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Case Report

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Primary Extra Gastrointestinal Stromal Tumors of Vagina: A Rare Case Report without Gastroenterology Manifestations

Lin Xia[1]; Xuanyu Chen[2]; Jun Zhou[1]; Yun Yan[3]; Limian Song[1]*

[1]Department of Gynaecology and Obstetrics, Union Hospital, Tongji Medical College, Huazhong University of Science and Technology, Wuhan, Hubei Province, China.

[2]Department of Neuroscience and Regenerative Medicine, Medical College of Georgia, Augusta University, Augusta, GA, USA.

[3]Division of Neurogastroenterology/Motility, Medical College of Georgia, Augusta University, Augusta, GA, USA.

Abstract

Background: Primary extragastrointestinal stromal tumors (EGISTs) of vagina without invading gastrointestinal tracts are rare.

Case presentation: Here, we reported the case of a 72-year-old female who presented with vaginal bleeding for 2 weeks and a vaginal mass. MRI showed a 3.05 x 2.87 x 3.86 cm mass on the posterior wall of the vagina. Proctoscopy and gastroscopy showed normal findings. Histological analysis confirmed the presence of a spindle cell type of EGISTs. Immunohistochemical staining of the biopsy indicated positive CD117, CD34, and DOG-1 and negative for S-100, Desmin, and ER. The Ki67 labeling was 15% indicating a relatively low rate of cell proliferation. The subject responded and tolerated imatinib well without surgical removal of mass, and pelvic floor examination showed the absence of a visible vaginal mass at the 3 year-follow up.

Conclusions: Primary vaginal EGISTs unrelated to GI tracts are very rare. The effective utilization of imatinib therapy for treating EGISTs without surgery offers valuable insights into personalized treatment strategies for this disease.

Keywords: Extragastrointestinal stromal tumors; Vagina; c-kit; DOG-1; Imatinib.

Introduction

Gastrointestinal stromal tumors (GISTs) accounting for 1% of primary gastrointestinal malignancy are rare mesenchymal tumors with an incidence of 12 per million [1]. GISTs can manifest in various locations within the digestive tract [2], typically the stomach (60%) and the proximal small intestine (30%), but on rare occasions, may also emerge outside the gastrointestinal tract [3], which are referred to as extra-gastrointestinal stromal tumors (EGISTs) comprising less than 5% of GISTs. EGISTs mostly originate from the mesentery and retroperitoneum [3]. As compared to GISTs, patients with EGISTs have larger size of tumors, higher recurrence rate, and shorter survival time [3]. The recommended approach for treating EGIST without metastasis involves either

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Correspondance: *Limian Song, Department of Gynaecology and Obstetrics, Union Hospital, Tongji Medical College, Huazhong University of Science and Technology, Wuhan, Hubei Province, China.*

Tel: +86-027-84309767, Email: songlimian1985@163.com

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complete surgical removal with clear margins or, as an alternative, enucleation surgery [3]. These methods have proven effective in preventing both recurrence and the spread of the disease to distant sites. In this report, we present a case of primary EGISTs situated in the posterior wall of the vagina, which did not exhibit any gastroenterological manifestations. Remarkably, the patient demonstrated a visible vaginal mass reduction solely through imatinib treatment, without requiring surgical removal of the tumor at its location.

Case report

A 72-year-old woman presented to our clinic with a complaint of vaginal bleeding for approximately two weeks after falling down the stairs. On pelvic exam, a gray-red hard mass located on the posterior wall of the vagina was found. The boundary of the mass was unclear. Transvaginal ultrasound revealed a 3.4x2.7x2.8 cm heterogeneous, hypoechoic mass originating from the posterior wall of the vagina. Further investigations were performed due to suspicion of malignancy. Magnetic resonance imaging (MRI) showed an approximately 3.05x2.87x3.86 cm enhanced scanning unevenness reinforcement tissue mass with significantly limited diffusion on diffusion-weighted imaging (DWI), partially unclear demarcation with the anterior wall of the lower rectum (Figure 1). Proctoscopy and gastroscopy revealed normal findings**.** Other laboratory exams including a complete set of tumor markers, especially CA-19.9, CA-125, CEA, AFP, were within normal range.

Histological analysis of the biopsy confirmed the presence of an EGIST, a spindle cell type, with a mitotic rate of over ten per 5mm^2 (Figure 2). Further immunohistochemical staining showed that the tumor cells were positive for CD117, CD34, and DOG-1, but negative for S-100, Desmin, and ER. The Ki67 labeling was 15%. After the histological confirmation of GISTs, the subject was only prescribed imatinib without surgical resection of the tumor site. Over the course of the 3-year follow-up period, the subject responded well to the treatment and showed good tolerance to imatinib. Importantly, the last pelvic examine indicated no visible vaginal mass.

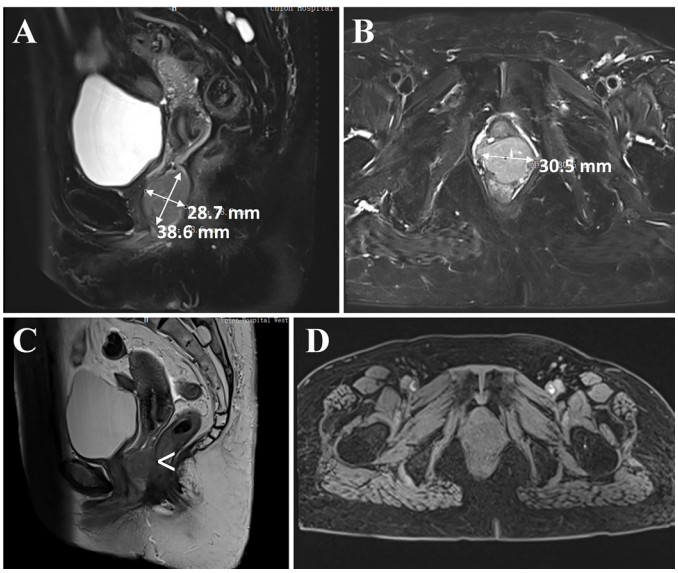

Figure 1: MRI revealed a tissue mass at the back of the vagina, with uneven enhancement and unclear borders with the lower rectum's anterior wall.

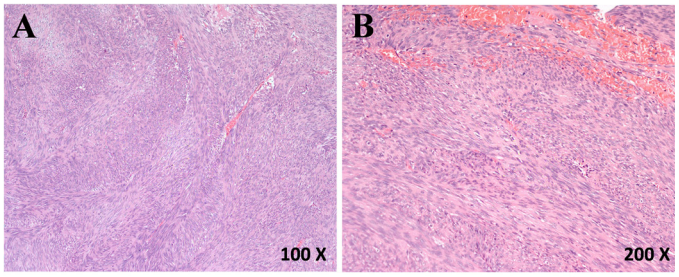

Figure 2: Pathological findings revealed a spindle cell type of extra gastrointestinal stromal tumors, with a mitotic rate of over ten per 5 mm^2 (hematoxylin and eosin staining).

Discussion

Extra gastrointestinal stromal tumors (EGISTs) often originate from the mesentery, peritoneum, scrotum, ovaria, bladder, prostate, and vagina. EGISTs primary from the vagina are particularly rare and often discovered incidentally during surgery or examination due to non-specific symptoms.

The main clinical gynecological manifestations of patients with EGISTs in the vagina include vaginal bleeding, the passage of tumoral tissue from vagina [4] and painless mass [5]. The size of the mass can range from 3 to 8 cm [4,5]. The diagnosis of GISTs is based on the morphology and distinctive immunohistochemical reactions. There are three main types of GISTs [6], spindle cell type (70%), which composes of eosinophil cells and is similar to leiomyoma; epithelioid type (20%) and mixed type (10%). CD117 (C-kit) and GOG-1 have the most diagnostic value in GISTs, and CD117 is not expressed in all non-GIST tumors. One study showed that 100% (20/20) tumor were immunoreactive for c-kit and 100% (10/10) for DOG-1 [7], and another large cohort of population showed that 96.5% (798/27) cases of GISTs exhibited KIT gene mutations [8]. However, a small percentage (5-7%) of GISTs may show negative expression of CD117. In these cases, genetic mutation tests are recommended for accurate diagnosis. Our case showed that tumor cells were positive for CD117, CD34, and DOG-1, and negative for S-100, Desmin and ER. Since S-100 and Desmin are usually positive (70-80%) in leiomyoma and negative for GISTs. This immunopurified confirmed EGISTs rather than a leiomyoma. In addition, we examined the expression of Ki 67, which is correlated with the rate of cell proliferation and metastasis. The Ki67 labeling index of 15% in our case suggested a relatively low rate of cell proliferation.

The majority of the EGISTs cases in the pelvic involved the GI tracts, as shown in a 10-year retrospect pelvic GISTs study where 65% (13/20) cases originated from small bowel, 15% (3/20) originated from rectum, 15% (3/20) originated from stomach and 1 had unavailable primary site information [7]. Another study showed that 56.2% (9/16) of incidentally found GISTs in gynecological surgery were located in the stomach wall, cecum, omentum and mesentery [9]. These cases also presented with GI symptoms, such as abdominal pain [7]. This case is unique as the patient only complained vaginal bleeding without any GI symptoms. Also, although the size of the tumor was relatively small and localized. However, the immunoreactive indicated the tumor was malignant as discussed above. Furthermore, we could not find any abnormal mass in other organs, especially the GI tract.

Surgical removal of tumor is the mainly standard initial treatment for localized GISTs [10], however, it is an adverse prognostic factor because the risk the tumor rupturing during surgery. And the recurrence was as high as 90% after surgical resection [11]. Imatinib therapy has been shown to be effective in treating GISTs by targeting the kit receptors as a C-Kit inhibitor. However, the response and tolerance to imatinib therapy can vary among patients, making individual optimization of treatment crucial for achieving optimal outcomes [12]. In this case, the subject responded well to imatinib therapy alone, as indicated by the absence of a visible mass during the pelvic examination and did not require surgery as initially recommended. Since interruption of imatinib treatment can worse disease progression, it is important to continue imatinib therapy indefinitely for the subject who tolerates it well [12]. These findings underscore the importance of individualizing therapy for GISTs to achieve optimal outcomes.

Conclusion

In conclusion, our case report showed a rare case of EGISTs primary from the vagina without involving GI tracts. Our diagnostic process, which included careful consideration of immunophenotype, played a crucial role in identifying these rare and atypical EGISTs. Additionally, our successful use of imatinib therapy in treating EGISTs without surgery provides valuable insight into the individualized treatment plans for EGISTs.

Declarations

Author contributions: LX and XC analyzed the data and conducted the literature review and manuscript preparation. JZ performed histological analysis of the biopsy. YY reviewed and revised the manuscript. LS designed, acquired, and provided critical feedback and revisions on the manuscript preparation.

Conflict of interest: The authors declare no conflicts of interest.

Availability of data and material: All the data generated or analyzed during this study are included in the article.

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