



RECTAL GIST PRESENTING AS GANGRENOUS RECTAL PROLAPSE: A RARE CASE REPORT

General Surgery

**Dr Sriranganath
H S***

Post Graduate, Department of General Surgery, JJMMC, Davangere-577004, Karnataka
*Corresponding Author

Dr Jagadeesha BVC

Department of General Surgery, JJMMC, Davangere-577004, Karnataka

**Dr Mahesh
Kariyappa**

Department of General Surgery, JJMMC, Davangere-577004, Karnataka

ABSTRACT

Gastrointestinal stromal tumors or "GIST" are mesenchymal neoplasms expressing KIT(CD117) tyrosine kinase and showing the presence of activating mutations in KIT or PDGFR α (platelet-derived growth factor alpha). GIST of anal canal is an extremely rare tumor, accounting for only 3% of all anorectal mesenchymal tumors and 0.1–0.4% of all GIST. GIST with large tumor size and high mitotic activity are highly malignant, but the biological behavior of anorectal GIST is less clear. Abdominoperineal resection (APR) or conservative surgery is the best treatment option. Imatinib mesylate, a tyrosine kinase inhibitor, has shown promising results in its management. We present a case of rectal GIST which presented as irreducible rectal prolapse. The patient underwent abdominoperineal resection (APR) and was confirmed on histopathology to have rectal GIST with tumor size more than 5 cm in maximum dimension

KEYWORDS

Introduction

Gastrointestinal stromal tumor or "GIST" was a name given in 1983 to a group of gastrointestinal tumors which were otherwise unclassifiable as being of smooth muscle or neurogenic origin [1]. They are mesenchymal neoplasms expressing KIT(CD117) tyrosine kinase and showing presence of activating mutations in KIT or PDGFR α (platelet-derived growth factor alpha) [2]. It is the commonest gastrointestinal mesenchymal tumor [3] with the commonest site being stomach (50–60%), followed by small intestine (30–40%), colon (7%), and oesophagus (1%) [4]. GIST of anal canal and rectum are often grouped together and account for nearly 5% of all GIST [4, 5]. However, of these only 2–8% are from rectum, making GIST of anal canal an extremely rare tumor [6, 7].

Case Report

A 70-year-old male presented with an irreducible gangrenous rectal prolapse along with a prolapsed rectal growth. He had history of prolapsed from past 1 year which he used to reduce himself. There was no history of bleeding per rectum. The patient was not a known case of piles/diabetes mellitus/hypertension/tuberculosis/or any other chronic ailment. He had no urinary complaints. There were no other complaints referable to chest and cardiac or nervous system.

On examination he was moderately built and of average nourishment. Karnofsky performance scale was more than 80. There was no pallor/icterus/pedal edema/lymphadenopathy. On digital rectal examination friable gangrenous irreducible mass measuring 10*8 cms which bled on manipulation. It was found to be arising from the anterior wall of the prolapsed rectum.

Abdominal examination was normal. His hematological and biochemical parameters were all within normal limits. patient underwent abdominoperineal resection with end colostomy and histopathological examination report revealed rectal GIST with tumor size more than 5 cm in maximum dimension Mitotic figures were less than 5/50 HPF.



Fig1: Prolapsed rectal gist **Fig2: Friable growth-intraoperative picture**



Fig3: Post op picture.- perineal site **Fig4: End colostomy**

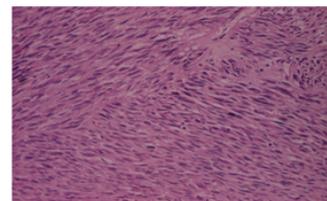


Fig5: Histopathological picture

Discussion:

The incidence of anal cancer in the western world is between 7 and 9 per million population. It contributes to only 1.5% of all malignancies of the digestive system [8]. Of these, GIST form only 3% of anorectal mesenchymal tumors [7]. However, the exact data in eastern world may be different, as in China, unlike in western countries, rectal cancer accounts for approximately 70% of colorectal cancers [9]. GIST are currently thought to originate from interstitial cells of Cajal. The presence of interstitial Cajal-like cells has been reported in several extraintestinal organs including urinary bladder, prostate, gallbladder, omentum, uterus, fallopian tube, and atrial and ventricular myocardium [10, 11]. This may explain the development of extraintestinal GIST [12, 13]. Mutational statuses of c-KIT and PDGFR α genes are the basis for the diagnosis of this neoplasia and represent the criteria for surgical therapy, expected chemotherapy response, and clinical outcomes [6]. GIST with large tumor size and high mitotic activity are highly malignant, but the biological behavior

of anorectal GIST is less clear. GIST of size <2 cm and mitosis <5 per 50 HPF were indolent, whereas those with size >5 cm and/or mitosis >5 per 50 HPF were highly malignant.

GIST are best treated by surgery and are not radio- or chemosensitive. However, controversy exists whether abdominoperineal resection (APR) or conservative surgery is the best alternative [14]. Though the incidence of local recurrence is lower after APR, the distant metastasis and survival are not significantly different [15]. Patterns of recurrence and metastasis for anorectal GIST are the same as for GIST elsewhere, and the disease usually has a long or protracted course. Therefore long followups are essential and local recurrences if any can be reoperated, if resectable. As regards adjuvant or salvage therapy imatinib mesylate, a tyrosine kinase inhibitor, has shown promising results in the management of patients with GIST [14].

Anorectal GIST, though rare, should be considered in the differential diagnosis of tumors in this region, especially if the pre-operative biopsy is equivocal. Gross and histopathological are both important, as prognosis depends on tumor size as well as grade. However, prognosis is usually better than for corresponding carcinomas in the region. Immunohistochemistry is a must, as CD-117 score is not only diagnostic but also guides adjuvant therapy and is an important prognostic marker.

References:

1. F. van der Aa, R. Sciot, W. Blyweert et al., "Gastrointestinal stromal tumor of the prostate," *Urology*, vol. 65, no. 2, p. 388, 2005. View at Publisher · View at Google Scholar · View at Scopus
2. M. C. Heinrich, C. D. Blanke, B. J. Druker, and C. L. Corless, "Inhibition of KIT tyrosine kinase activity: a novel molecular approach to the treatment of KIT-positive malignancies," *Journal of Clinical Oncology*, vol. 20, no. 6, pp. 1692–1703, 2002. View at Publisher · View at Google Scholar · View at Scopus
3. A. Ghobadi, W. Kabbani, B. Barker, and J. E. Dowell, "Rectal GI stromal tumor mimicking a prostate mass," *Journal of Clinical Oncology*, vol. 25, no. 36, pp. 5827–5828, 2007. View at Publisher · View at Google Scholar · View at Scopus
4. M. Miettinen, M. Sarlomo-Rikala, and J. Lasota, "Gastrointestinal stromal tumours," *Annales Chirurgiae et Gynaecologiae*, vol. 87, no. 4, pp. 278–281, 1998. View at Google Scholar · View at Scopus
5. M. Miettinen, M. Furlong, M. Sarlomo-Rikala, A. Burke, L. H. Sobin, and J. Lasota, "Gastrointestinal stromal tumors, intramural leiomyomas, and leiomyosarcomas in the rectum and anus: a clinicopathologic, immunohistochemical, and molecular genetic study of 144 cases," *American Journal of Surgical Pathology*, vol. 25, no. 9, pp. 1121–1133, 2001. View at Publisher · View at Google Scholar · View at Scopus
6. G. R. Nigri, M. Dente, S. Valabrega et al., "Gastrointestinal stromal tumor of the anal canal: an unusual presentation," *World Journal of Surgical Oncology*, vol. 5, article 20, 2007. View at Publisher · View at Google Scholar · View at Scopus
7. J. A. Tworek, J. R. Goldblum, S. W. Weiss, J. K. Greenson, and H. D. Appelman, "Stromal tumors of the anorectum: a clinicopathologic study of 22 cases," *American Journal of Surgical Pathology*, vol. 23, no. 8, pp. 946–954, 1999. View at Publisher · View at Google Scholar · View at Scopus
8. J. C. Bendell and D. P. Ryan, "Current perspectives on anal cancer," *Oncology*, vol. 17, no. 4, pp. 492–503, 2003. View at Google Scholar · View at Scopus
9. D. B. Zhao, Y. K. Wu, Y. F. Shao, C. F. Wang, and J. Q. Cai, "Prognostic factors for 5-year survival after local excision of rectal cancer," *World Journal of Gastroenterology*, vol. 15, no. 10, pp. 1242–1245, 2009. View at Publisher · View at Google Scholar · View at Scopus
10. F. van der Aa, T. Roskams, W. Blyweert, and D. De Ridder, "Interstitial cells in the human prostate: a new therapeutic target?" *Prostate*, vol. 56, no. 4, pp. 250–255, 2003. View at Publisher · View at Google Scholar · View at Scopus
11. K. W. Min and M. Leabu, "Interstitial cells of Cajal (ICC) and gastrointestinal stromal tumor (GIST): facts, speculations, and myths," *Journal of Cellular and Molecular Medicine*, vol. 10, no. 4, pp. 995–1013, 2006. View at Google Scholar · View at Scopus
12. B. D. Gun, M. O. Gun, and Z. Karamanoglu, "Primary stromal tumor of the omentum: report of a case," *Surgery Today*, vol. 36, no. 11, pp. 994–996, 2006. View at Publisher · View at Google Scholar · View at Scopus
13. C. H. Lee, Y. H. Lin, H. Y. Lin, C. M. Lee, and J. S. Chu, "Gastrointestinal stromal tumor of the prostate: a case report and literature review," *Human Pathology*, vol. 37, no. 10, pp. 1361–1365, 2006. View at Publisher · View at Google Scholar · View at Scopus
14. J. C.-M. Li, S. S.-M. Ng, A. W.-I. Lo, J. F.-Y. Lee, R. Y.-C. Yiu, and K.-L. Leung, "Outcome of radical excision of anorectal gastrointestinal stromal tumors in Hong Kong Chinese patients," *Indian Journal of Gastroenterology*, vol. 26, no. 1, pp. 33–35, 2007. View at Google Scholar
15. C. R. Changchien, M. C. Wu, W. S. Tasi et al., "Evaluation of prognosis for malignant rectal gastrointestinal stromal tumor by clinical parameters and immunohistochemical staining," *Diseases of the Colon and Rectum*, vol. 47, no. 11, pp. 1922–1929, 2004. View at Publisher · View at Google Scholar · View at Scopus