International Journal of Medical Science and Clinical Research Studies

ISSN(print): 2767-8326, ISSN(online): 2767-8342

Volume 03 Issue 06 June 2023

Page No: 1172-1175

DOI: https://doi.org/10.47191/ijmscrs/v3-i6-27, Impact Factor: 6.597

# Diagnosis and Treatment of Prostatic Sarcoma: Advances in the Understanding of a Rare and Aggressive Disease

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**ABSTRACT**

Prostatic sarcoma is a rare and aggressive disease that accounts for less than 1% of all diagnosed malignant prostate tumors. It mainly affects older men and its epidemiology is still not well defined due to the lack of comprehensive registries. The clinical presentation of prostatic sarcoma is variable, with urinary symptoms, pelvic or lumbar pain and systemic symptoms. Diagnosis requires a comprehensive evaluation including clinical history, physical examination, prostate biopsy and imaging studies. Treatment is individualized and multidisciplinary, including surgery, radiotherapy, chemotherapy and targeted therapies. However, due to the rarity of this disease, evidence on the efficacy of different treatments is limited. More research is needed to better understand prostatic sarcoma, develop more effective therapeutic strategies and improve outcomes for affected patients.

**ARTICLE DETAILS**

**Published On:**
**23 June 2023**

**Available on:**
**https://ijmscr.org/**

## INTRODUCTION

Prostatic sarcoma is a rare malignant neoplasm affecting the soft tissues of the prostate. It is characterized by the uncontrolled proliferation of mesenchymal cells, which are of embryonal origin and differentiate into various types of connective tissue.

Prostatic sarcoma represents only 0.7%[1] of all prostate cancers and is considered a highly aggressive clinical entity. This disease can occur in any age group, although it is more common in older men.

Histologically, prostatic sarcoma can present a wide variety of subtypes, such as leiomyosarcoma, rhabdomyosarcoma and fibrosarcoma, among others. Each subtype has distinctive characteristics in terms of cell morphology and growth patterns.1

It is generally characterized by specialized prostate stromal expansion and a non-neoplastic glandular component, typically expressing CD34 and progesterone receptors.2

The phyllodes tumor is characterized by its infiltrative growth and its potential for metastatic spread to the bladder, rectum, lung, abdominal wall, bone and lymph nodes, rhabdomyosarcoma is a tumor that occurs in childhood, first decade of life, and typically does not express CD34 or progesterone, leiomyosarcoma usually expresses in the immunohistochemical study actin and many of them desmin, it lacks epithelial component, carcinosarcoma is a biphasic tumor, composed of epithelial and mesenchymal malignant elements, it is distinguished by having an epithelial immunophenotype. Low-grade fibromyxoid sarcoma has no associated benign epithelial component and no or only focal expression of CD34.[15]

### Epidemiology

The epidemiology of prostatic sarcoma is characterized by its rarity and low incidence compared to other types of prostate cancer. It constitutes less than 1% of all diagnosed malignant prostate tumors. This disease primarily affects men with an estimated mean age of 45 years, although cases have been reported in various age groups.2

The incidence of prostatic sarcoma varies geographically, and a specific distribution in terms of race or ethnicity has not been established. Cases have been documented in different regions of the world, but the lack of comprehensive epidemiological registries makes it difficult to accurately estimate its prevalence.3

Although the etiology of prostatic sarcoma is not yet fully elucidated, several potential risk factors have been suggested. Some studies have reported a possible association with exposure to ionizing radiation, as well as prior hormone

therapy for prostate cancer. However, more research is needed to better understand these possible connections.4 It has been observed that prostatic stromal sarcomas have an expansion of the specialized prostatic stroma, related to a non-neoplastic glandular component, which expresses CD34 and progesterone receptors, supporting the theory that the lesions are associated with an abnormal hormone-dependent response of the prostatic stroma. A relationship has been observed in the progressive accumulation of p53, suggesting an increase in the clonal dominance of dedifferentiated tumor cells carrying p53 mutations. P53 mutation has therefore been observed as a potential marker of immunoreactivity in the 'pretreatment' stage to predict failure of local treatment with ionizing radiation.5,6,7

Since prostatic sarcoma is a rare disease, information on its clinical presentation pattern and prognostic factors is limited. Symptoms may vary according to the size and location of the tumor, making early detection and accurate diagnosis difficult.5

In addition, prostatic sarcoma is known for its aggressiveness and propensity for metastatic spread, which significantly affects patient prognosis and survival. The overall survival rate is generally low, and the disease is often diagnosed at advanced stages, limiting treatment options and the efficacy of therapeutic interventions.5

**Clinic**

The clinic of prostatic sarcoma is characterized by a series of clinical manifestations that may vary in intensity and presentation depending on the stage of the disease and the location of the tumor in the soft tissues of the prostate.

In the early stages, prostatic sarcoma may be asymptomatic or present nonspecific symptoms that make early diagnosis difficult. As the tumor grows, patients may experience urinary symptoms, such as difficulty urinating, increased urinary frequency, weak or interrupted stream, sensation of incomplete bladder emptying, and presence of blood in the urine (hematuria).5

In addition to urinary symptoms, some patients may experience pain in the pelvis or lumbar region due to tumor invasion into surrounding tissues. The presence of metastases in distant organs, such as bones, lungs or liver, may cause additional symptoms, such as bone pain, shortness of breath or jaundice.

Rectal examination reveals an enlarged prostate and Prostate Specific Antigen is usually in normal values.5

Prostatic sarcoma, due to its aggressiveness and propensity for metastatic spread, can lead to unexplained weight loss, persistent fatigue, generalized weakness, and decreased appetite. These systemic symptoms are indicative of advanced disease and are associated with an unfavorable prognosis. 6

It is important to note that the clinical features of prostatic sarcoma may overlap with those of other prostatic diseases, such as prostatic adenocarcinoma, which can make differential diagnosis difficult. Therefore, a thorough evaluation including detailed clinical history, physical examination, laboratory tests (including prostate-specific antigen - PSA - levels) and imaging studies, such as MRI or CT scan, is crucial to establish an accurate diagnosis.6

The clinic of prostatic sarcoma is characterized by a wide range of clinical manifestations that can affect the urinary system, cause pain in the pelvis or lumbar region, and result in systemic symptoms related to disease progression and metastasis. Early diagnosis and accurate detection are critical to ensure an appropriate therapeutic approach and improve the prognosis of patients affected by this rare and aggressive disease.6

**Diagnosis**

The diagnosis of prostatic sarcoma requires a thorough clinical evaluation and the use of multiple diagnostic tools to establish an accurate differential diagnosis. Due to the rarity and variable presentation of this disease, a multidisciplinary approach involving specialists in urology, oncology, and pathology is crucial.7

They are often missed in transrectal prostate biopsy due to their unusual architecture.[15]

The first step in the diagnosis of prostatic sarcoma involves a thorough evaluation of the patient's medical history, including medical history, presenting symptoms, and potential risk factors. This provides an overview of the disease and helps guide further investigations.7

A detailed physical examination is then performed to evaluate the prostate and detect possible abnormalities, such as palpable masses, changes in consistency or the presence of pain in the prostate region.8

The most commonly used test for the diagnosis of prostatic sarcoma is prostate biopsy, which involves obtaining a sample of prostate tissue for histopathological examination. The biopsy is performed under imaging guidance, such as transrectal ultrasound or MRI, to accurately direct the sampling of suspicious areas. Histopathological evaluation of the tissue allows determination of the presence of malignant cells and helps to classify the subtype of prostatic sarcoma present.8

The diagnosis of choice for this sarcoma is by histopathological and immunochemical report. The most common histological pattern is leiomyosarcoma and rhabdomyosarcoma' . 9

In addition to biopsy, other complementary studies may be performed to assess the extent and stage of the disease. These include blood tests to measure prostate-specific antigen (PSA) levels and other tumor markers, such as lactate dehydrogenase (LDH). Imaging studies, such as MRI, CT scan or bone scan, may also be performed to assess the presence of metastases in other organs or tissues.10

It is important to note that the differential diagnosis of prostatic sarcoma should consider other prostatic diseases, such as prostatic adenocarcinoma, as well as benign tumors or inflammatory processes of the prostate. This is achieved by comparing clinical, histopathological and imaging

findings, and requires the expertise of a specialized medical team.11

In summary, the diagnosis of prostatic sarcoma is based on a comprehensive evaluation that includes clinical history, physical examination, tissue sampling by biopsy, analysis of tumor markers in blood, and imaging studies to assess the extent of disease. A multidisciplinary approach and the expertise of medical specialists are essential to establish an accurate differential diagnosis of prostatic sarcoma.12

Prognosis depends on several factors such as stage, presence of metastasis at the time of diagnosis, with distant metastasis being the main indicator of survival and recurrence. 11

Radical resection with negative margins results in better survival in these patients. 5

Patients with metastases at the time of diagnosis should have systemic chemotherapy added. 10

Despite the aforementioned treatments, survival is no longer than 7 months.

**Treatment**

The treatment of prostatic sarcoma requires an individualized, multidisciplinary therapeutic approach based on the stage of the disease, tumor extent and individual patient characteristics. Because this disease is rare and presents particular challenges, there are no established standard treatment guidelines, and therapeutic strategies are based on clinical experience and the limited evidence available.12

Radical prostate surgery is one of the mainstays of treatment for localized prostatic sarcoma. This surgical procedure seeks complete removal of the tumor and may involve removal of the prostate, seminal vesicles and regional lymph nodes. However, due to the high aggressiveness and propensity for local invasion and metastasis of prostatic sarcoma, surgery may not be feasible in all cases and may require a multimodality approach.13

Radiotherapy is another therapeutic modality used in the treatment of prostatic sarcoma. It can be administered before or after surgery as adjuvant therapy or as the main treatment in patients who are not candidates for surgery. Radiotherapy may include irradiation of the prostate and surrounding tissues with external radiation or implantation of radioactive seeds directly into the prostate (brachytherapy).14

Radiotherapy is not recommended in the first instance, as it has not shown favorable results, since there is disease progression and recurrences.16

Chemotherapy with cisplatin and etoposide or four- to five-cycle cisplatin and doxorubicin is considered a therapeutic option in cases of advanced or metastatic prostatic sarcoma, especially when other approaches have been unsatisfactory. However, due to the lack of well-established clinical trials and the paucity of data on the efficacy of chemotherapy in prostatic sarcoma, its role in treatment is not fully defined.12

In addition to conventional chemotherapy, new targeted therapies and immune therapies are being investigated for the treatment of prostatic sarcoma. These approaches are based on knowledge of specific molecular alterations and the tumor microenvironment to develop drugs that selectively target tumor cells and stimulate the body's immune response.13

Importantly, due to the low incidence and lack of adequate clinical studies, evidence on the efficacy of different treatments in prostatic sarcoma is limited. Therefore, careful evaluation of the potential risks and benefits of each therapeutic option in the context of each individual patient is required, as well as continuous monitoring and follow-up to assess response to treatment and make adjustments accordingly.16

Treatment of prostatic sarcoma involves an individualized, multidisciplinary approach that may include radical prostate surgery, radiotherapy, chemotherapy, targeted therapies and immune therapies. The choice of treatment is based on the stage of disease, tumor extent and patient characteristics, and must take into account the lack of strong clinical evidence due to the rarity of this disease. Close follow-up and the involvement of a specialized medical team are essential to optimize therapeutic outcomes in prostatic sarcoma.17

## CONCLUSION

In conclusion, prostatic sarcoma is a rare and aggressive disease that accounts for less than 1% of all diagnosed malignant prostate tumors. It mainly affects elderly men and its incidence varies geographically without a specific distribution in terms of race or ethnicity.

The epidemiology of prostatic sarcoma is complex due to the lack of comprehensive epidemiologic registries, which makes it difficult to accurately estimate its prevalence and geographic distribution. Further research is needed to better understand the associated risk factors and patterns of clinical presentation of this disease.

The clinical presentation of prostatic sarcoma is variable and may include urinary symptoms, pelvic or lumbar pain, systemic symptoms and distant organ metastases. The diagnosis of prostatic sarcoma requires a comprehensive evaluation involving clinical history, physical examination, prostate biopsy and imaging studies to establish an accurate and differential diagnosis.

The treatment of prostatic sarcoma is based on an individualized and multidisciplinary therapeutic approach, which may include radical prostate surgery, radiotherapy, chemotherapy, targeted therapies and immunological therapies. However, due to the rarity of this disease and the lack of adequate clinical studies, evidence on the efficacy of the different treatments is limited.

Overall, prostatic sarcoma represents a clinical challenge due to its aggressiveness, propensity for metastatic spread, and lack of robust epidemiologic and therapeutic information. Further research and a collaborative approach are needed to improve understanding of this disease, develop more effective therapeutic strategies, and improve outcomes for patients affected by prostatic sarcoma.

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