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Case Report

Prostatomegaly in a young adolescent: unusual presentation with rare diagnosis

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ABSTRACT

Prostatic mesenchymal tumours are extremely uncommon benign tumors and very few cases have been published. Our case presented with diarrhea and increased urinary frequency with normal serum PSA levels. On multi-parametric MRI diffusely enlarging prostate with loss of zonal anatomy with possibility of inflammatory etiology was suggested. Trus guided biopsies were reviewed, which followed by immunohistochemistry aided in the diagnosis of ganglioneuroma. Surgical resection represents the only choice for both diagnosis and treatment. Because of the benign nature of ganglioneuroma, adjuvant chemo or radiotherapy is not indicated but regular follow-up is necessary for an early risk of potential local recurrence. Prostatic ganglioneuroma is the first case being reported to the best of our knowledge. The aim of the study was to document a newly diagnosed entity at this site and segregate more literature about it.

Keywords: Prostate, Young age, Ganglion cells, Neurons, Mesenchymal tumour

INTRODUCTION

There is an extensive spectrum of mesenchymal neoplasms of the prostate, which account for less than 1% of all prostatic tumors. These include benign as well as malignant tumors that arise from the specialized prostatic stroma like stromal tumors of uncertain malignant potential (STUMPs) and site-specific neoplasms such as smooth muscle tumors, fibrous or myofibroblastic neoplasms, neurogenic tumors, vascular tumors and an unending list of sarcomas.

Some tumors show classic sites of origin within the prostate, like STUMPs commonly involve the peripheral zone at the prostatic base, leiomyomas typically originate from the central prostate toward the apex.[1-3] Other prostatic neoplasms such as gastrointestinal stromal tumors, solitary fibrous tumor, paragangliomas, and neurogenic tumors arise primarily from periprostatic soft tissues. Most mesenchymal tumors of the prostate present as large tumors that causes nonspecific symptoms with normal prostatic specific antigen levels. These mesenchymal neoplasms may demonstrate variable histopathologic and immune-histochemical features along with indistinct imaging findings.

Diagnosis is difficult because of the rarity and lack of awareness of the tumors and the significant overlap in histopathologic features. Accurate diagnosis is essential for optimal management owing to markedly different tumor biology as well as prognostic implications.[4]

CASE REPORT

A 23 year non-diabetic and non-hypertensive male presented to in-patient department with the chief complaints of long standing on and off loose stools since 2 years and increased frequency of micturition for last 6

months. General examination was normal, however on per rectal examination an enlarged prostate was felt. His routine hematological and biochemical investigations were within the normal limits. Serum PSA levels were normal (1.36 ng/ml). Pelvic USG and urosonography revealed irregularly enlarged heterogenous prostatic mass measuring 40.3×58.4×48.5 mm and prostatic volume was-59.7 cc (Figure 1).

Multiparametric MRI was also performed which revealed diffusely enlarged prostatic with loss of zonal anatomy, breach in prostatic capsule with infiltration into adjacent seminal vesicles (Figure 1).

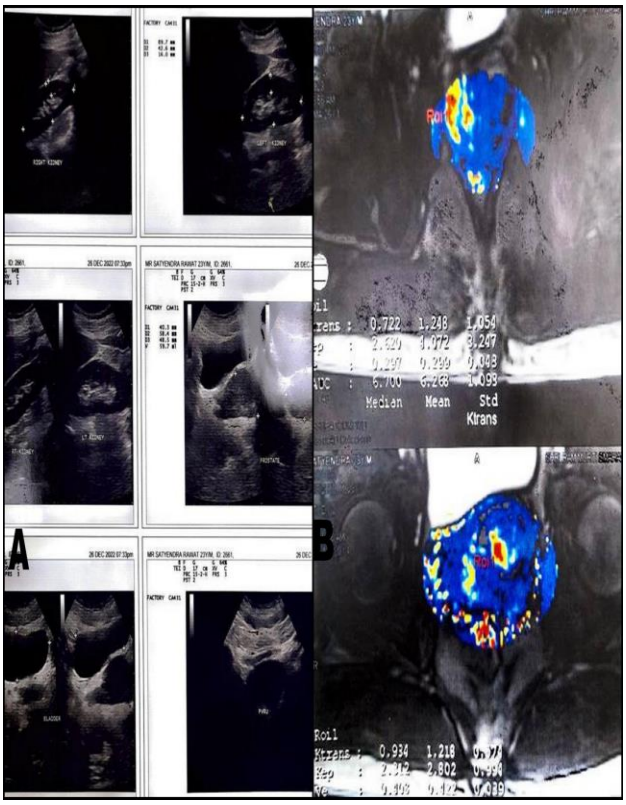

Figure 1: (A) USG showing irregularly enlarged heterogenous prostatic mass measuring 40.3×58.4×48.5 mm; (B) multiparametric MRI was also performed which revealed diffusely enlarged prostatic with loss of zonal anatomy, breach in prostatic capsule with infiltration into adjacent seminal vesicles.

Two ill-defined T2 hypointense lesions were seen in right and left posterior mid zone region. It suggested possibility of inflammatory etiology which needed histological confirmation. Simultaneously mantaoux tuberculin test and widal were done which were negative. Blood, sputum, urine and semen cultures were negative.

After this TRUS guided biopsies were planned and were reported as fibromuscular hyperplasia of prostate. The biopsies were reviewed again and we found bundles of smooth muscle fibres intersected by loosely dispersed, proliferated thin elongated cells in an edematous stroma. The cells had a wavy-spindled nuclei and lightly stained cytoplasm. Few scattered ovoid to polygonal cells with abundant eosinophillic granular cytoplasm and small round nuclei (s/o ganglion cells) were also noted interspersed in it. Surrounding normal glandular acini with corpora amylacea was also seen (Figure 2).

Thus immunohistochemistry was applied and ganglion cells were diffusely positive for NSE, synaptophysin and chromogranin. Background elongated cells and Ganglion cells were positive for S-100 (Figure 2). The stromal cells were negative for CD 34 and CD 117 thus ruling out solitary fibrous tumour and gastrointestinal stromal tumour. Light microscopy followed by immunohistochemistry aided in diagnosing ganglioneuroma. Patient underwent surgical resection as per treatment protocol and no chemotherapeutic agents were administered.

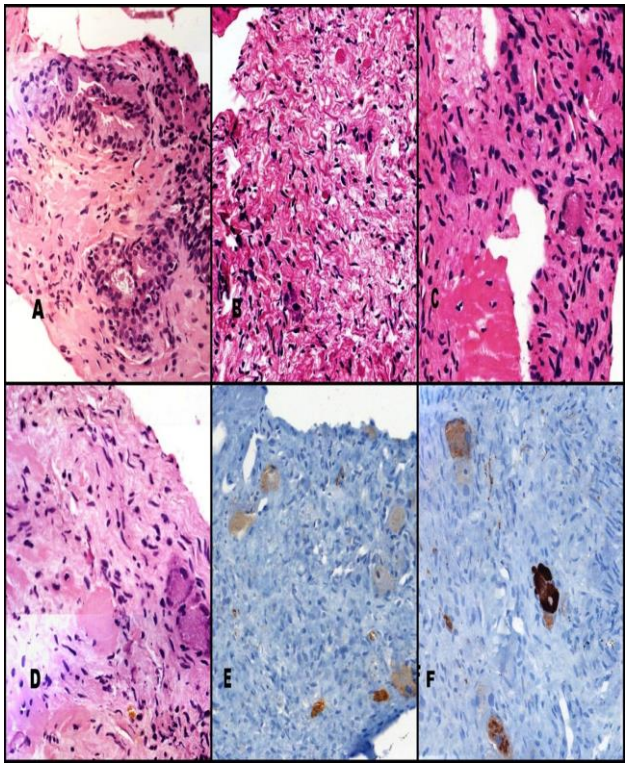

Figure 2: (A) Section show few benign prostatic glands with fibromuscular hyperplasia (40X H and E); (B) section show loosely dispersed, proliferated thin elongated cells in a mildly edematous stroma. (40X H and E); (C) The cells have a wavy-spindled nuclei and lightly stained cytoplasm. Few scattered ovoid to polygonal cells with abundant eosinophillic granular cytoplasm and small round nuclei (s/o ganglion cells) are also noted (40X H and E); (D) other areas displaying ganglion cells with similar morphology (40X H and E); these ganglion cells are uniformly positive for synaptophysin (40X IHC); and (E) these ganglion cells and few stromal cells are diffusely positive for NSE (40X IHC).

DISCUSSION

Ganglioneuromas (GNs) are rare and benign neural tumors arising from the neural crest, Schwannian stroma and connective tissue of paravertebral sympathetic plexus.[5] It belongs to the most differentiated tumour in neuroblastic tumour spectrum.[6] It mainly occurs in the posterior mediastinum or retroperitoneum, but can occur also in other locations like adrenal gland, cervical and parapharyngeal area, urinary bladder, prostate, bone, pancreas, skin, orbit, paratesticular area, appendix, gastrointestinal tract and presacral region.[7,8] Both males and females are equally predisposed. It is usually found in patients between 10-40 years, although any age can be affected.[9]

Intra-abdominal GNs present with pain and compressive symptoms, due to local mass effect on adjacent organs. Gastrointestinal ganglioneuroma may be associated with several inherited diseases like tuberous sclerosis, cowden syndrome, juvenile polyposis and familial adenomatous polyposis.[10] Patients may also present with sweating and hypertension due to elevated catecholamines levels which can be detected as urinary metabolites. Others may produce abdominal cramping and diarrhea due to increased secretion of vasoactive intestinal peptides. Females may manifest virilizing symptoms as a result of raised testosterone-producing Leydig cells. Our case presented with diarrhea initially followed by urinary urgency and had normal catecholamines.

Grossly GNs are well circumscribed lesions which ranges between (3 to >15 cm) with mean size of 7 cm. On serial sectioning tumour appears homogenous grey-white with firm consistency.[11] It is composed of intersecting bundles of Schwann cells and scattered as well as small clusters of ganglion cells in a myxoid or hyalinized stroma. Cells may show mild atypia and multinucleation.

Surgical resection is the best diagnostic and therapeutic choice of treatment. Adjuvant systemic chemotherapy or local radiotherapy are usually not indicated due to benign nature of GNs. They have a tendency to remain silent for a long time, and are often associated with a long-term disease-free survival.[12] However, it should be pointed out that rarely an apparent metastatic focus of GNs can be encountered in a lymph node adjacent to main tumour mass or at a distant site. It is assumed that this represents neuroblastomas in which the metastasis and the primary tumour matured.[13] Rare cases may undergo sarcomatous transformation to a malignant peripheral nerve sheath tumor. Therefore, annual follow-up with neurologic examination and pelvic magnetic resonance imaging is necessary.[12,13]

CONCLUSION

Prostatic GNs are extremely rare benign tumors and to best of our knowledge this one is the first case being reported in the literature. They present with non-specific lower urinary symptoms and slow growing abdominal masses. Cross-sectional imaging modalities, particularly MRI helps in detection, accurate localization and determination of extent of spread. Histo-pathological examination and immunohistochemistry usually allows the diagnosis to be established. Surgical resection is important as it represents the only therapeutic choice.

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