

Rare Case of Pemphigus Vulgaris Presenting as Acute Small Bowel Obstruction

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Abstract: *The clinical presentation of pemphigus vulgaris presenting as acute small bowel obstruction, unlike upper gastrointestinal presentations such as upper gastrointestinal bleed or dysphagia. This study aims to present a detailed case report on the infrequent occurrence of pemphigus vulgaris presenting as acute small bowel obstruction. The objectives are to describe the clinical presentation, diagnostic challenges, surgical intervention, postoperative management, and to review relevant literature to highlight the complexity and rarity of this case.*

Keywords: Pemphigus Vulgaris, acute small bowel obstruction

1. Introduction

The term pemphigus refers to a group of autoimmune blistering diseases of skin and mucous membranes that are characterized histologically by intraepidermal blisters due to acantholysis (i.e., separation of epidermal cells from each other) and immunopathologically by in vivo bound and circulating immunoglobulin directed against the cell surface of keratinocytes. Mucous membranes are usually affected before any cutaneous lesions manifest. The most common gastrointestinal symptoms include: Odynophagia, dysphagia, emesis, hematemesis Oral ulcers, then cutaneous manifestations May involve: oropharynx, skin, esophagus, conjunctiva, nasal mucosa, larynx, urethra, vulva, cervix Esophageal involvement is often asymptomatic but may present with esophagitis.

2. Case Report

A 53 years old female patient with pemphigus vulgaris on regular immunosuppression who presented with severe abdominal pain and distension, features of bowel obstruction. The symptoms were sudden in onset associated with 5 episodes of vomiting and mild fever. Also, she complaints of obstipation for the past 2 days. No similar previous history or history of medical gastroenterologist consultations. She is also not a known case of diabetes mellitus or hypertensive or with any other comorbidities. No other aggravating or relieving factors. No history of recent abdominal surgery / and other malignancy / family history.

Examination: patient was toxic, with painful distended abdomen. No guarding or rigidity present over the abdomen. Patient was tachypneic and was responding to all commands. Her bowel sounds were absent. And on per rectal examination showed mucus staining with normal anal tone. Her vitals were normotensive and normothermic but tachypneic and tachycardia was present and her blood gas analysis showed metabolic acidosis.



Figure 1: CT abdomen showing small bowel obstruction



Figure 2: hematoma evacuated from lumbar region which was causing small bowel obstruction

Procedure:

Patient was operated in Emergency Operation theatre under general anesthesia and midline laparotomy was done. there was a hematoma in the lumbar region with pus collection around it. Both were cleared and the bowel viability was checked and wash given and the wound closed under aseptic conditions.

3. Discussion

The immunobullous disorders represent a group of conditions characterized by antibody-mediated autoimmune responses against structural elements of the skin resulting in blistering of the skin and mucosae. Antibody targets include proteins in the hemidesmosomes and the basement membrane zone (pemphigoid group), desmosomes (pemphigus group) and epidermal and tissue-type transglutaminase (dermatitis herpetiformis). If left untreated, immunobullous disorders may be associated with significant morbidity and mortality and thus prompt accurate diagnosis and treatment are mandatory. Esophageal or small bowel involvement of pemphigus vulgaris is rare, and when present, the most common presenting symptoms reported in the medical literature are odynophagia and dysphagia. Here, we present a pemphigus vulgaris presenting with upper gastrointestinal obstruction because of idiopathic hematoma causing small bowel obstruction.

4. Conclusion

Pemphigus vulgaris which occurs commonly in 6th decade, mostly it presents with outright skin manifestations but involvement of gastrointestinal system is rare. This case gives us insight into challenging and difficult decisions made in the background of pemphigus vulgaris presenting as acute small bowel obstruction. We should approach such dermatological conditions with open mind rather they won't present with such acute conditions of bowel

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Institutional Review Board Statement:

The study did not require ethical approval due to the nature of the study (case report).

Informed Consent Statement:

Written Informed consent was obtained from the patient to publish this paper.

Data Availability Statement:

The data used in this study was accurately cited within the manuscript. Further inquiries can be directed to the corresponding author.

Conflicts of Interest:

The authors declare no conflict of interest.

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