Extragastrointestinal stromal tumor of prostate

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INTRODUCTION

Gastrointestinal stromal tumors (GISTs) are the most common mesenchymal tumors of the GI tract. This neoplasm arises from the interstitial cells of Cajal, which are the intestinal pacemaker cells in the gut. Approximately two-thirds of GISTs originate from the stomach and one-fifth from the small intestine, while a few occur in the rectum, colon, or esophagus. Extra GISTs (EGISTs) that originate in the prostate are exceptionally rare. To the best of our knowledge, only five cases have been reported in the literature as primary prostatic GISTs [Table 1]. The aim of our report is thus to present the case of an 84-year-old male with a primary EGIST involving the prostate and to review the literature for enhancing the understanding of this exceedingly rare prostatic disease.

CASE REPORT

An 84-year-old male patient was referred to King Abdulaziz University Hospital with an indwelling urethral catheter to manage his refractory acute urinary retention.
with failure of repeated trials of spontaneous voiding. A review of other systems was unremarkable. The patient was not a smoker but had a history of diabetes mellitus and hypertension. Digital rectal examination revealed a hugely enlarged prostate encroaching upon the lumen of the rectum with a smooth surface, firm in consistency, and nontender. Transrectal ultrasound showed a markedly enlarged prostate with an estimated volume of 360 ml [Figure 1]. Prostate-specific antigen (PSA) was 5.4 ng/ml. In addition, immediate preoperative cystoscopy demonstrated only a moderate enlargement of the prostate, which was disproportionate to its actual size. The patient underwent a transvesical open prostatectomy, and the plane of inoculation could be easily demarcated between the “surgical capsule” and “prostatic adenoma.” Postoperative abdominal computed tomography showed residual prostatic tissue with an estimated weight of 78 g, multiple diffuse colonic diverticulosis with no evidence of diverticulitis, and multiple scattered subcentimeter mesenteric lymph nodes with no evidence of abdominal metastasis [Figure 2]. The visualized osseous structure showed lytic lesions in the left pedicle of the L2 vertebra. Histopathological gross examination of the submitted prostatic tissue showed multiple, irregular, grayish-pink fragments of tissue measuring 17.0 cm × 16.0 cm × 5.0 cm in aggregate. Vague nodularities with foci of hemorrhage and necrosis were seen on a cross section of the tissue. Microscopic examination showed that most of the tissue fragments were colonized by cellular spindle cell neoplastic proliferation with a fascicular growth pattern. The neoplastic spindle cells showed a moderate degree of atypia. There were multiple foci of coagulative tumor necrosis. The mitotic rate was up to 3/50 high-power

![Figure 1: (a and b) Transrectal ultrasound of gastrointestinal stromal tumors originating in the prostate: hugely enlarged prostate with an estimated volume of 360 ml](image1)

![Figure 2: (a-c) Postoperative abdominal computed tomography imaging of gastrointestinal stromal tumors originating in the prostate: lytic lesions seen in the left pedicle of the L2 vertebra](image2)

| Table 1: Review of case reports on gastrointestinal stromal tumors originating in the prostate gland |
|---------------------------------|-------|-------------------------------|-----------------|----------------|----------------|-----------------|----------------|----------------|
| **Title & author**              | **Year** | **Country**                  | **Case report** | **Immunoreactivity** | **Treatment** | **Outcome** | **Metastasis** |
| Gastrointestinal Stromal Tumor of Prostate in an 85-Year-Old Male: Case Report | 2017 | Saudi Arabia                 | An 85-year-old male patient presented with refractory acute urinary retention with failure of repeated trials of spontaneous voiding. | (DOG-1, CD117, and CD34) | Transvesical Open prostatectomy | No recurrence | None |
| Liu Sulai, Primary gastrointestinal stromal tumor of the prostate: A case report and literature review | 2014 | China                       | A 55-year-old male presented to the Hospital with dysuria and urinary frequency that had persisted for approximately 6 months. | (CD34, DOG1, Vim and CD117) | Imatinib Radical prostatectomy | No recurrence | None |
| Zhi-Hong Zhang, A young man with primary prostatic extra-gastrointestinal stromal tumor: a rare case report and review of the literature | 2014 | Tianjin, China              | A 31-year-old man was admitted to our hospital with dysuria. He had frequency, urgency for 4 months and intermittent gross hematuria for 2 weeks. | (DOG-1, CD117, and CD34) | Imatinib | Bad condition | None |
| Yinghao S, Extragastrintestinal stromal tumor possibly originating from the prostate | 2007 | Shanghai, China            | A 49-year-old male was admitted because of vague perineum pain for 10 days. 75-year-old man with dysuria, urinary retention and hesitancy | CD117, CD34, Desmin | Radical prostatectomy | No recurrence | None |
| Lee CH, Gastrointestinal stromal tumor of the prostate: a case report and literature review | 2006 | Taipei, Taiwan             | A 49-year-old man with acute urinary retention | CD117, α-SMA | Transurethral prostatectomy + Radical prostatectomy | Imatinib | Reduced mass volume and liver nodules |
| Van Der Aa F, Gastrointestinal stromal tumor of the prostate | 2005 | Leuven, Belgium          | A 49-year-old man with acute urinary retention | CD117, α-SMA | Transurethral prostatectomy | Good condition | None |
fields (HPFs). Immunohistochemistry showed the neoplastic cells to be diffusely positive for CD117 (c-kit), DOG1, and CD34, while being focally and weakly positive for smooth muscle actin. Results were negative for desmin, S-100, and LMWCK. The Ki-67 index was >10%. The final histopathological diagnosis was a spindle cell neoplasm consistent with a GIST [Figure 3].

**DISCUSSION**

The current case report describes an extremely rare case of primary GIST originating from the prostate. This diagnosis was based on radiological studies, pathological findings, and immunohistochemical results. However, due to the fragmented nature of the specimen, the exact size of the mass and the completeness of resection could not be determined. Various urinary symptoms have been associated with a prostate GIST, and, in our case, severe urinary retention was presented. Pathologic features indicated the diagnosis of a typical prostatic GIST, i.e., an immunoprofile characteristic of spindle cell proliferation that had infiltrated into the rectum. PSA was 5.4 ng/ml, minimally elevated. Although imatinib is widely used in similar cases, it was not considered a suitable treatment for this particular patient. Standard neoadjuvant chemotherapeutic drugs have been used in GISTs but were found ineffective in shrinking the tumor. Therefore, a transvesical open prostatectomy was performed to mitigate symptoms and to establish a pathological diagnosis.

A GIST may arise anywhere in the alimentary tract, and the recognition of a primary GIST is crucial. The clinical presentation of GISTs is erratic. Approximately 70% of patients are symptomatic, while 20% are asymptomatic, and 10% of tumors are detected only on autopsy. Signs and symptoms are not pathognomonic; hence, about half of GISTs are metastatic at the time of diagnosis. GISTs have an uncertain prognosis ranging from benign to frankly malignant. EGIST represents only 5%–10% of all GIST cases. In the literature, affected patients’ ages range from 31 to 75 years (mean 51.8 years) when reported. According to the National Institutes of Health consensus classification scheme for risk stratification in GIST, this particular tumor has an intermediate-to-high risk for aggressive behavior. Since a primary prostatic EGIST is rare, we propose that an extension into the rectum or peri-rectal space should be ruled out. Upper and lower endoscopy should be done to rule out GI involvement, as well as bone scans. At present, size and mitotic count appear to be meaningful predictors of malignant behavior. Tumors with a diameter <5 cm are typically at low risk, while those with a diameter >5 cm are malignant. Mitotic rates <5 per 50 HPFs usually characterize GISTs as benign.

**CONCLUSION**

We believe that this case report will assist clinicians by providing an additional differential diagnosis of prostate tumors, emphasizing the importance of the inclusion of GISTs in the differential diagnosis of a spindle cell lesion encountered in the prostate.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/
have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

REFERENCES
1. Sibley PE, Harper ME, Joyce BG, Peeling WB, Griffiths K. The immunocytochemical detection of protein hormones in human prostatic tissues. Prostate 1981;2:175-85.
2. Appelman HD. Morphology of gastrointestinal stromal tumors: Historical perspectives. J Surg Oncol 2011;104:874-81.
3. Yinghao S, Bo Y, Xiaofeng G. Extragastrointestinal stromal tumor possibly originating from the prostate. Int J Urol 2007;14:869-71.
4. Liu S, Yu Q, Han W, Qi L, Zu X, Zeng F, et al. Primary gastrointestinal stromal tumor of the prostate: A case report and literature review. Oncol Lett 2014;7:1925-9.
5. Zhang ZH, Feng GW, Liu ZF, Qiao L, Zhang T, Gao C, et al. A young man with primary prostatic extra-gastrointestinal stromal tumor: A rare case report and review of the literature. Int J Clin Exp Pathol 2014;7:1764-70.
6. Lee CH, Lin YH, Lin HY, Lee CM, Chu JS. Gastrointestinal stromal tumor of the prostate: A case report and literature review. Hum Pathol 2006;37:1361-5.
7. Van der Aa F, Sciot R, Blyweert W, Ost D, Van Poppel H, Van Oosterom A, et al. Gastrointestinal stromal tumor of the prostate. Urology 2005;65:388.
8. Balachandran VP, Cavnar MJ, Zeng S, Bamboat ZM, Ocuin LM, Obaid H, et al. Imatinib potentiates antitumor T cell responses in gastrointestinal stromal tumor through the inhibition of Ido. Nat Med 2011;17:1094-100.
9. National Cancer Institute. Physician Data Query (PDQ). Gastrointestinal Stromal Tumors Treatment; 2017.
10. Stamatakos M, Douzas E, Stefanaki C, Safioleas P, Polyzou E, Levidou G, et al. Gastrointestinal stromal tumor. World J Surg Oncol 2009;7:61.
11. Emory TS, Sobin LH, Lukes L, Lee DH, O'Leary TJ. Prognosis of gastrointestinal smooth-muscle (stromal) tumors: Dependence on anatomic site. Am J Surg Pathol 1999;23:82-7.
12. Fletcher CD, Berman JJ, Corless C, Gorstein F, Lasota J, Longley BJ, et al. Diagnosis of gastrointestinal stromal tumors: A consensus approach. Hum Pathol 2002;33:459-65.
13. Herawi M, Montgomery EA, Epstein JL. Gastrointestinal stromal tumors (GISTs) on prostate needle biopsy: A clinicopathologic study of 8 cases. Am J Surg Pathol 2006;30:1389-95.