

Case report

Primary and confined gist of the prostate: Case report and review of literature

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The symptoms of prostate hyperplasia, such as tenesmus, nocturia, and urinary retention are common in elderly men. A healthy male patient, 92 years old, in urologic follow up for benign prostatic hyperplasia culminated to acute urinary retention and indication of a retropubic prostatectomy. Exams documented a prostate of 180 grams, PSA 2.45 ng/dl. A digital rectal exam documented grade IV prostate with an adenomatous and fiber elastic aspect, without nodules. Surgery and thorough immunohistochemical studies of the specimen were performed, and primary GIST (gastrointestinal stromal tumor) of the prostate was confirmed. Posterior staging did not document another focus of the illness, and a complete resection of the primary mass was performed. The patient and his family were offered all the treatment options and opted for an exclusive clinical follow up, without adjuvant chemotherapy or radiotherapy. This decision was made based upon all the collateral effects that chemotherapy and radiotherapy would produce with no gain to the patient's quality of life. In patients with localized disease, contingent on individual analysis of performance status as well as social aspects, exclusive clinical follow up can be offered according to their life expectancy.

Keywords: Prostate sarcomas, gastrointestinal sarcomatoid tumors, pelvic syndrome, imatinib, palliative procedures, clinical follow up.

INTRODUCTION

The clinical manifestation of pelvic syndrome is increasing exponentially, as the age of the male population advances. Pelvic syndrome is characterized by symptoms of benign prostate hyperplasia, and by additional symptoms such as nocturia, tenesmus, intermittent voiding, and infra vesical obstruction (Stamatiou 2009).

Gastrointestinal Sarcomatoid Tumors (GISTs) are tumors of mesenchymal origin. GISTs are thought to originate from the Cajal cells, which serve as immunomodulators of the digestive tract (DeMatteo 2000).

These are extremely aggressive tumors, as from the moment of diagnosis up to 47% of patients presents metastases. The primary treatment is based upon the

complete resection of the masses. The extra abdominal tumors account for 10% of the total of masses in extensive series. Surveillance has increased markedly after the introduction of imatinib mesylate, especially in metastatic diseases (Kasper 2006). Primary sarcomas of the prostate represent only 0.1% of all malignant prostatic diseases. There are few publications focused on the association of the prostate and GISTs. In this article, we describe a case of a primary prostate GIST that clinically simulates pelvic syndrome with benign prostatic hyperplasia, submitted to the retropubic resection of the mass.

CASE REPORT

Healthy male patient, 92 years old, in urologic follow up for benign prostatic hyperplasia. Pelvic syndrome developed with symptoms of progressive pelvic

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Figure 1. Pelvic Tomography with a Voluminous Prostate

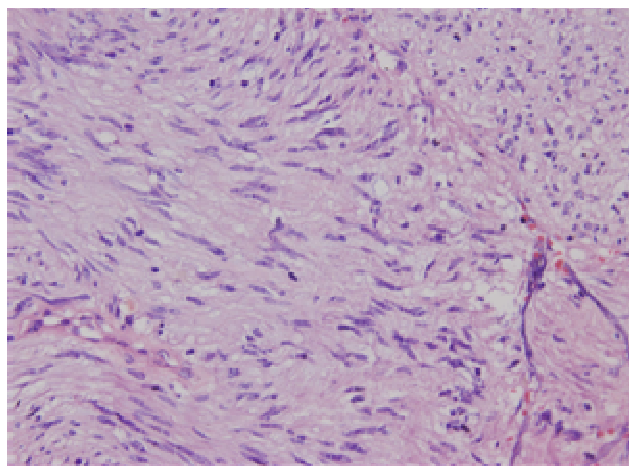


Figure 2. Histopathology, Haematoxylin Eosin, 400X, documents dense tissue, perinuclear vacuolization but with no prostatic acinii or stroma

discomfort, tenesmus, intestinal constipation, fecal bore alterations, and urinary urge, culminating to acute urinary retention. Ultrasonographic exams documented a prostate of 180 grams, and a laboratory PSA of 2.45 ng/dl. Digital rectal exam (DRE) revealed grade IV prostate, adenomatous, fiberelastic, without nodules. The disease did not extend beyond prostate or other rectal mass diagnosed. Abdomen and pelvis CT were obtained with the documentation of a large prostatic mass compressing the rectum but restricted to the organ's capsule (Figure 1). No other abdominal organ was affected by any other disease. Conventional retropubic prostatectomy by Millin's technique was performed without intercurrents. The specimen was a voluminous and friable mass with macroscopic areas of necrosis, weighing 225 grams. The postoperative evolution was satisfactory, as the patient left the hospital in the third postoperative day.

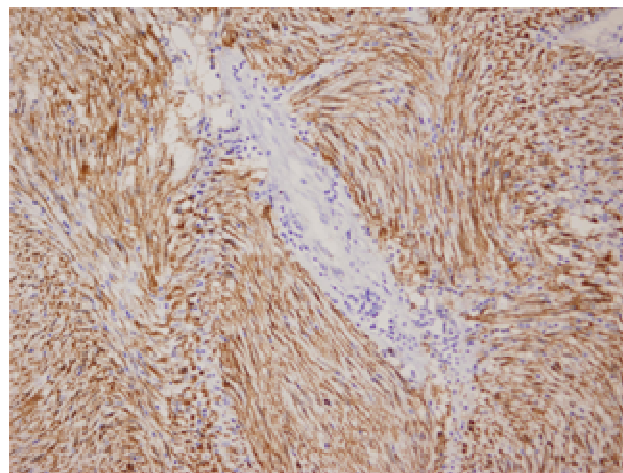


Figure 3. Immunohistochemical reaction to C KIT. Extremely positive 400X, Perinuclear vacuolization and dense fibers arrangement.

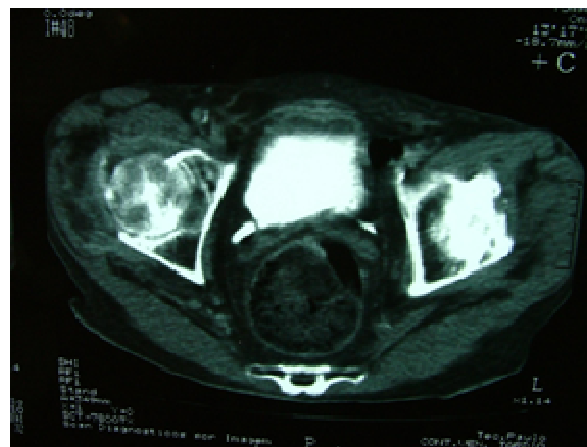


Figure 4. Pelvic Tomography, post operative site with no residual mass

Histopathologic analysis confirmed tissue with an abundant mitotic activity and areas of mixomatous degeneration with an exclusive sarcoma aspect. Molecular analysis documented super expression of the C KIT 117 protein, negative Desmine and S 100 proteins reaction (figures 2 and 3).

Histological and immunohistochemistry studies confirmed highly aggressive GIST confined to the prostate. The classical GIST histological composition is of an arrangement of dense cellular spindle cells in fascicles and perinuclear vacuolization as well as mixomatous stromal degeneration. It is associated the super expression of the protooncogene C-KIT and CD 34, which are not positive in leiomyomas (Miettinen 1998).

New postoperative exams consisting of abdomen and pelvis CT and colonoscopy identified the prostate as the primary site with no signals or symptoms of local recurrence or metastasis (figure 4). Once the diagnosis of

GIST was confirmed and total extirpation of the tumor was performed, treatment options were presented to patient and his family. Recommendation was made for exclusive clinical follow up, with no complementary chemotherapy or radiotherapy. The patient died from natural causes five years after the procedure, without recurrence of the illness.

DISCUSSION

Tumors of pelvic localization can cause pelvic discomfort syndrome with sub intestinal occlusion, mimicking symptoms of benign prostate hyperplasia. At the time of diagnosis it is important to distinguish between colorectal neoplasms, prostate abscesses, and cysts of seminal vesicles (Gasparotto 2008). Since Frank Van Der AA published the first case in 2005, GIST of the prostate, isolated or synchronous to other tumors, has been more frequently diagnosed (Lee 2006).

GISTs are tumors of the gastrointestinal tract. The terminology was introduced in 1983, categorized originally as a large group of mesenchymal tumors of smooth muscle of neurogenic origin which could not be classified in other types and confirmed throughout immunohistochemistry (Saul 1987 and Miettinen 1988). The largest published worldwide experience in GISTs was performed by DeMatteo and colleagues at the Memorial Sloan Kettering Center for Cancer. In this original article of 200 cases, the majority of GISTs originated in the stomach and small intestine. Extra peritoneal sites represent 10%, and in 9% there was no diagnosis of the primary site (De Matteo, 2000). Prostate sarcomas were observed in 0.1%-0.2% of all the malignant prostate neoplasms and of this small group 1 in each 4 are leiomyosarcomas (Miettinen 1998).

Besides being rare, the GIST diagnosis is extremely costly. The main hypothesis is that GISTs originate from mutations of the interstice of the Cajal cells, which are "pacemaker" cells from the gastrointestinal system (Min 2010).

Recent studies also revealed the presence of these cells inside the urinary tract's interstice in smooth muscle cells from bladder, prostate, and urethra. The GIST's histological composition is of an arrangement of dense cellular spindle cells in fascicles with perinuclear vacuolization and mixomatous stromal degeneration. It is associated with the super expression of the protooncogene C-KIT and CD 34, which are not positive in leiomyomas. Of these tumors, 70 to 80% are CD 117 positive, while 30 to 40% are CD 34 positive. Positive results for Desmine and S 100 protein are rare in this case (Saul 1987, Min 2010).

Once the GIST diagnosis is confirmed, it is important to ensure complete surgical resection of the primary mass because, as determined by DeMatteo and colleagues (2000), the overall disease-free surveillance rate is 35%

in the first five years. Further, there is an increase in both recurrence and metastasis when there are residual masses. The partial response to chemotherapy ranges from 12 to 43% with the use of doxorubicin; however, with the introduction of imatinib, a tirosinoquinase inhibitor, the positive response to chemotherapy is increasing up to 64% (Nilsson 2005, Reynoso 2010 and Bakshi 2004). Radiotherapy is ineffective, being indicated only in local recurrences and individual target therapy (Papaetis 2010).

In our opinion, the use of chemotherapy or radiotherapy would be harmful, considering the patient's age. After explaining to the family and patient all adjuvant options, we opted for an exclusive clinical follow up. This decision was based upon the known side effects of chemotherapy and radiotherapy, the complete primary mass resection, and patient's life expectancy.

Conclusion

In conclusion, our group advocates offering exclusive clinical follow up in particular cases where quality of life and localized staging promote adequate management of the tumor.

REFERENCES

- DeMatteo, RP, Lewis JJ, Leung D, Mudan SS, Woodruff JM, Brennan MF, (2000). Two Hundred Gastrointestinal Stromal Tumors, Recurrence Patterns and Prognostic Factors for Survival. *Ann. Surg.* 231: 51- 58.
- Gasparotto D, Rossi S, Bearzi I, Doglioni C, Marzotto A, Hornick JL, Grizzo A, Sartor C, Mandolesi A, Sciot R, Debiec-Rychter M, Dei Tos AP, Maestro R, (2008). Multiple primary sporadic gastrointestinal stromal tumors in the adult: an underestimated entity, *Clin. Cancer Res.* 14: 5715-5721.
- Kasper B, Kallinowski B, Herrmann T, Lehnert T, Mechttersheimer G, Geer T, Ho AD, Egerer G (2006). Treatment of gastrointestinal stromal tumor with imatinib mesylate: a retrospective single-center experience in Heidelberg. *Dig. Dis.* 24:207-211.
- Lee CH, Lin YH., Lin HY, Lee CM, Chu JS (2006). Gastrointestinal Stromal Tumor of The Prostate: A case report and a Literature Review. *Hum. Patol.* 37:1361- 1365.
- Miettinen M, Sarlomo-Kikala M, Lasota J (1998). Gastrointestinal Stromal Tumors. *Ann. Chir. Gynecol.* 87:278-281.
- Min KW (2010). Gastrointestinal stromal tumor: an ultrastructural investigation on regional differences with considerations on their histogenesis. *Ultrastruct. Pathol.* 34:174-188.
- Nilsson B, Bümming P, Meis-Kindblom JM, Odén A, Dortok A, Gustavsson B, Sablinska K, Kindblom LG (2005). Gastrointestinal stromal tumors: the incidence, prevalence, clinical course, and prognostication in the preimatinib mesylate era--a population-based study in western Sweden. *Cancer.* 103: 821-829.
- Saul SH, Rast ML, Brooks JJ (1987). The immunohistochemistry of gastrointestinal stromal tumors. Evidence supporting an origin from smooth muscle. *Am J Surg Pathol.* 11:464-473.
- Stamatiou K (2009). Management of benign prostatic hypertrophy-related urinary retention: current trends and perspectives. *Urol. J.* 6:237-244.
- Van der Aa F, Sciot R, Blyweert W, Ost D, Van Poppel H, Van Oosterom A, Debiec-Rychter M, De Ridder D (2005). Gastrointestinal stromal tumor of the prostate. *Urol.* 65:388.