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A Rare Variant of Prostate Cancer in a Young Adult Male of Asian Origin

Authors
Dr Piyush Gupta[1], Dr Uday S Dadhwal[2]
[1]Clinical Tutor (Surgery), Armed Forces Medical College, Pune 411040 India
Email: *piyush_gupta18@rediffmail.com*
[2]Assoc Prof (Surgery), Armed Forces Medical College, Pune 411040 India
Email: *usd999@rediffmail.com*

ABSTRACT

Prostatic cancer is a disease seen commonly in elderly males. Small cell carcinoma of prostate is a rare variant and its occurrence in young men is extremely rare. This aggressive malignancy may develop in previously diagnosed prostatic adenocarcinoma or as a recurrence after surgery or radiotherapy. Thus, it poses a diagnostic dilemma at the time of presentation and even if diagnosed, limits the management in view of rapid loco-regional and systemic spread. This is case report exemplifies one such unusual occurrence and the challenges confronted in small cell carcinoma of prostate in a young adult. The prognosis remains poor as the treatment is yet not standardized. Chemotherapy may have some role in the management.

Keywords: *Carcinoma prostate, Ca prostate in young, Small cell carcinoma prostate.*

Introduction

Prostate carcinoma is the most common non cutaneous cancer among males. The commonest prostatic malignancy is acinar adenocarcinoma that constitutes 95% of all prostatic malignancies. Small cell carcinoma prostate (SCCP) is a rare type of prostatic malignancy which was first described by Wenk et al in 1977.[1] Small cell variety accounts for less than 1% of all prostatic malignancies with an incidence of 0.8% to 1.1%.[2,3] Till date a total of 241 cases have been reported in all age groups of SCCP,[4] with less than 20 cases reported in younger men below the age of 40 years.[5] SCCP is known to have an aggressive course, and both regional and distant metastases are common.

Case

A 20 year male presented with low grade fever for 10 days and sudden onset retention of urine with

overflow incontinence and haematuria of two days duration. He was clinically found to have a grade IV tender prostatomegaly. The prostate felt hard, with irregular surface and mobile overlying rectal mucosa. Blood picture showed leucocytosis (23,000/mm^3 with polymorphs 92%) and numerous pus cells in urine. Serum prostatic specific antigen (PSA) was 70 ng/ml (normal <4ng/ml). On Ultrasonography imaging, a 290 cm^3 heterogeneously enhancing prostate mass compressing the left ureter and rectum was seen. A suprapubic cystostomy was done to relieve the bladder obstruction as urethral catheterization could not be done. Percutaneous nephrostomy was done to relieve left hydroureteronephrosis. He was managed conservatively for acute prostatitis. With antibiotics he showed an improvement in his condition and his fever became low grade. After an initial improvement in symptoms, his condition gradually deteriorated after two weeks of treatment. The fever became continuous, associated with chills and he developed pain radiating to perineum and left thigh. Repeat sonography of pelvis revealed the size of the prostate to be substantially increased to 497 cm^3.

Patient was then taken up for a surgical intervention for a prostatic abscess and transurethral drainage of prostate was attempted. Intra-operatively, the cystoscope could be passed with difficulty due to large size of prostate. The cystoscopic directed prostatic biopsy was taken that showed non friable tissue taken for biopsy during this process the scope got cracked because of the hard tumor. A small amount of purulent fluid was also drained. A subsequent MR imaging of pelvis (Fig 1& 2) revealed the extent of the lesion as a large prostatic mass with multiple pelvic nodes involving obturator and iliac group of lymph nodes and evidence of skeletal metastasis to lumbar vertebrae. Patient deteriorated progressively with development of paraparesis and continuous hematuria. He succumbed to this very aggressive and fast growing malignancy 3 months after initial presentation. Histopathological evaluation revealed a malignant small round cell tumor with endocrine differentiation (Fig 3) which was confirmed on Immunohistochemistry; Vimentin (clone V 9) positive, neuron specific enolase (clone M1 GN3) positive and was negative for Desmin (clone 33), CytoKinins, LCA, SMA, CD99 and chromogranin.

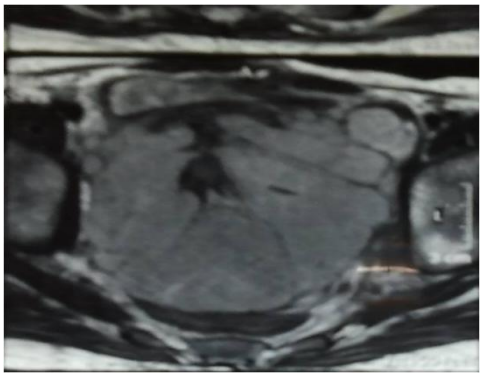

Fig 1

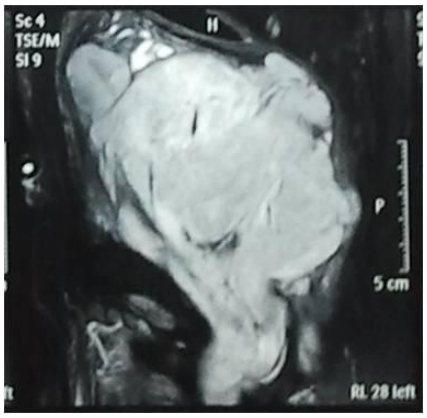

FIG 2

FIG 1 & 2: Cross sectional and sagittal views of MRI pelvis: Large pelvic mass lesion likely to be

of prostatic origin. size – 14 x 10 x 10 cm . Hypointense on T1 and hyperintense on T2 W images. Mass seen to displace urinary bladder anteriorly and encasing lt UV junction. Both seminal vesicles displaced cranially. Soft tissue is seen destroying posterior elements of L3 vertebra with epidural extension. Multiple pelvic lymph nodes noted.

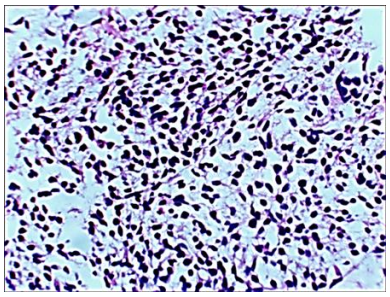

Fig 3. Section reveals aggregates of small round cells with high N:C ratio and hyperchromatic nuclei. No mitotic figures or rosette seen. These aggregates are scattered in a background of smooth muscle. IHC for NSE and vimentin was positive

Discussion

Carcinoma prostate is a disease of elderly and is extremely rare in the young population. The variants of prostate cancer represent 5 to 10% of all carcinomas of prostate. This is the second documentation of such an occurrence from India.[6] SCCP is a high-grade malignant neoplasm with neuroendocrine differentiation. The prognosis of SCCP is poor with a median survival of less than 5 months.[7] Virtually, the entire spectrum of cellular differentiation has been observed within prostatic epithelium, and rare neoplasms exhibit these unusual forms of differentiation as the chief component of the tumour. In most patients with pure SCCP, the PSA level is not elevated despite large metastatic burden. However there are many reports which commensurate to our finding of a raised level of PSA.[8] Unlike conventional prostatic adenocarcinoma, SCCP is known to invade the surrounding organs, regional lymph nodes, and distant organs as noted in this case.

The histological features of SCCP include cell size less than 3 resting lymphocytes, scant cytoplasm with nuclear moulding and finely dispersed vesicular chromatin with absent or small nucleolus. Pathological confirmation of a suspected small cell carcinoma is based on neuroendocrine markers like - chromogranin A, synaptophysin, neuron-specific enolase, and CD56. Typically, one or more of these markers are positive in SCCP. However, in a minority of cases (approximately 10%), all neuroendocrine markers are negative. The treatment guidelines for this rare malignancy are not well defined. The cases presenting in early stages and limited disease would benefit from a radical prostatectomy, however there are isolated reports of beneficial application of etoposide and irinotecan based chemotherapy with external beam radiotherapy. [9,10] Small cell carcinomas do not benefit from androgen deprivation like orchidectomy.[11]

Conclusion

Acute retention, with large hard and irregular prostate may indicate cancer of prostate even in young age. Early careful cystoscopic biopsy is diagnostic. If still localized, surgery or radiotherapy can be attempted in this rare lethal condition

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