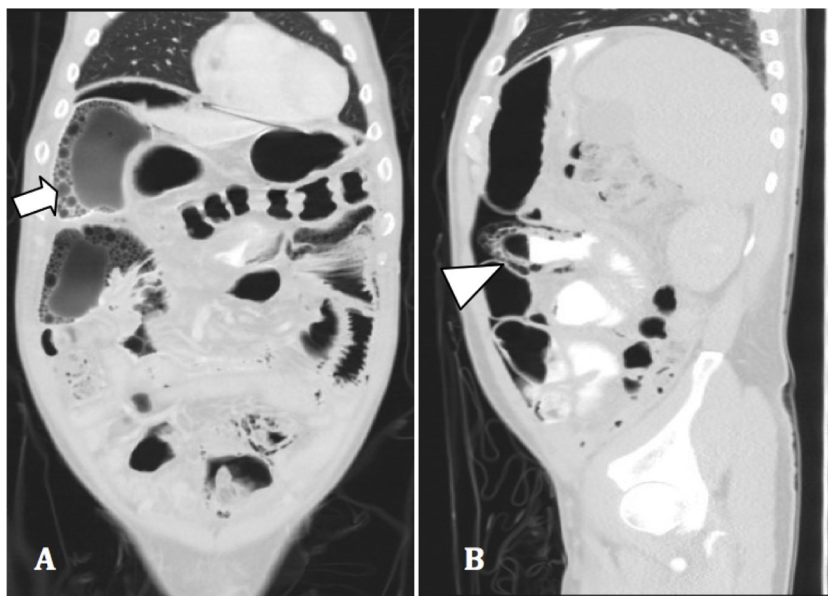


Fig. 2. CT scan of the abdomen showing (A) coronal section with segmental pneumatosis intestinalis (arrow) & in (B) sagittal view (arrow head).

observed segmental areas of pneumatosis intestinalis. In addition to multiple mesenteric hematomas and telangiectatic patches were seen over the small bowel (Figs. 3 and 4). Stretch marks were observed over some of the small bowel loops (Fig. 5). The large bowel, duodenum and stomach were grossly normal. No free fluids or bowel contents were found. Lymph Node biopsy was taken for histopathology. The decision was made to terminate the laparotomy without further intervention.

Post-operatively the patient was doing well. Although his abdomen was distended he had no abdominal pain. He was vitally stable and afebrile. In addition he was having and tolerating full diet and mobilizing fully. Patient was booked for upper & lower G.I endoscopy after discharge.

His colonoscopy was unremarkable. His upper G.I endoscopy has showed areas of ischemic changes in the duodenum from which a biopsy was taken & showed features of amyloid deposition that was confirmed by Congo red stain.

3. Discussion

Pneumatosis intestinalis (PI) is not a disease but a radiographic finding with a poorly understood pathogenesis [1]. The term “pneumatosis intestinalis” was first used by Duo Vernoi during postmortem observations. A PI diagnosis in surviving patients was first established by Hahn in 1899. Radiologically, it was first described by Baumann-Schender in 1939. The condition originally described by Duo Vernoi is what we now consider primary PI. The term “secondary PI” was coined by Koss in 1952, who analyzed 213 pathological specimens and attributed 85% of the cases to a secondary disease [2,3].

Several hypotheses have been proposed regarding the development of PI, although its pathogenesis is still controversial. Two main hypotheses regarding the fundamental pathogenesis of PCI are mechanical and bacterial [4]. The mechanical hypothesis postulates that PI develops when defects in the mucosa, in combination with increased intraluminal pressure, allow gas to infiltrate the gas-



Fig. 3. Showing dilated small bowel loops with segmental area of pneumatosis intestinalis & telangiectatic patches.

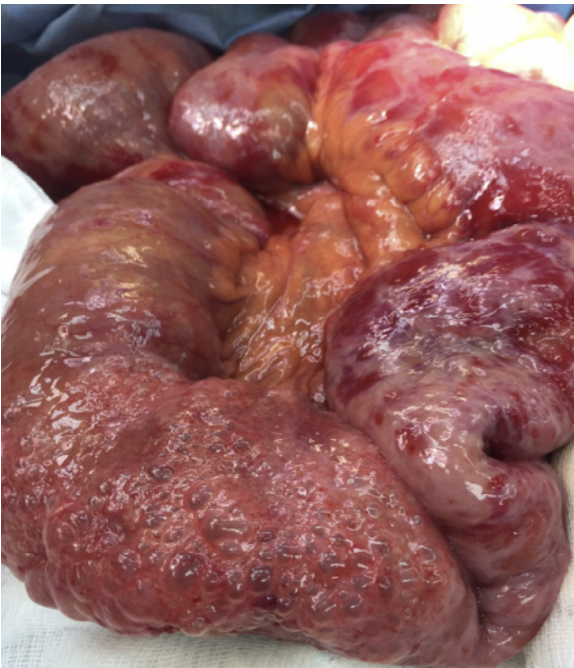


Fig. 4. Showing dilated small bowel loops with segmental area of pneumatosis intestinalis & telangiectatic patches.

trointestinal (G.I) tract wall. A subgroup of patients with severe pulmonary conditions may present with PI arising from pulmonary causes, such as cough and rapid changes in intra-abdominal pressure. The bacterial hypothesis proposes that PI develops when gas-producing bacteria gain entry into the GI tract wall and produce gas pockets. Much of the supporting evidence for these two hypotheses is derived from observational studies, and mechanical and bacterial mechanisms may occur simultaneously [5–7].

PI can be divided into primary/idiopathic (15%) or secondary (85%) [8], based on the factors thought to play a role in its devel-



Fig. 5. showing dilated small bowel loops with stretch marks.

opment. Many GI, non-GI diseases as well surgical and endoscopic trauma can lead to secondary PI [9,10].

One of rare causes that can lead to secondary PI is G.I amyloidosis as proven in our case. Amyloidosis is a generic term that refers to the extracellular tissue deposition of a variety of serum proteins, many of which circulate as constituents of plasma [11]. Amyloidosis of the gastrointestinal tract may be limited to the gut or part of systemic involvement. In a retrospective review of 2334 patients with amyloidosis, 76 patients (3%) had biopsy-proven amyloid involvement of the gastrointestinal (GI) tract of which approximately 80% had systemic amyloidosis and 20 percent had amyloidosis of the gastrointestinal tract without evidence of an associated plasma cell dyscrasia or other organ involvement [12]. GI disease in amyloidosis results from either mucosal or neuromuscular infiltration. In addition, an extrinsic autonomic neuropathy may also affect gut function.

Patients with symptomatic gastrointestinal amyloidosis usually present with one of four syndromes: gastrointestinal bleeding, malabsorption, protein-losing gastroenteropathy, and, less often, gastrointestinal dysmotility [13].

The diagnosis of GI amyloidosis requires a tissue biopsy with positive staining of amyloid by Congo red or the presence of amyloid fibrils on electron microscopy. Although the gastrointestinal complications can result in significant morbidity, they are not usually the cause of death, which is most often due to renal failure, restrictive cardiomyopathy, or ischemic heart disease. However, the presence of hepatic manifestations has a poor prognosis as it likely reflects relatively severe systemic disease. Therapy is directed at the gastrointestinal manifestations and at the underlying cause of amyloidosis [13].

When dealing with PI cases, it is most important to distinguish between life-threatening and non-urgent pathologies. A high index of suspicion is therefore required to rule out the life-threatening causes which include mesenteric ischemia, bowel necrosis or bowel obstruction.

X-rays are of great importance because they are readily available in every emergency department. It may show free air under hemi-diaphragms. This sometimes, however, does not represent a true bowel perforation as these cysts are either submucosal or subserosal & it may rupture [2]. CT has greater sensitivity in diagnosing PI. It may also provide data on other abdominal pathologies, complications such as porto-venous gas, or pneumoperitoneum with gas as little as 1–2 ml of free intraperitoneal air [14,15]. CT scan may also help distinguish between primary and secondary PI. Primary PI will have a bubbly appearance, whereas secondary PI will have a linear radiolucency within the wall of the GI tract, parallel to the intraluminal gas. Laparoscopy is quite useful to confirm, if the physical findings are suspicious, at the same time it has the convenience of conversion to a laparotomy if needed [14].

Most patients with PI never come to clinical attention. The signs and symptoms are either related to the PI or to the underlying

disease. Symptoms also depend on the segment of the intestine involved. Depending on the clinical picture, PI may be treated conservatively by oxygen, IV hydration and antibiotics or if needed immediate surgery [15].

4. Conclusion

G.I amyloidosis is a rare cause of secondary pneumatosis intestinalis. The presentation of the disease varies from patient to patient, therefore, the management should be tailored accordingly.

Conflict of interest

There are no conflict of interest.

Funding

There has been no funding involved in this research.

Ethical approval

Approval obtained from local research committee. Approval was given by consenting the patient.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Authors' contribution

Filza Khalid—author.
Hadiel Kaiyasah—co-author.
Wafa Binfadil—discussion.
Maiyasa Majid—discussion.
Wessam Hazim—contributors.
Yousif ElTayeb—contributors.

References

- [1] P. Sooby, R. Harshen, R. Joarder, An unusual triad of pneumatosis intestinalis, portal venous gas and pneumoperitoneum in an asymptomatic patient, *J. Surg. Case Rep.* (April (4)) (2015), pii:rjv035. 10.1093/jscr/rjv035. PMID: 25858266.
- [2] M.S. Morris, A.C. Gee, S.D. Cho, K. Limbaugh, S. Underwood, B. Ham, M.A. Schreiber, Management and outcome of pneumatosis intestinalis, *Am. J. Surg.* 195 (2008) 679–682, discussion 682–683.
- [3] R. Voboril, Pneumatosis cystoides intestinalis—a review, *Acta Medica (Hradec Kralove)* 44 (2001) 89–92.
- [4] R.K. Yellapu, H. Rajekar, J.D. Martin, T.D. Schiano, Pneumatosis intestinalis and mesenteric venous gas—a manifestation of bacterascites in a patient with cirrhosis, *J. Postgrad. Med.* 57 (2011) 42–43.
- [5] S.D. St Peter, M.A. Abbas, K.A. Kelly, The spectrum of pneumatosis intestinalis, *Arch. Surg.* 138 (2003) 68–75.
- [6] E. Wayne, M. Ough, A. Wu, J. Liao, K.J. Andresen, D. Kuehn, N. Wilkinson, Management algorithm for pneumatosis intestinalis and portal venous gas: treatment and outcome of 88 consecutive cases, *J. Gastrointest. Surg.* 14 (2010) 437–448.
- [7] F.F. Bolukbas, C. Bolukbas, Pnomatozis sistoides intestinalis, *Guncel Gastroenterol.* 8 (2004) 182–185.
- [8] K.M. Kim, C.H. Lee, K.A. Kim, C.M. Park, CT colonography of pneumatosis cystoides intestinalis, *Abdom. Imaging* 32 (2007) 602–605.
- [9] Y. Sakurai, M. Hikichi, J. Isogaki, S. Furuta, R. Sunagawa, K. Inaba, Y. Komori, I. Uyama, Pneumatosis cystoides intestinalis associated with massive free air mimicking perforated diffuse peritonitis, *World J. Gastroenterol.* 14 (2008) 6753–6756.
- [10] E. Türk, E. Karagülle, I. Ocak, D. Akkaya, G. Moray, Pneumatosis intestinalis mimicking free intraabdominal air: a case report, *Ulus Travma Acil Cerrahi Derg.* 12 (2006) 315–317.
- [11] E. Nordling, M. Abraham-Nordling, Colonic amyloidosis, computational analysis of the major amyloidogenic species, serum amyloid A, *Comput. Biol. Chem.* 39 (2012) 29.
- [12] A.J. Cowan, M. Skinner, D.C. Seldin, et al., Amyloidosis of the gastrointestinal tract: a 13-year, single-center, referral experience, *Haematologica* 98 (2013) 141.
- [13] S. Tada, M. Iida, T. Yao, et al., Endoscopic features in amyloidosis of the small intestine: clinical and morphologic differences between chemical types of amyloid protein, *Gastrointest. Endosc.* 40 (1994) 45.
- [14] Z. Arikanoglu, E. Aygen, C. Camci, S. Akbulut, M. Basbug, O. Dogru, Z. Cetinkaya, C. Kirkil, Pneumatosis cystoides intestinalis: a single center experience, *World J. Gastroenterol.* 18 (February (5)) (2012) 453–457, <http://dx.doi.org/10.3748/wjg.v18.i5.453> (PMID:22346251).
- [15] Daniela Berritto, Raffaello Crincoli, Francesca Iacobellis, Francesca Iasiello, Nunzia Luisa Pizza, Francesco Lassandro, Lanfranco Musto, Roberto Grassi, Primary pneumatosis intestinalis of small bowel: a case of a rare disease, *Case Rep. Surg.* 2014 (2014) 350312 (Published online 2014 November 17. 10.1155/2014/350312 PMID: PMC4248370).

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