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Rare Variants of Prostatic Carcinoma: The Management Approach from Multicentre Experience in Malaysia

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Abstract

Objective: Up to 99% of prostate cancer were adenocarcinoma. Stromal prostate sarcoma only contributes 0.1% of primary prostate malignancy, whereas basaloid cell carcinoma is even rare prostatic neoplasm. We retrospectively evaluated the details of these rare prostatic malignancies, the management approaches, and the clinical outcomes of our case series. **Methods:** A total of six cases with rare variants of prostate malignancy were managed and retrospectively reviewed to evaluate the symptoms at presentation, diagnostic methods, and staging protocol from two tertiary hospitals in Malaysia from 2019 to 2023, comprising four cases of prostate sarcoma and two cases of basaloid cell carcinoma of the prostate. Histopathological findings and treatment modality were analysed. Overall survival was assessed. **Results**: In this study of six patients with rare prostatic malignancies, four had prostatic sarcoma (three had rhabdomyosarcoma and one had high-grade unspecified sarcoma) and two had basaloid cell carcinoma (BCC). The median age at presentation was 48 years for prostatic sarcoma and 35 years for BCC. Clinical presentations included urinary obstruction (n=4), perineal pain (n=1), and gross haematuria (n=1). All patients underwent transrectal ultrasound-guided biopsy and comprehensive staging, including CT, MRI, and bone scans. Treatment varied by disease type. Two patients with localized or locally advanced prostatic sarcoma received radical prostatectomy; one required postoperative chemoradiation due to positive surgical margins. The patient with locally advanced BCC underwent transurethral resection of the prostate (TURP) for symptom relief, followed by adjuvant radiotherapy (50 Gy) and androgen deprivation therapy (ADT). Systemic treatments included vincristine, Adriamycin, and cyclophosphamide for rhabdomyosarcoma, and cisplatin, Adriamycin, and ifosfamide for other sarcomas. The BCC patient received concurrent chemoradiotherapy with carboplatin, paclitaxel, and intensity-modulated radiotherapy (70 Gy). All advanced disease patients were treated with LHRH agonists. The median follow-up was 24 months. Median survival was 43 months for localized or locally advanced cases and 2 months for metastatic cases. Among prostatic sarcoma patients, median survival was 37 months for rhabdomyosarcoma and 1 month for other sarcoma subtypes. BCC patients had a median survival of 14

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months. All patients with localized or locally advanced prostate cancer survived till today, including one prostatic sarcoma patient with positive surgical margin post-prostatectomy with lung metastases during follow-up. Three patients with metastatic disease at presentation experienced death during follow-up. **Conclusion:** Managing rare prostatic malignancies is complex and requires personalized approaches. Radical surgical resection remains the most effective treatment for locally advanced prostatic cancer disease. Systemic treatments offer limited benefits, especially for metastatic cases. Further research is needed to refine management strategies and improve patient outcomes.

Keywords

Sarcoma, Basaloid Cell Carcinoma, Prostate Cancer

Introduction

Prostate cancer ranked as the third most common cancer in Malaysia with a lifetime risk of 1 in 117 men[1]. Up to 99% of prostate cancer were histologically adenocarcinoma. Stromal prostate sarcoma only contributes 0.1% of primary prostate malignancy, whereas basaloid cell carcinoma is even rare prostatic neoplasm. According to the American Joint Committee on Cancer (AJCC), 6 rare histological prostate adenocarcinoma histological variants comprise mucinous, ductal, signet ring cell, adenosquamous, sarcomatoid and neuroendocrine[2]. However, there are limited publications describing the rare variants of prostate cancer specifically sarcoma and basaloid cell carcinoma (BCC). Sarcoma originates from mesodermal tissue and only contributes less than 1% of primary prostate malignancy. Commonly the sarcoma arises from the retroperitoneum or abdomen instead of the prostate. It is associated with poor prognosis with approximately 50% of patients dying of their disease in two years despite surgical intervention. Prognostic factors for tumor recurrence and progression have been identified including tumor grade, size, depth of invasion, and surgical margin status, but because primary prostate sarcoma is rare, clinical variables affecting outcome are primarily based on small case series[3].

Basaloid cell carcinoma (BCC) of the prostate also known as adenoid cystic carcinoma, was first published in 1974 as a distinctive variant of prostate carcinoma. Histopathologically, it is similar to the adenoid cystic tumor of salivary glands, composed of mainly myoepithelial and ductal cells. This rare carcinoma is an aggressive form of poorly differentiated squamous cell carcinoma consisting of medullary patterns of cells with central necrosis. It is difficult to detect histologically, with uncertain behavior, and less than 100 cases are reported in the current literature compared to over a million cases of acinar adenocarcinoma reported yearly.

The rarity of these prostate tumors poses a challenge regarding the treatment options for these prostate cancers. In this study, we aim to report our management approach and clinical outcomes for patients with rare variants of prostate cancer from 2 institutions in Malaysia and evaluate factors that determine the prognosis of malignancy.

Materials and Methods

The Department of Urology, Hospital Tengku Ampuan Afzan, and Hospital International Islamic University Malaysia, Malaysia conducted this descriptive study. All patients diagnosed with rare variants of prostatic carcinoma from January 2019 to December 2023 were included in this retrospective review. The patients' details, clinical presentation, and diagnosis were retrieved from the medical records. Tumor-associated factors such as histology, grading, size, and clinical stage were examined. Treatment-related factors such as the type of operation conducted and the role of adjuvant radiotherapy, chemotherapy, or androgen deprivation therapy were evaluated. Patient survival was analysed from the date of diagnosis till death or until the most recent patient follow-up under the hospital (Table 1). The study was approved by the National Medical Research Register with the research ID (RSCH ID-24-05098-UG7).

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Table 1: Patients' profile

Pt No	Age (Years)	Histology	Size (cm)	Stage	Surgery	XRT	Chemo	Hormonal	Status	Survival (Mos)	Follow-up (Mos)
1	65	BCC	7	Local	TURP	Post-op	Salvage	Yes	Alive	25	25
2	68	BCC	8	Metastatic	No	Palliative	Palliative	Yes	Dead	3	3
3	36	Rhabdomyosarcoma	9	Local	Exenteration	Yes	No	Yes	Alive	60	60
4	35	Rhabdomyosarcoma	10	Local	Exenteration	Post-op	Salvage	Yes	Alive	46	46
5	40	Non-specific carcinosarcoma	10	Metastatic	No	No	Palliative	Yes	Dead	1	1
6	38	Rhabdomyosarcoma	7	Metastatic	No	Palliative	Palliative	Yes	Dead	4	4

Results

Among the six patients reviewed, four had prostatic sarcoma (three with high rhabdomyosarcoma and one with high-grade unspecified sarcoma) and two had basaloid cell carcinoma. The median age at presentation was 48 years for prostatic sarcoma and 35 years for BCC. Clinical presentations varied: four patients presented with urinary obstruction, one with perineal pain, and one with gross hematuria. All patients underwent transrectal ultrasound-guided biopsy. Routine staging included thoracic-abdominopelvic computed tomography (CT), magnetic resonance imaging (MRI) of the prostate, and bone scans.

In this study, we reviewed the outcomes of six patients with rare variants of prostate malignancy, including prostatic sarcoma and basaloid cell carcinoma (BCC). Of these, two patients with localized or locally advanced prostatic sarcoma [Figures 1 and 2] underwent radical prostatectomy. In contrast, one patient [Figure 3] with locally advanced BCC opted for transurethral resection of the prostate (TURP) due to obstructive urinary symptoms, followed by adjuvant radiotherapy.

Among the prostatic sarcoma cases, two patients had localized or locally advanced disease, with tumor sizes exceeding 8 cm. One patient had a basaloid cell carcinoma with tumor infiltration into the bladder neck. The remaining three patients presented with bone and visceral metastatic disease.

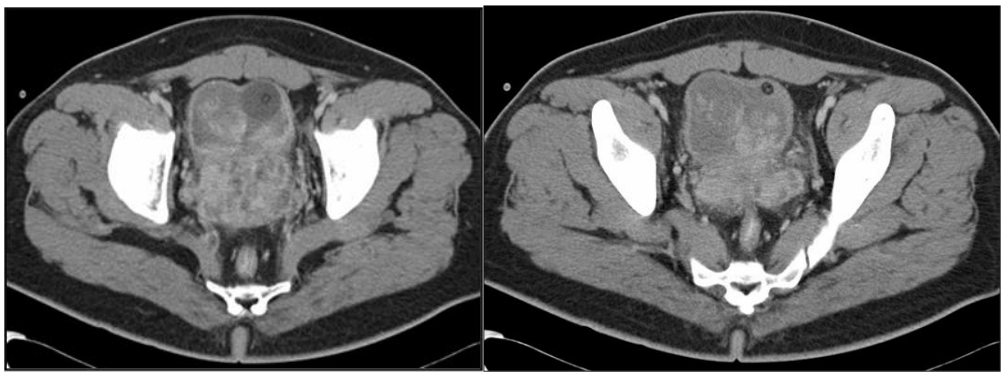

Figure 1: Attached are the axial views of CT TAP for patients with locally advanced prostatic sarcoma infiltrating into and seminal vesicles with bladder clots.

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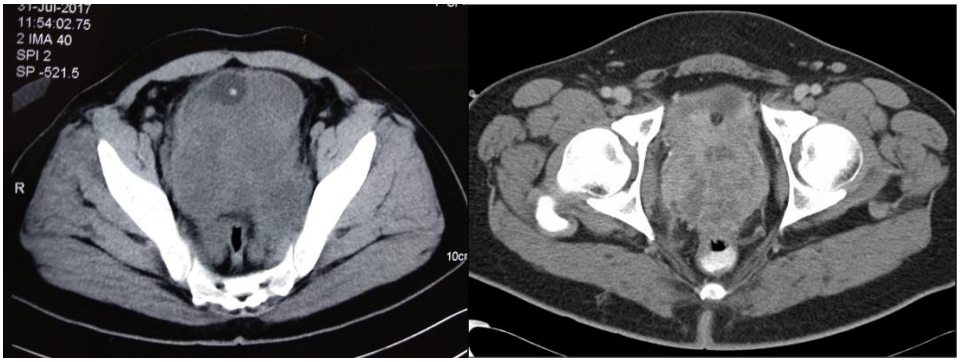

Figure 2: Attached are the axial views of CT for patient with advanced huge prostatic sarcoma infiltrating into bladder and rectum.

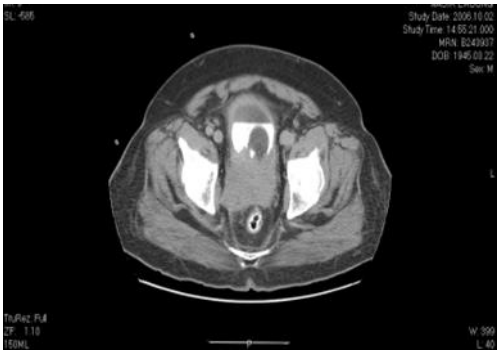

Figure 3: Attached are the axial views of CT for a patient with locally advanced basaloid cell carcinoma of prostate with infiltration into bladder.

For management, two patients with localized or locally advanced prostatic sarcoma underwent radical prostatectomy. One of these patients received postoperative chemoradiation due to a positive surgical margin. The patient with locally advanced BCC was initially considered for radical cystoprostatectomy because of the aggressive nature of the disease. However, he chose to undergo transurethral resection of the prostate (TURP) for symptomatic obstructive urinary relief, followed by adjuvant radiotherapy (50 Gy) and androgen deprivation therapy (ADT) with a luteinizing hormone-releasing hormone (LHRH) agonist. Systemic treatment varied according to the disease subtype. Patients with metastatic prostatic sarcoma received chemotherapy: vincristine, adriamycin, and cyclophosphamide for rhabdomyosarcoma; and cisplatin, adriamycin, and ifosfamide for other sarcoma subtypes. The BCC patient received concurrent chemoradiotherapy with carboplatin and paclitaxel, with a targeted radiotherapy dose of 70 Gy using intensity-modulated radiotherapy. All patients with advanced prostate carcinoma were treated with palliative chemotherapy and LHRH agonists every three months.

The median follow-up period was 24 months. The median survival was 43 months for localized or locally advanced cases and 2 months for metastatic cases. Within the prostatic sarcoma subgroup, median survival was 37 months for rhabdomyosarcoma and 1 month for other sarcoma subtypes. Patients with BCC had a median survival of 14 months. Three patients with metastatic disease at presentation experienced death: The patient with BCC and underwent TURP had oligometastasis 16 months and adjuvant radiotherapy and received palliative chemotherapy and continued ADT. Another patient with prostatic sarcoma, who had positive surgical margins post-radical prostatectomy, developed lung metastases during 26 months of follow-up.

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Discussion

This study provides valuable insights into the management and outcomes of rare prostate carcinoma variants, specifically prostatic sarcoma and basaloid cell carcinoma (BCC). Despite their infrequency, these malignancies present considerable clinical challenges, emphasizing the need for customized management strategies.
Prostatic sarcomas, originating from various tissues within the urogenital system, exhibit diverse clinical presentations. Effective management requires adapting treatment approaches to these varied presentations. According to Mondiani et al., tumor site is a crucial prognostic factor in sarcomas across the genitourinary system, based on an evaluation of 22 cases[3]. Most prostatic sarcoma patients initially present with obstructive urinary tract symptoms due to the tumour's size compressing the urethra. The majority of our patients also presented with urinary obstructive symptoms such as acute urinary retention and poor flow.

Our findings suggest that conventional diagnostic tools, such as prostate-specific antigen (PSA) assays, are less effective for diagnosing and monitoring these rare variants. The PSA marker, generally reliable for adenocarcinomas, was ineffective for prostatic sarcoma and BCC due to the absence of epithelial components in these tumours. This observation aligns with previous studies indicating that these variants often exhibit distinct biological behaviours not reflected by PSA levels[4].

Imaging techniques, including computed tomography (CT) and magnetic resonance imaging (MRI), remain essential for staging and monitoring. MRI is particularly valuable for assessing soft tissue planes and aiding surgical planning. However, our study found that PSMA positron emission tomography (PET) scans had limited utility in these cases, likely due to the unique pathophysiological characteristics of rare prostatic malignancies compared to more common types. Interestingly, despite a positive surgical margin for the patient post-prostatectomy, the PSMA scan showed no evidence of local or distant metastases. This scenario proves that the PSMA scan has a limited role in detecting local or distant recurrence for rare variants of prostatic malignancy.

Histological subtypes of prostatic sarcoma are significant prognostic indicators. For example, adults with non-rhabdomyosarcoma subtypes generally experience poor outcomes, especially at metastatic stages. Our data indicate that patients with non-rhabdomyosarcoma sarcomas had a median survival of only one month compared to four months for patients with other subtypes, despite both groups receiving palliative chemotherapy. This finding is consistent with Sexton et al.'s observations that tumor grade and size do not significantly impact outcomes in these cases[4].

Our results suggest a potential survival advantage for patients with positive surgical margins who receive a multimodal treatment approach compared to those who undergo surgery alone. Sexton et al. also noted possible benefits from combined modality therapy. Dundore's review of carcinosarcomas proposed that lung metastases are more prevalent in carcinosarcomas than in adenocarcinomas, suggesting hematogenous rather than lymphatic dissemination[4]. Thus, both aggressive local and systemic therapies are warranted. Hence, our patient with distant metastasis post radical prostatectomy was offered chemotherapy and he is alive till now with static disease during follow-up.

Treatment outcomes varied significantly among our cases. Radical prostatectomy proved effective for localized or locally advanced prostatic sarcoma and BCC, particularly when negative surgical margins were achieved. Patients with locally advanced prostate carcinoma who underwent radical prostatectomy showed improved overall survival compared to those receiving alternative treatments. Notably, the patient with BCC who had a transurethral resection of the prostate (TURP) followed by adjuvant radiotherapy faced a higher risk of local recurrence, highlighting the potential advantages of radical cystoprostatectomy for managing locally advanced disease. While some patients with localized prostatic sarcoma initially achieved disease-free status, all eventually succumbed to metastatic disease within a few years[4].

Given BCC's rarity, there are no established treatment standards due to limited clinical experience. Cozzi et al encompassed 106 patients from thirty-three studies, reflecting various management approaches[6]. Radical surgical resection remains the primary treatment for localized or locally advanced BCC. Poor prognostic factors for BCC, such as positive surgical margins, perineural invasion, and local tumor infiltration into adjacent organs, often necessitate postoperative radiotherapy[7]. In our study, a patient with

BCC, whose tumor had infiltrated surrounding seminal vesicles and the rectum, opted against radical cystoprostatectomy and was thus treated with postoperative radiotherapy after TURP.

For patients with advanced disease, prognosis remains generally poor. We observed limited efficacy of androgen deprivation therapy and chemotherapy in those with metastatic prostatic sarcoma and BCC. Notably, patients with advanced prostatic sarcoma had a significantly shorter median survival compared to those with BCC, reflecting the more aggressive nature and poorer response to systemic therapies associated with sarcoma.

Conclusion

Managing rare prostatic malignancies is complex and requires personalized approaches. Radical surgical resection remains the most effective treatment for localized disease. Systemic treatments offer limited benefits, especially for advanced cases. Further research is needed to refine management strategies and improve patient outcomes.

References

1. Lim J, Malek R, Jr S, Toh CC, Sundram M, Woo SYY, Yusoff NAM, Teh GC, Chui BJT, Ngu IS, Thevarajah S, Koh WJ, Lee SB, Khoo SC, Teoh BW, Zainal R, Tham TM, Omar S, Nasuha NA, Akaza H, Ong TA; M-CaP Study. Prostate cancer in multi-ethnic Asian men: Real-world experience in the Malaysia Prostate Cancer (M-CaP) Study. Cancer Med. 2021 Nov;10(22):8020-8028. doi: 10.1002/cam4.4319.
2. Edge SB, Byrd DR, Compton CC, Fritz AG, Greene FL, Trotti A (eds). The AJCC Staging Manual, 7th edn. New York, NY, Springer, 2007.
3. Bronkema C, Arora S, Sood A, Dalela D, Keeley J, Borchert A, et al. Rare Histological Variants of Prostate Adenocarcinoma: A National Cancer Database Analysis. Journal of Urology ;204(2):260–6.
4. Morse MJ, Whitmore WF. Genitourinary sarcomas in adults. In: Shiu MH, Brennan MF, eds. Surgical Management of Soft Tissue Sarcomas. Philadelphia, PA: Lea and Febiger; 1989:189 –207.
5. Sexton WJ, Lance RE, Reyes AO et al. Adult prostate sarcoma. Journal of Urology :2001. 166:521-525.
6. Dundore, P. A., Cheville, J. C., Nascimento, A. G., Farrow, G. M., & Bostwick, D. G. (1995). Carcinosarcoma of the prostate. Report of 21 cases. Cancer, 76(6), 1035-1042.
7. Cozzi, S.; Bardoscia, L.; Najafi, M.; Botti, A.; Blandino, G.; Augugliaro, M.; Manicone, M.; Iori, F.; Giaccherini, L.; Sardaro, A.; et al. Adenoid Cystic Carcinoma/Basal Cell Carcinoma of the Prostate: Overview and Update on Rare Prostate Cancer Subtypes. Curr. Oncol. 2022, 29, 1866–1876. https://doi.org/10.3390/curroncol29030152
8. Tsuruta, K.; Funahashi, Y.; Kato, M. Basal cell carcinoma arising in the prostate. Int. J. Urol. 2014, 21, 1072–1073.