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Diaphragm Disease of Small Bowel - The Unsolved Mystery and an Emerging Disease

Dr. Rudraiah HGM, Dr. Prakash MG, Dr. Siddharth Vijay Kalke*, Dr. Basavaraja C, Dr.Chatura Kr[1]. Dr.Aniruddha Desai.

Department of General Surgery JJM Medical College and Bapuji Hospital Davangere, Karnataka.

1- Chief Histopathologist and Professor Department of Pathology JJM Medical College and Bapuji Hospital Davangere, Karnataka.

Corresponding Author: Dr. Siddharth Kalke, Post Graduate Student**,** Pradeep Ayurvedalaya near Dr.Vidyasagar Clinic MCC B block Davangere, Karnataka.

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Abstract

Diaphragm disease is typical of NSAID-related injury of the small intestine but remains an uncommon, although not vanishingly rare, occurrence: 2% of patients taking NSAIDs and COX-2 inhibitors long-term, Diaphragm disease developes. 20-year-old patient presented to casualty of our hospital with pain abdomen,distension,vomiting since 4 days. Patient came with 4 day old multiple ileal perforations with severe fecal peritonitis. Emergency laparotomy was done and patient was managed postoperatively with prolonged ventilator support, inotropic support, higher antibiotics for severe sepsis. On Postoperative day 14 very unlikely the patient developed enterocutaneous fistula when the patient was already mobilizing, tolerating orally and passing stools. So a very unlikely picture of low output enterocutaneous fistula was decided to be managed conservatively with improving patients nutritional status. After 6 weeks the fistula didn't heal so fistulogram was done and re-exploration was planned. Post re- exploration again patient developed an enterocutaneous fistula which healed in another 3 weeks and the stoma was functioning. Granulomatous conditions affecting the bowel are Tuberculosis, Crohns Disease and Diaphragm disease of small bowel. As in our case recurrent enterocutaneous fistulas made us doubt the diagnosis thinking it is crohns disease and whether to start azathioprine and steroids. The histopathologist was very sure of the diagnosis as there were was pyloric gland metaplasia or neuronal hyperplasia. It was extremely challenging and difficult to manage the disease over a period of 10 weeks when the disease entity is so new and hardly any literature is available. Hence our case report serves as a future reference point for other surgeons who will come across such rare entity.

Keywords: Diaphragm disease of small bowel, crohns disease, granulomatous bowel conditions.

Introduction

Diaphragm disease (DD) is a rare gastrointestinal disease due to prolonged intake of non steroidal anti-inflammatory drugs (NSAIDs).[1]. It shows circumferential lesions (<5mm), most commonly in the small intestine, causing multiple strictures. NSAID-related enteropath is under research nowadays.[2]. Diaphragm disease of small

intestine is rare disease and its prevalence is not studied.[3,4]. There is no pathogenesis in literature; however, it is believed that (NSAIDs) play an important role.[5,6]. Inhibition of of cyclo-oxygenase 1 (COX-1) and therefore prostaglandins in small intestine result in mucosal injury, fibrosis, and mucosal barrier breakdown resulting in an Inflammatory response leading to circumferential ulceration and diaphragm formation.[4,6]. This disease manifests with nonspecific symptoms, including abdominal pain, nausea, vomiting, and features suggestive of complete or incomplete small bowel obstruction.

Case Description

20-year-old patient presented to casualty of our hospital with pain abdomen, distension, vomiting since 4 days. Patient also complained of absolute constipation since 4 days. On evaluation patient had air under diaphragm on erect xray abdomen. Patient gave history of prolonged intake of NSAIDS for pain abdomen. Emergency laparotomy was planned. Multiple ileal perforations each of size 3*2cm with a blue ring surrounding each perforation was seen. Severe fecal peritonitis was seen. Resection and end to end anastamosis was done. Thorough bowel wash was given and intra-abdominal drains were placed .Postoperatively patient improved symptomatically and tolerated orally and passed stools. On post operative day 14 patient developed low output enterocutaneous fecal fistula which was decided to be managed conservatively with nutritional support. After 6 weeks of Total parenteral nutrition (TPN) and control of sepsis fistulogram was done and patient was planned for re-exploration. On re- exploration enterocutaneous fistula from the ascending colon, transverse colon was seen and also new ileal perforations were seen. Previous anastamosed site was healthy. Primary closure of perforations with excision of fistulous tract was done and a diversion ileo-transverse anastamosis was done with proximal ileal ileostomy. Postoperatively patient again developed enterocutaneous fistula which healed in next 3 weeks and the ileostomy was functioning. Over a period of 10 weeks patient developed severe sepsis with ventilator associated pneumonia, massive left pleural effusion and bedsores. Patient was managed aggressively with TPN, wound care, control of severe sepsis with higher antibiotics and ventilator support.

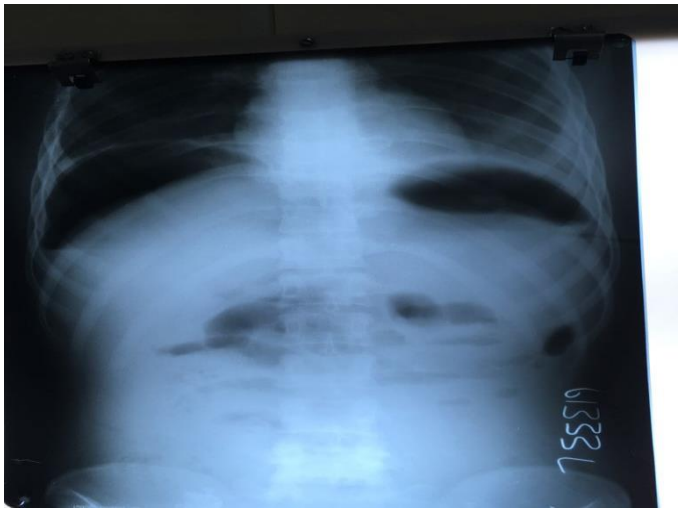

Figure 1: showing air under diaphragm on admission on an erect xray abdomen and pelvis.

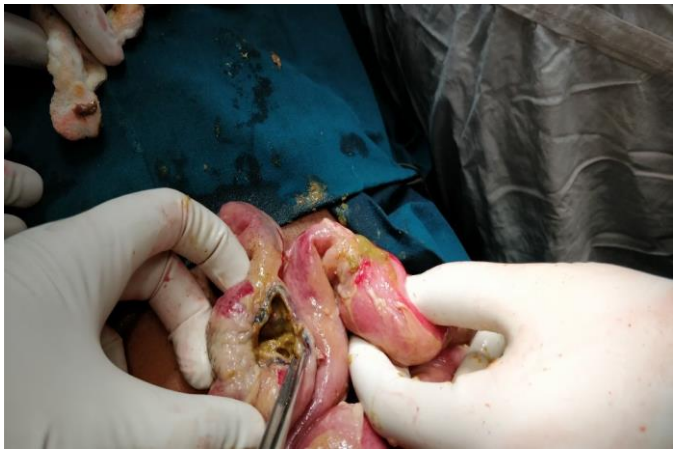

Figure 2: Showing intraoperative picture of the ileal perforation. Note the large size of the perforation.

Histopathology

The gross features recorded in 30 cms length of small intestine were ring-like depression on the external surface of the intestine which showed five perforations with areas of fat wrapping (Fig 3) and on cut section two

diaphragms defined as an intraluminal, circumferential, and protruding ridge of tissue with luminal stenosis and presence of circumferential ulceration were seen.(Fig 4)

Microscopy showed focal and diffuse pattern of injury with fissuring ulcer, transmural lymphoid infiltrate outside the ulcer(Fig 5), neutrophil abscess, granulomas with foreign body giant cells and organizing serosal exudate. Patchy areas corresponding to the strictures showed ulceration and submucosal fibrous thickening with fat wrapping on the serosa. (Fig 6)

Gross and microscopic features, in view of NSAID history and obstructive symptoms suggested the diagnosis of diaphragm disease.

Figure 3: Gross Mounted Specimen

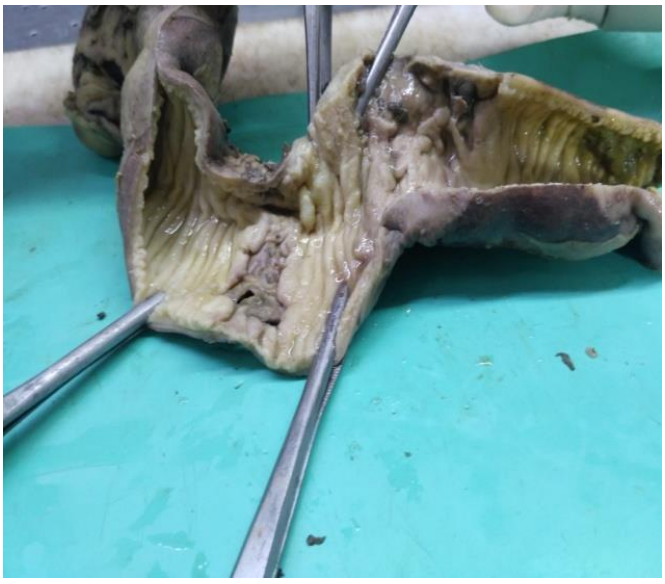

Figure 4: Gross Mounted Specimen

Figure 3 and 4 The gross features recorded in 30 cms length of small intestine were ring-like depression on the external surface of the intestine which showed five perforations with areas of fat wrapping (Fig 3) and on cut section two diaphragms defined as an intraluminal, circumferential, and protruding ridge of tissue with luminal stenosis and presence of circumferential ulceration were seen.(Fig 4)

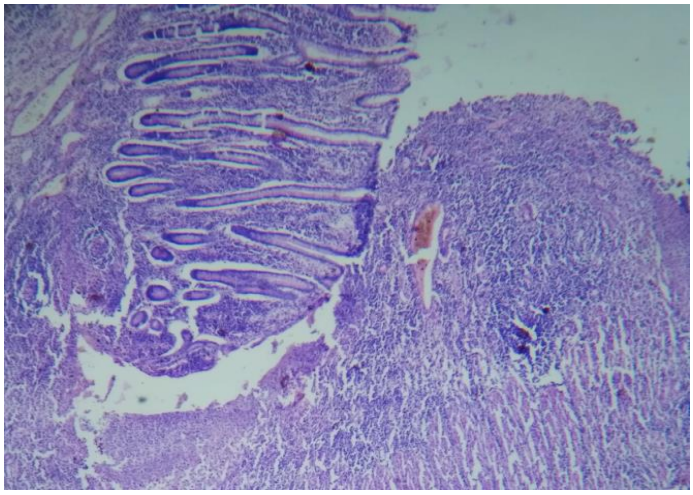

Figure 5: Microscopic Appearance Of Diaphragm

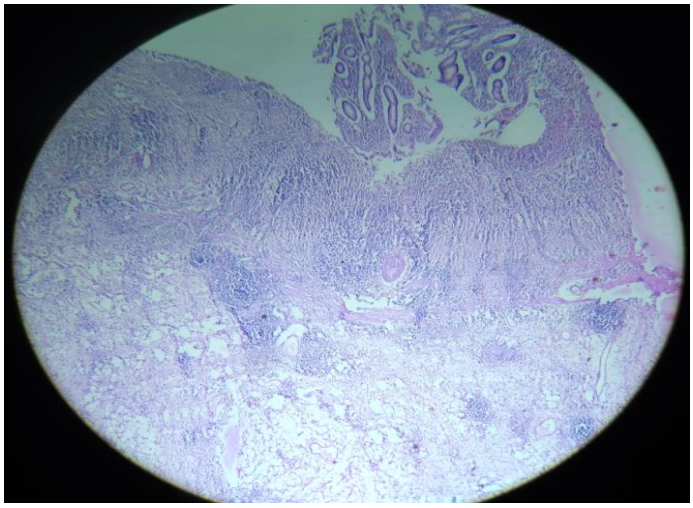

Figure 6: Microscopic Appearance

Figure 5 And 6 showing focal and diffuse pattern of injury with fissuring ulcer, transmural lymphoid infiltrate outside the ulcer(Fig 5), neutrophil abscess, granulomas with foreign body giant cells and organizing serosal exudate. Patchy areas corresponding to the strictures showed

ulceration and submucosal fibrous thickening with fat wrapping on the serosa. (Fig 6).

Treatment

Patient came with 4 day old multiple ileal perforations with severe fecal peritonitis. Emergency laparotomy was done and patient was managed postoperatively with prolonged ventilator support, inotropic support,higher antibiotics for severe sepsis. On Postoperative day 14 very unlikely the patient developed enterocutaneous fistula when the patient was already mobilizing, tolerating orally and passing stools. So a very unlikely picture of low output enterocutaneous fistula was decided to be managed conservatively with improving patients nutritional status. After 6 weeks the fistula didn't heal so fistulogram was done and re- exploration was planned. Post re- exploration again patient developed a enterocutaneous fistula which healed in another 3 weeks and the stoma was functioning.

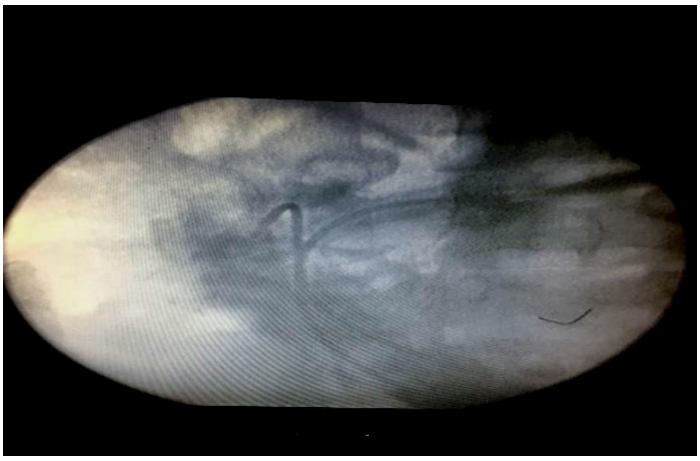

Figure 7: showing Fistulogram with fistula extending in the ascending colon.

Discussion

Diaphragm disease is typical of NSAID-related injury of the small intestine but remains an uncommon, although not vanishingly rare, occurrence: In 2% of patients taking NSAIDs and COX-2 inhibitors long-term, Diaphragm disease developed.[12] Women are more commonly affected than are men, and the small intestine is more commonly affected than is the colon.[13] The route of ingestion of NSAIDs does not preclude the formation of diaphragms: Diaphragm disease has been reported in a bypassed bowel segment, suggesting systemic effect of NSAIDs.[14] Granulomatous conditions affecting the bowel are Tuberculosis, Crohns Disease and Diaphragm disease of small bowel. As in our case recurrent enterocutaneous fistulas made us doubt the diagnosis thinking it is crohns disease and whether to start azathioprine and steroids. The histopathologist was very sure of the diagnosis as there were was pyloric gland metaplasia or neuronal hyperplasia. It was extremely challenging and difficult to manage the disease over a period of 10 weeks when the disease entity is so new and hardly any literature is available. But due to over the counter use of NSAID's in today's world this granulomatous condition is on the rise and soon will possess a major health hazard.

Conclusion

Diaphragm disease was first described by Lang et al [7] in 1988. He reported seven patients who had been taking NSAIDs, who had developed small bowel strictures resembling perforated diaphragms. Since this description, diaphragm disease has become a well recognised though relatively rare complication of NSAIDs use.[8,9,10]. The most common presentation of diaphragm disease is with anaemia in combination with obstructive symptoms including abdominal pain, distention, vomiting and weight loss. A definitive preoperative diagnosis of diaphragm disease is extremely difficult to make. However, if there is a high suspicion of diaphragm disease, then conservative treatment with withdrawal of the NSAIDs can be adopted,[11] though if narrow or long fibrostenotic strictures have become established, then surgical resection may be inevitable, as once fibrous scar tissue has matured, removal of the cause of injury is unlikely to allow resolution. This case report highlights an important condition, small bowel diaphragm disease, which will

become more prevalent with the continued and widespread use of NSAIDs. Why recurrent multiple enterocutaneous fecal fistulas happen is a mystery unsolved and the question arising is that in the near future will it be a bigger threat than crohns disease. A history of long NSAIDs use and the presence of anaemia should lead clinicians to consider diaphragm disease in the differential diagnosis, particularly when oesophagogastrodudenoscopy and colonoscopy are negative.

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