Association of Idiopathic Type 3 Hypertrophic Pyloric Stenosis with Gastric Volvulus

Authors: Tannaz Safari Vejin (MD-candidate, MPH), T; Ian Dwinell (MD, MBA, PGY-4); Charles Thompson III (MD, MBA, FACS, FASMBS)

Mount Sinai South Nassa, Department of Surgery, General Surgery, Oceanside, New York, US

Introduction

Gastric volvulus (GV) is a medical anomaly characterized by the stomach's atypical rotation, leading to obstruction and potentially compromising blood supply. The majority of these rotations occur organoaxially, where the stomach turns around its longitudinal axis, accounting for 60-80% of cases. Various factors can induce GV, including defects in the diaphragm, the presence of paraesophageal hernias, and the relaxation of specific gastric ligaments. In rare instances, GV has been associated with idiopathic hypertrophic pyloric stenosis (IHPS)).[1,2,3,4,5,7,8,12]

IHPS in adults manifests in three main types, with Type 3 being idiopathic. While IHPS's primary form arises without an underlying cause, the secondary variant results from other pathologies, such as gastrointestinal tumors or post-operative adhesions. Diagnostic hallmarks for IHPS include pyloric hypertrophy and its associated symptoms such as postprandial nausea and vomiting.[6]

In this report, we chronicle the cases of two patients diagnosed with mesenteric-axial in the context of Idiopathic Type 3 pyloric stenosis. Both underwent robotic gastroduodenal resection surgery using Billroth II Anastomosis (BRII) and subsequent pathological examination revealed hypertrophic pyloric tissue. These cases underscore a potential relationship between IHPS and GV, a link previously unexplored in-depth in medical literature.

Case Description & Procedure

Two adult patients with symptoms indicative of GV were diagnosed with IHPS Type 3. Diagnostic evaluation included UGI studies, endoscopy, and CT scans. Both patients underwent a robotic BRII procedure with pylorus removal and antrectomy.

Nuances specific to each patient were:

- **First Patient (A):** Incidental appendiceal mucocele led to appendectomy

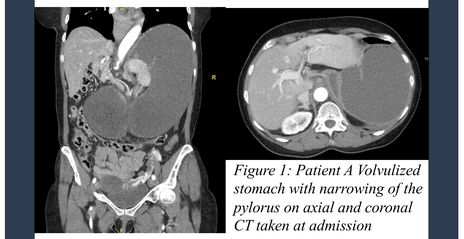


Figure 1: Patient A Volvulized stomach with narrowing of the pylorus on axial and coronal CT taken at admission

- **Second Patient (B):** concurrent large hiatal hernia repair was performed.
  A holistic patient care approach was maintained, encompassing pre-operative and diagnostic assessment, surgical execution, and post-operative progress. Pathology report for any resected tissues were analyzed histopathologically to confirm IHPS, focusing on muscularis mucosa hypertrophy.

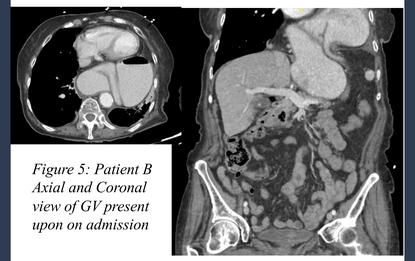


Figure 5: Patient B Axial and Coronal view of GV present upon on admission

Results

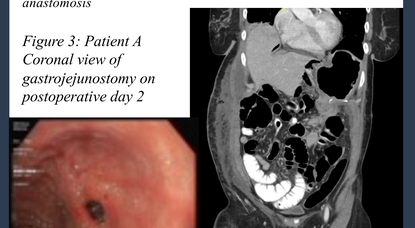
Both patients recovered from their robotic BRII procedures without incident. Pathologic findings revealed a hypertrophic pylorus, with hypertrophy of the muscularis mucosa. Postoperatively, both patients received UGI series to reveal free passage of contrast into the jejunum.

This comprehensive case report reveals the intricate diagnostic considerations and the successful surgical repair of Patient A's gastric obstruction and concurrent complications, possibly furthering our understanding of GV and its associated complications. Previous studies and case reports have mainly focused on importance of early diagnosis and uses of laparoscopic and open surgical repairs for GV and IHPS .[7,12, 13]

Additionally, IHPS is extremely seldom stumbled upon and therefore less thoroughly speculated for its origin of cause let alone methods of surgical repair. Therefore, literature on this topic is significantly limited and demands further investigation and contemplation particularly in the field of general surgery.[7,8,9,1012,13] As far as we know only one case in previous literature reported using BR I anastomosis robotic surgery repair, and ours the only one reporting the use of BRII anastomosis repair in our two patients presenting with IHPS.[13] More investigation and post-operative observation is needed in exploring the efficacy of using robotic surgical repair, in particular BR I and II anastomosis for patients with GV and IHPS.

Figure 4: Patient A's UGI study using Esophagogastroduodenoscopy (EGD) both intra-op and 1 month post-op showing intact BRII anastomosis

Figure 3: Patient A Coronal view of gastrojejunostomy on postoperative day 2



Conclusion

The association between IHPS and GV, as presented, underlines the necessity of keeping an expansive differential diagnosis, especially in adult patients with GI symptoms. This report underscores the merit of combining state-of-the-art diagnostic tools with innovative surgical techniques, to ensure the best outcomes for patients. The medical fraternity can significantly benefit from this knowledge, enhancing diagnostic precision and patient care for such rare and complex presentations.

The concurrent presentation of idiopathic type 3 IHPS with GV in adult patients, as described in these cases, is a rare medical phenomenon. Early detection and a comprehensive diagnostic approach, utilizing tools like GI endoscopy, CT scans, and UGI studies, are crucial for accurate identification. Robotic surgical interventions, especially the BRII anastomosis, have proven effective in treating both conditions concurrently, highlighting the importance of adaptable surgical techniques. The association between IHPS and GV emphasizes the need for clinicians to consider potential overlaps between different gastrointestinal disorders. Ensuring multidisciplinary involvement, encompassing both surgical and medical teams, is key for optimal patient outcomes.

References

1. Barussaud ML, Mesrotte G, Canaguera E, et al. Gastric volvulus: diagnostic and therapeutic management. J Visc Surg. 2014;151(3): 193-200. doi: 10.1016/j.jviscsurg.2014.05.005
2. Ebert EC. Gastrointestinal manifestations of connective tissue disorders. J Clin Gastroenterol. 2006;40(9): 769-775. doi: 10.1097/01.mcg.0000225641.84405.b3
3. Inoue T, Okada H, Okada H, et al. Current status in the diagnosis and treatment of gastric volvulus in Japan. Surgery. 2014;155(1): 21-27. doi: 10.1016/j.surg.2013.06.038
4. Lukaszczyk JJ, Janczak DT, Szulc R. Gastric volvulus--diagnosis, treatment, and difficulties: Hassan, S. M., Mubeek, A., Maddassir, S., & Haq, F. (2018). Adult idiopathic hypertrophic pyloric stenosis - a common presentation with an uncommon diagnosis. *Journal of Community Hospital Internal Medicine Perspectives*, *8*(2), 64–67. https://doi.org/10.1080/20009666.2018.1444905
6. Hallas M, Lee T, Lerner T. Diagnosis and therapy of primary hypertrophic pyloric stenosis in adults: case report and review of literature. J Gastrointest Surg. 2006 Feb;10(2):265-9. doi: 10.1016/j.gassur.2005.06.003. PMID: 16455460.
7. Tamburini, N., Andolfi, C., Vigoda, C., Nasti, M., Resta, G., Marino, S., Rubino, S., Cavallesco, G., Occhionorelli, S., Vasquez, G., & Anania, G. (2021). The Surgical Management of Acute Gastric Volvulus: Clinical Outcomes and Quality of Life Assessment. *Journal of laparoendoscopic & advanced surgical techniques. Part A*, *31*(3), 347–350. https://doi.org/10.1089/lap.2020.0770
8. Rashid, F., Thangarajah, T., Mulvey, D., Lanvin, M., & Iftikhar, S. Y. (2010). A review article on gastric volvulus: a challenge to diagnosis and management. *International journal of surgery (London, England)*, *8*(1), 18–24. https://doi.org/10.1016/j.ijsu.2009.11.002
9. Hattori, A., Kawabata, H., Umeda, Y., Tsuboi, J., Yamada, R., Hamada, Y., & Tanaka, K. (2020). Adult hypertrophic pyloric stenosis that improved by spontaneous double-channel pylorus formation. *JGH open : an open access journal of gastroenterology and hepatology*, *5*(2), 317–319. https://doi.org/10.1002/jgh3.12454
10. Hallas, M., Lee, T., & Lerner, T. (2006). Diagnosis and therapy of primary hypertrophic pyloric stenosis in adults: case report and review of literature. *Journal of gastrointestinal surgery : official journal of the Society for Surgery of the Alimentary Tract*, *10*(2), 265–269. https://doi.org/10.1016/j.gassur.2005.06.003
11. Pintor-Tortolero, J., García-León, A., & Padilla-Ruiz, J. (2021). Surgery for Strangulated Gastric Volvulus Should Not Be Delayed. *Clinical gastroenterology and hepatology : the official clinical practice journal of the American Gastroenterological Association*, *19*(7), A28–A29. https://doi.org/10.1016/j.cgh.2020.04.049
12. Abdessamad EI, KAOUKABI, Mohamed MENFAA, Samir HASBI, Fouad SAKIT, Abdelkrim CHOHO, "Acute Gastric Volvulus on Hiatal Hernia", *Case Reports in Surgery*, vol. 2020, Article ID 8147278, 5 pages, 2020. https://doi.org/10.1155/2020/8147278
13. Lin, H. P., Lin, Y. C., & Kuo, C. Y. (2015). Adult idiopathic hypertrophic pyloric stenosis. *Journal of the Formosan Medical Association = Taiwan yi zhi*, *114*(7), 659–662. https://doi.org/10.1016/j.jfma.2012.07.001

Funding: The authors have no relevant financial relationships or in-kind support to disclose. Disclosure statement: All authors involved in this manuscript had no conflict of interest to disclose. Keywords: Gastric Volvulus, Adult-onset hypertrophic pyloric stenosis, idiopathic hypertrophic pyloric stenosis, Case report, hiatal hernia, gastric outlet syndrome, pyloric stricture, Billroth 2 anastamosis, robotic surgery repair, appendiceal mucocele, paraesophageal hernia. Abbreviations: GV: gastric volvulus, IHPS; idiopathic hypertrophic pyloric stenosis, BRII; BillRoth II anastamosis, EGD; esophagogastroduodenoscopy (EGD), UGI; upper gastrointestinal, RLQ; Right lower quadrant, ED; Emergency department, HTN; hypertension, IV; intravenous, GI; Gastrointestinal, DVT; Deep Vein Thrombosis, PACU; post-anesthesia care unit, OR; operating room