PROSTATIC STROMAL SARCOMA: RARE CASE REPORT

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Abstract
Prostatic stromal sarcoma is rare and aggressive malignant mesenchymal tumor accounting for 0.1-0.2% of all prostate malignancy. Here we report a case of middle aged male with lower urinary tract symptoms and normal PSA levels. Ultrasonography findings showed grade 3 prostatomegaly. An open prostatectomy specimen was sent and a diagnosis of Prostatic Stromal Sarcoma was made based on histopathological and immunohistochemical findings.

Keywords: Prostate, Stromal sarcoma, mesenchymal

Introduction
Prostate malignancy is the second most common cancer in men. Prostatic stromal sarcoma are rare mesenchmal tumors accounting for only 0.1-0.2% of all malignant prostate tumors. They have a poorer prognosis in comparison with prostate cancer.[1,2] Non epithelial neoplasms of prostate include both benign and malignant tumors specific to the prostate that are specialized stromal tumors. The majority of tumors are characterized by spindle cell pattern with significant overlap in morphological pattern. Use of immunohistochemistry is necessary for accurate diagnosis and treatment. An accurate distinction is warranted because of significant differences in their therapeutic implications.[3] Several authors stated that the symptom of prostatic stromal sarcoma is urinary retention and PSA levels remain at a normal level. Though, with the low incidence, the clinical, investigative information is still unclear.[4]

Case Report
A 52 years old male presents with lower urinary tract symptoms of urinary retention, burning micturition and hematuria since 2 years. Per abdomen examination showed urinary bladder distension upto two fingers below umbilicus. Ultrasonographic findings revealed grade 3 prostatomegaly with prostate measuring 131cc in size. On MRI pelvis, there was a 6x4x4cm heterogeneously enhancing mass lesion involving left side of prostate gland, seminal vesicle and left sided bladder wall. PSA levels was 2.5ng/ml (Normal 0-4ng/ml). Urine examination revealed hematuria and increased leucocytes. Open Prostatectomy was performed. Preoperatively, median lobe of prostate was enlarged. Biopsy was received in the department of pathology in multiple pieces measuring 16x12.5x4cm. Histopathological findings revealed spindle to ovoid stromal cells with mild to moderate nuclear pleomorphism and low mitotic activity. Immunohistochemical study showed progesterone, androgen, CD 34, Vimentin and S100 positivity while CK, PSA, ER was found out to be negative. A final diagnosis of Prostatic stromal sarcoma was made.
Discussion

Primary prostate sarcomas are rare and highly aggressive neoplasm, with limited therapeutic options. Prostate sarcoma occurs more often in younger men, before 40 years, in comparison with prostate cancer, which affects mostly older men. Prostatic sarcoma classified into leiomyosarcoma, rhabdomyosarcoma, malignant fibrous histiocytoma, and unclassified sarcoma. Rhabdomyosarcomas are the most common sarcomas arising in the prostatic stroma of children. Further, unclassified sarcoma can be divided into prostatic stromal sarcoma (PSS) and stromal tumors of uncertain malignancy potential (STUMP) on the basis of mesenchymal invasion, mitosis and extent of mesenchyme overgrowth. The first case of prostatic stromal sarcoma was reported in 1998 by Gaudin. To the best of our knowledge, there are about 30 case reports till 2018. Prostatic stromal sarcoma are rare mesenchymal tumors. Age of the patient ranges from 25 to 86 years. Patient presents with non specific clinical symptoms of urinary retention, hematuria and palpable rectal mass. Due to the lack of typical clinical symptoms, the tumor is easily overlooked or misdiagnosed as benign prostatic hyperplasia, as a consequence, prostate was significantly enlarged when the tumor was discovered in most of the cases. Serum PSA levels are usually normal because of their non-acinar origin. On transrectal ultrasonography, a markedly enlarged volume and irregular margins are important characters of prostate sarcoma. Imaging does not help much, it is neither specific for the type of lesion nor definitely depicts its malignant potential. Limited published data regarding the CT enhancement characteristics of the adult prostate sarcoma showed that tumors had a heterogeneous enhancement or delayed enhanced compared with the normal prostate tissue, whereas the necrosis and cystoids areas were unenhanced. The imaging features of magnetic resonance imaging (MRI) are similar to those of transrectal ultrasonography and CT, apart from, the peripheral solid part of the lesion showed a slightly hyperintense on T1WI and T2WI, a hyperintense on DWI. Both solid or mixed with cystic areas along with areas of necrosis and hemorrhage can be seen grossly in Prostatic stromal sarcoma. However, size does not correlate with the grade and prognosis of the tumor. Histopathological findings are characterized by proliferation of spindle and ovoid stromal cells, some of which poses atypical nuclei, scattered mitotic figures, and necrotic foci. Prostatic stromal sarcoma is further divided into low-grade and high-grade tumors based on moderate to high cellular atypia and hypercellularity in high-grade tumors. It should be noted that both PSPUMP and PSS typically are positive for CD34, vimentin, and express progesterone receptor, but not estrogen receptors. It was also reported that in most cases, a significant increase in the Ki-67 labeling index is observed if a high grade of intraepithelial neoplasia is associated with the tumor. Hence Prostate sarcoma should be included in the differential diagnosis of a large prostatic mass with heterogeneous/ low enhancement of the contrast material and metastasis without lymphadenopathy in a young patient with normal PSA levels. Tumors can invade adjacent organs, with the bladder and rectum being the most common but till now, there is no published case for the involvement of pelvic lymph nodes. Vascular invasion...
and metastases to the liver and lung have also been reported in high-grade sarcomas. [1] Surgery remains the main choice of treatment modality, complete tumor resection with a free margin favors a long-term survival.[7] There is no current data regarding the effectiveness of chemotherapy or radiotherapy, since only limited numbers of cases have been evaluated so far. [7] Prognosis is generally poor, with a medium survival of less than 15 months and only 10% of patients surviving more than 3 years. [7]

Knowledge of the various lesions occurring at this site along with study of morphologic features is the most important tools in the differential diagnosis and making an early and proper diagnosis. [3]

Conclusion

Due to rarity and limited understanding of prostatic stromal sarcoma, they may pose a diagnostic challenge, due to histological overlap between them or their rarity. The ancillary studies including immunohistochemistry (IHC) have often limited utility and the main criteria for diagnosis lies on morphology findings by hematoxylin and eosin (H and E) staining.

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Ethical Approval

Not applicable as it is a case report

References