



Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

Pneumoperitoneum, pneumoretroperitoneum, pneumomediastinum and extensive subcutaneous emphysema in a patient with ulcerative colitis: A case report



Usman T. Siddiqui^{a,*}, Hira Shahzad^a, Asad Jamil Raja^b

^a Medical College, Aga Khan University Hospital, Karachi, Pakistan

^b Department of Surgery, Aga Khan University Hospital, Karachi, Pakistan

ARTICLE INFO

Article history:

Received 3 August 2015

Received in revised form

15 September 2015

Accepted 26 September 2015

Available online 9 October 2015

Keywords:

Ulcerative colitis

Pneumoperitoneum

Pneumoretroperitoneum

Subcutaneous emphysema

Inflammatory bowel disease

ABSTRACT

INTRODUCTION: Pneumo-mediastinum and subcutaneous emphysema are rare presentations of lower gastrointestinal tract perforation.

PRESENTATION OF CASE: We are presenting the case of a middle aged man diagnosed with UC who presented with dyspnea and subcutaneous emphysema, attributed to multiple perforations including the stomach and colon.

CASE DISCUSSION: Patients with ulcerative colitis (UC) are at an increased risk of perforations due to friability of colonic mucosa given the chronic inflammation and relapsing flares. Chronic use of steroids further predisposes to stress ulcers. These pathologies sometimes coexist and identification of each is crucial for the appropriate treatment plan.

CONCLUSION: The case allows for a learning opportunity focusing on coexisting pathologies which may be differentiated based on anatomical knowledge and patient presentation.

© 2015 The Authors. Published by Elsevier Ltd. on behalf of IJS Publishing Group Ltd. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

1. Introduction

Development of pneumomediastinum and subcutaneous emphysema are concomitant presentations of disease or injury to the respiratory tract. Spread of free air along fascial planes from sites distal to the mediastinum has very rarely been reported to cause such a presentation. This free air might result as a consequence of iatrogenic injuries or chronic inflammatory conditions which might cause perforation of a hollow viscus in the gastrointestinal (GI) tract [1–6]. For an underlying GI tract etiology to induce such a presentation, risk factors have been defined for patients undergoing invasive procedures particularly endoscopies and colonoscopies [1–6]. However, spontaneous isolated presentation of air leakage from a chronically diseased hollow organ remains an uncommon finding.

Ulcerative colitis (UC) is a chronic, progressive and relapsing disorder with unknown etiology, characterized by inflammation and ulcerations in the mucosa and submucosa of colon and rectum [7–9]. Patients are at an increased risk of perforation due to widespread inflammation, advanced crypt distortion, formation of crypt abscesses, and mucin depletion which lead onto fulminant colitis or toxic megacolon [10]. We are presenting the case of a

middle aged man diagnosed with UC who presented with dyspnea and subcutaneous emphysema, attributed to multiple spontaneous colonic perforations.

2. Case report

A 53 year old gentleman with ulcerative colitis diagnosed 15 years back, presented to the Emergency Department with progressive shortness of breath on walking for the past 2 days and abdominal distention. Shortness of breath had started spontaneously, was initially severe but stabilized in intensity and consequently limited physical activity. He also complained of generalized weakness and chronic diarrhea for the past 3 months. The patient had a relapse of ulcerative colitis and had about 10+ stools per day with blood. He was started on Azathioprine for one month which he completed and cortisone which he was still taking. He was now having 2–3 stools per day without bleeding or abdominal pain. His weakness had progressively worsened to the point that he was unable to walk with support for the last 1 day. Review of systems was otherwise unremarkable.

On physical examination, he was found to be tachycardic (heart rate 122 beats/min) without fever or lymphadenopathy. Abdominal examination showed a soft distended abdomen which was mildly tender on deep palpation in the periumbilical and epigastric region with questionable rebound tenderness. There was loss of liver dullness at the lower intercostal levels. Digital rectal

* Corresponding author.

E-mail address: usmantsiddiqui@hotmail.com (U.T. Siddiqui).

[Type text]

[Type text]

Siddiqui UT et al.



Fig. 1. CT scan abdomen coronal images showing extensive pneumoperitoneum and subcutaneous emphysema in the chest, neck, axilla and shoulders.

examination revealed a rubbery, irregular, leathery mucosa of the rectum but no blood. Chest examination exhibited subcutaneous emphysema bilaterally along the chest wall. When questioned, the patient admitted to have first noticed it 3 days prior to presentation, initially limited to his face. Further examination showed subcutaneous emphysema in the shoulders, neck, upto the angle of the mandible and in the cervical back region. On the basis of examination findings, patient's history was revisited and was confirmed to be negative for trauma or recent medical procedures. The patient also had no difficulty in breathing at rest or during talking to suggest air leakage from the respiratory tract.

Laboratory workup showed Hemoglobin of 11.6 gm/dl³ (Normal: 13–15), platelets of 582 (Normal: 150–400) and white blood cell count of 15.0 (Normal: 4.0–10.0) with 89% neutrophil count. The patient had a blood urea nitrogen (BUN) of 54 with a creati-

nine of 2.0 which improved to 1.6 with subsequent hydration. A chest radiograph showed extensive subcutaneous emphysema in the chest and neck with some air present under the diaphragm indicating pneumoperitoneum. A computed tomography scan showed extensive subcutaneous emphysema in the neck, shoulders and axilla along with pneumomediastinum, pneumoperitoneum and pneumoretroperitoneum (Fig. 1). Based on the history, physical exam findings and radiographic imaging, a diagnosis of colonic perforation was made and the patient was urgently rushed to the operating room for exploratory laparotomy.

Intraoperatively, we identified gross pus in the peritoneal cavity with multiple perforations in the transverse, descending and sigmoid colon. Bone and muscle of left iliac crest exposed and surrounding abscess was drained and subsequently irrigated with antibiotic solution. A 2 cm perforation at the incisura angularis of

stomach was identified and was repaired with a fat graft. The transverse colon was found to be adherent to the duodenum without any evidence of perforation. A total colectomy with ileostomy was performed and the specimen sent for pathology. The patient was successfully extubated after surgery and did well postoperatively.

Pathology reports of the biopsy specimen showed severe active inflammation, cryptitis, crypt abscess, inflammatory pseudopolyps and crypt distortion. Appendix showed acute and chronic inflammation on the serosal surface. A total of ten lymph nodes were recovered and all showed benign reactive changes without any evidence of malignancy.

3. Discussion

Pneumo-mediastinum and subcutaneous emphysema are rare presentations of lower gastrointestinal tract perforation and have been previously reported in patients with colorectal cancer or diverticulitis, typically linked to retroperitoneal colonic perforation or toxic megacolon, and are extremely uncommon without preceding endoscopic procedures [1–6]. To our knowledge, very few cases have been reported to date. One such case was reported by Annaházi et al. [11] of a 19 year old male with UC who had presented with subcutaneous and mediastinal emphysema, treated medically with steroids, immunomodulators and antibiotics and emergent surgical intervention was avoided. Colonoscopy performed a week later failed to reveal any gross colonic perforation but did show evidence of extensive ulcerations. Rarely similar reports from pediatric population have been discussed previously [10].

Our patient presented with multiple perforations following a relapsing flare of ulcerative colitis which had been settled down with conservative treatment a month prior to his presentation to the ED. The abundance of air seen, showing the “falciform ligament sign” (Fig. 1) [12] is probably from the stomach perforation which was identified intraoperatively. This might have arisen from the chronic use of steroids that the patient was on due to the aggressive disease. During exploratory laparotomy, some of these perforations were noticed along the transverse and descending colon. These findings would explain the occult peritonitis and pneumoperitoneum. Additional perforations were localized in the sigmoid colon, which would rationalize the presence of air within the retroperitoneal area. Furthermore, the manifestation of mediastinal and cervical emphysema can be justified as the air in retroperitoneum which dissected adjoining tissue planes as it escaped along the perivascular adventitia to the anterior pararenal space, and further through the diaphragmatic hiatus along the adventitia of great vessels to the mediastinum and pretracheal fascia to the neck [6].

Medical treatment aims at induction of clinical and endoscopic remission during active disease flares and maintenance therapy to slow progression of disease with steroids, immunomodulators and biological agents [7–9]. Therapeutic endpoint is defined as healing of the mucosa [8]. However, due to progressive and relapsing nature of the disease, approximately 30% of the patients with UC eventually require surgery despite vigorous medical treatment [13]. These include both elective and emergent procedures. Indicators of emergent surgery include patients who present with toxic megacolon, perforation, uncontrolled rectal bleeding or fulminant disease activity in spite of intensive medical therapy [7].

Perforation in our patient's case occurred due to an aggressive flare of his condition and relapse of disease which caused multiple perforations, in the stomach and extending throughout the length of the colon. This necessitated repair of the stomach perforation and a total colectomy. The rectum was however spared and an ileostomy was performed.

Perforations can be often managed medically with bowel rest and intravenous antibiotics [1,2,10]. However, in the light of deteriorating condition and development of signs of peritonitis, surgical intervention becomes mandatory [1]. Toxic mega-colon, perforation, uncontrolled rectal bleeding, sepsis or fulminant disease activity notwithstanding intensive medical treatment, are some of the common indicators of emergent surgery in UC. The standard procedure in such emergent cases is a subtotal colectomy with end ileostomy which is easier to perform than a total proctocolectomy (TPC) with an ileal J-pouch anal anastomosis performed electively. The advantages of such an approach allow for an efficient control over the (potentially) septic emergency and a second restorative procedure later on [7].

This case provides a unique presentation of commonly encountered GI emergencies such as steroid induced stress ulcer perforation as well as colonic perforations secondary to inflammatory conditions such as UC in a single patient. This allows for a learning opportunity focusing on coexisting pathologies which may be differentiated based on anatomical knowledge and patient presentation. It also allows for a multi-pronged treatment strategy; in our case, a colectomy was both therapeutic and prophylactic considering the duration of disease in our patient which significantly raised the chances of colonic cancer.

Conflicts of interest

The authors have no conflict of interest.

Funding

There is no funding supporting this project.

Ethical approval

The study was exempted from the institutional review board since it was a retrospective case analysis with no ethical problems.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

All the authors contributed in the conception, data acquisition, drafting and critically revising the manuscript.

Guarantor

Usman T. Siddiqui.

References

- [1] M. Cappello, C. Randazzo, S. Peralta, G. Cocorullo, Subcutaneous emphysema, pneumomediastinum and pneumoperitoneum after diagnostic colonoscopy for ulcerative colitis: a rare but possible complication in patients with multiple risk factors, *Int. J. Colorectal Dis.* 26 (3) (2011) 393–394, <http://dx.doi.org/10.1007/s00384-010-1005-7>.
- [2] G.B. Schmidt, M.W. Bronkhorst, H.H. Hartgrink, L.H. Bouwman, Subcutaneous cervical emphysema and pneumomediastinum due to a lower gastrointestinal tract perforation, *World J. Gastroenterol.* 14 (24) (2008) 3922–3923, <http://dx.doi.org/10.3748/wjg.14.3922>.
- [3] T. Hur, Y. Chen, G.H. Shu, J.M. Chang, K.C. Cheng, Spontaneous cervical subcutaneous and mediastinal emphysema secondary to occult sigmoid diverticulitis, *Eur. Respir. J.* 8 (1995) 2188–2190.

- [4] T. Nedrebø, Subcutaneous emphysema in gastrointestinal tract perforation, *Tidsskr. Nor. Laegeforen.* 112 (1992) 2855–2856.
- [5] R. Pretre, A. Rohner, Pneumomediastinum and subcutaneous cervical emphysema as signs of rectosigmoid perforation, *Gastroenterol. Clin. Biol.* 16 (1992) 460–462.
- [6] S.D. Fitzgerald, A. Denk, M. Flynn, W.E. Longo, A.M. Vernava 3rd, Pneumopericardium and subcutaneous emphysema of the neck. An unusual manifestation of colonoscopic perforation, *Surg. Endosc.* 6 (1992) 141–143.
- [7] S.H. Baik, W.H. Kim, A comprehensive review of inflammatory bowel disease focusing on surgical management, *J. Korean Soc. Coloproctol.* 28 (3) (2012) 121–131.
- [8] G. Latella, C. Papi, Crucial steps in the natural history of inflammatory bowel disease, *World J. Gastroenterol.* 18 (29) (2012) 3790–3799, <http://dx.doi.org/10.3748/wjg.v18.i29.3790>.
- [9] I. Ordás, L. Eckmann, M. Talamini, D.C. Baumgart, W.J. Sandborn, Ulcerative colitis, *Lancet* 380 (9853) (2012) 1606–1619, [http://dx.doi.org/10.1016/S0140-6736\(12\)60150-0](http://dx.doi.org/10.1016/S0140-6736(12)60150-0).
- [10] A.A. Cooke, A.V. Deshpande, A. Shun, E.V. O’Loughlin, Pneumomediastinum and subcutaneous emphysema in a child with ulcerative colitis, *Pediatr. Emerg. Care* 26 (2) (2010) 129–131, <http://dx.doi.org/10.1097/PEC.0b013e3181ce3022>.
- [11] A. Annaházi, I. Polyák, F. Nagy, T. Wittmann, T. Molnár, Ulcerative crepitus—a case with subcutaneous emphysema and pneumomediastinum without colonic perforation or toxic megacolon in ulcerative colitis successfully treated conservatively, *J. Crohns Colitis* 6 (6) (2012) 717–719, <http://dx.doi.org/10.1016/j.crohns.2012.01.022>, Epub 2012 Feb 24.
- [12] S.H. Kim, S.S. Shin, Y.Y. Jeong, S.H. Heo, J.W. Kim, H.K. Kang, Gastrointestinal tract perforation: MDCT findings according to the perforation sites, *Korean J. Radiol.* 10 (January–February (1)) (2009) 63–70.
- [13] E. Langholz, P. Munkholm, M. Davidsen, V. Binder, Course of ulcerative colitis: analysis of changes in disease activity over years, *Gastroenterology* 107 (1994) 3–11.

Open Access

This article is published Open Access at sciedirect.com. It is distributed under the [IJSCR Supplemental terms and conditions](#), which permits unrestricted non commercial use, distribution, and reproduction in any medium, provided the original authors and source are credited.