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**Research Paper**

# Sarcoma of the Prostate – An Uncommon Diagnosis: - A Case Presentation in Aba, South Eastern Nigeria.

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***ABSTRACT***
*The pathological types of prostate cancer include*

- *Adenocarcinoma – about 99% of cases*
- *Neuroendocrine cancer*
  - *Small cell cancer*
  - *Large cell cancer*
- *Transitional cell cancer of the prostate*
- *Squamous cell cancer of the prostate*
- *Sarcoma of the prostate*

*Sarcoma of the prostate is a stromal tumor arising from messenchymal tissues of the prostate with uncertain malignant potential.*
*It has various subtypes. It is rare, accounting for less than 0.1% of prostate malignancies (Yanget et al, 2018, Coudin et al. 1998 and Rojaset et al, 2013)*
*It commonly presents with obstructive symptoms because of its usual large size.*
*Often, the PSA is within normal range but sometimes elevated when in coexistence with BPH, Prostatitis and/or Adenocarcinoma.*
*It tends to present at an advanced stage due to:*

- *Normal PSA level*
- *Nonspecific symptoms*
- *Aggressive behavior.*

*We present the case of a 67 year old patient, who presented with acute urinary retention and subsequent gross haematuria.*
*A prostate biopsy done following abnormal findings on digital rectal examination revealed a sarcoma of the prostate.*

*Subsequently, immunohistochemistry was done with a diagnosis of Malignant Peripheral Nerve Sheath Tumor (MPNST).*
*Multiparametric MRI done revealed a locally advanced tumor.*
*This case highlights the diagnostic difficulties and non availability of standard treatment protocols.*
*This emphasizes the need for comprehensive evaluation of men with persistent lower urinary tract symptoms (LUTS) and normal PSA values and multidisciplinary approach in management.*

***KEY WORDS***
*Prostate, Sarcoma, Malignant peripheral nerve sheath tumor and Aba.*

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## I. INTRODUCTION

Primary prostate sarcomas are rare malignant tumors of the prostate usually of messenchymal origin accounting for less than 1% of all prostate cancers.
Often they present with non specific symptoms but present with obstructive symptoms due to large tumor size.
Sarcomas run an aggressive course often with normal PSA range.
The most common histopathological subtype in adults is Leiomyosarcoma while Rhabdomyosarcoma is the most common subtype seen in children.
Prognosis is generally poor and surgical resection and pelvic exenteration remain the Gold standard of treatment especially in the locally advanced setting.
Malignant peripheral nerve sheath tumors (MPNST) are a variant of sarcoma. They are very aggressive and constitute 5 – 10% of all sarcomas.
About 50% of MPNST are linked to Neurofibromatosis 1 (NF1) mutation and the rest due to prior radiation and sporadic cases.
MPNST tumors may arise De-novo from a peripheral nerve or as a malignant transformation of a pre existing Benign nerve sheath tumor especially neurofibroma.

## II. CASE PRESENTATION

A 67 year old Business man, a Christian of the Jehova's Witness sect, presented with acute urinary retention.
He was immediately relieved of retention by urethral catheterization.
Subsequently, gross haematuria followed after 2 hours but moderate in intensity.
Patient had been well doing his business with no previous lower urinary tract symptoms.
There was no history of smoking, no history of excessive alcohol intake. He took alcohol sparingly.
There was no positive family history and no history of prior radiation. He had no significant medical and surgical history.
The vitals were stable.
A digital rectal examination done revealed features suggestive of malignancy prompting a prostate biopsy following the control of haematuria.
Treatment given included normal saline bladder irrigation, intravenous fluids, intravenous tranexamic acid and Recombinant erythropoietin.
PSA value was 6ng per ml and haemologbin was 7g.
The histopathological result of the biopsy was pleomorphic sarcoma.
He was further investigated by immunohistochemistry and a diagnosis of malignant peripheral nerve sheath (MPNST) was made.
He was stabilized and sent for a staging MRI which showed a capsular breach with extension to the periphery of the prostrate suggesting a locally advanced stage of the cancer.
The outlined treatment was surgical resection with adjuvant external beam radiotherapy.
Patient declined surgery in view of his religious beliefs due to the possibility of blood transfusions.
He was referred for external beam radiotherapy.
However, he was seen 8 months later with generalized metastasis having been advised against radiotherapy by family members.
He died shortly after that.

## III. DISCUSSION

Primary prostate sarcomas are rare malignant tumors of messenchymal origin accounting for less than 1% of all prostate malignancies.
On the other hand, malignant peripheral nerve sheath tumors are a variant of sarcoma.
They are very aggressive soft tissue sarcomas constituting 5-10% of total sarcomas often linked to Neurofibromatosis (NFI) mutation or prior radiation.
Incidence of malignant peripheral nerve sheath tumors (MPNST) is 1:100,000 in the general population and 1:3500 among patients with inherited Neurofibromatosis 1.
MPNST may arise de-novo from a peripheral nerve or as a malignant transformation of a pre-existing Benign Nerve Sheath tumor especially Neurofibroma.
In 50% of cases, MPNST occurs in association with Neurofibromatosis 1 characterized by loss of function /mutation of the tumor suppressor gene Neurofibromin.
The protein Neurofibromin is impaired. Its key function is the function of a tumor suppressor gene through negative regulation of RAS/KAF/MEK/ERK pathways.
The rest of MPNST occur either sporadically or due to previous exposure to radiation.
The median age for NF1 associated MPNST is 20-40 years while that of sporadic MPNST is 30-60 years.
MPNST therefore affects young and middle aged adults and are associated with early metastasis and resistance to chemotherapy.
Yang, Ping et al, in their work on case series of 6 patients with primary sarcoma of the prostate found out that the primary sarcoma of the prostate presents with insidious onset and rapid progression, lacking standard treatment protocols and associated with poor prognosis.
They found that males between 37 – 50 years were commonly affected – significantly younger than the average age at which prostate cancer is diagnosed.
Enes Erul et al, on their work on primary prostatic stromal sarcoma – case report and literature review found out that existing literature reports most patients presenting between 30-40 years of age.
They posited that due to the aggressive nature of the disease, Radical Cystoprostatectomy or Prostatectomy combined with Lymph Node Dissection should be considered as part of the surgical management.
In USA, for locally advanced sarcomas, Neoadjuvant chemotherapy and Neoadjuvant radiotherapy are instituted to reduce tumor size and achieve clear surgical margin.
According to Samantha W-E Knight et al in their work on malignant peripheral nerve sheath tumor (MPNST) – a comprehensive review of pathophysiology, diagnosis and multidisciplinary management affirmed that treatment can be summarized as maximal surgical resection in combination with neuroadjuvant chemotherapy or radiotherapy followed by adjuvant chemotherapy or radiotherapy when positive resection margin exists or metastatic disease exists.
In our index patient, His age of 67 years was unusual for MPNST.
He was not agreeable to surgical management because of the possibility of blood transfusions in view of his religious beliefs.

## IV. CONCLUSION

Primary sarcoma of the prostate is indeed rare.
Malignant peripheral nerve sheath tumor is even a rarer disease.
Because of its rapid progression and aggressive course, early diagnosis and institution of surgical resection is the key to enhancing prognosis.
Again because of its presentation in a much younger age group coupled with normal PSA values, comprehensive evaluation is advocated for males below 40 years presenting with lower urinary tract symptoms.
The use of multidisciplinary management team comprising at least Urologists, Pathologists and Oncologists cannot be overemphasized.

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