Gastrointestinal stromal tumors (gist) of the anus: A single institution experience

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Dear Editor,

Gastrointestinal stromal tumors (GIST) of the anus are rare neoplasms regarding which limited data are known as few cases have been reported. Since 2000, only 15 cases of GIST of the anus, verified to have a c-kit mutation, have been published (1). The ideal treatment for anal GIST is unknown. Successful treatment has been reported with methods, such as abdominoperineal resection (APR) and pelvic exenteration, using wide local excision. The utility and efficacy of imatinib mesylate is unknown in the treatment of anal GIST. The aim of this study was to examine our institution's experience with this rare disease, examine its clinicopathologic factors, and observe recurrence patterns. This study was approved by the institution's review board, and all patients consented to the involvement in this research. The institutional review board approval was obtained and patients diagnosed with GIST of the anus from 2000 to 2017 were identified by querying the institution's tumor registry and searching the electronic medical records. The year 2000 was chosen to ensure that c-kit testing was performed. The medical records were reviewed for clinicopathologic data, including patient demographics, presenting symptoms, tumor size, local recurrence, distant metastasis, and treatment(s). Three cases of GIST of the anus positive for c kit mutation were identified in 1 female and 2 males. Patient 1 was aged 68 years at diagnosis and presented with complaints of fecal incontinence. Patient 2 was aged 69 years at diagnosis, and the GIST was noted upon colonoscopy. Patient 3 was aged 72 years at diagnosis, and the GIST was incidentally detected upon a positron emission tomography scan for tonsillar mantle cell lymphoma; this patient also reported fecal incontinence. All patients were noted to have tumors arising from the anal canal upon imaging and physical examination. Patients 1 and 3 received neoadjuvant imatinib followed by APR and transanal resection, respectively. They were subsequently treated with adjuvant imatinib and were reported to be alive without evidence of recurrence after 24 months and 27 months follow-up, respectively. Patient 2 had an endoscopic resection followed by recurrence after 51 months, which was treated with imatinib. A complete response was observed, but local recurrence was noted at 38 months; transanal resection was subsequently performed. No adjuvant therapy was given for 12 months during which imatinib was resumed. Patient 2 developed local recurrence 29 months after transanal excision and was reported to be alive with local recurrence and suspected distant metastasis 128 months after the initial diagnosis. For patients 1, 2, and 3, the tumor size was 3.4 (T2), 1.4 (T1), and 3 (T2) cm, respectively. Mitoses were 0/50, >5/50, and >5/50 high power fields, respectively. All were positive for c-kit and exon 11 mutations. All the patients experienced periorbital edema, and patient 2 developed pancytopenia secondary to imatinib. To our knowledge, this is the largest single institution series of anal GIST verified to have a c-kit mutation and reported to date. This is a rare malignancy with prognosis thought to be similar to GIST occurring in other parts of the digestive tract. An R0 resection and/or imatinib mesylate appear to be the mainstays of treatment. Case series similar to the present are important for sharing experiences of rare malignancies and improving care for patients.

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