

Sarcomatoid carcinoma of prostate involving the whole lower urinary tract and rectum

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Abstract Sarcomatoid carcinoma of prostate is an extremely rare but aggressive neoplasm. It is generally associated with a poor prognosis. About 100 cases have so far been reported in the English literature. We report the case of a 64-year-old male with a very rapidly progressive disease that ultimately involved the whole lower urinary tract and rectum. The management of this case along with etio-pathogenesis and literature review is discussed.

Key Words: Carcino-sarcoma, prostate, sarcomatoid carcinoma

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INTRODUCTION

Sarcomatoid carcinoma of prostate is a rare but aggressive neoplasm comprising of an admixture of both epithelial and mesenchymal components. Until now less than 100 cases has been reported so far in the English literature in the form of case reports or small case series.^[1] The origin of this tumor is uncertain and there is debate whether these are originating from a single malignant transformation event with evolution of high grade adenocarcinoma into sarcoma or a simultaneous occurrence of two malignant processes, i.e., collision of epithelial and mesenchymal elements.^[2,3]

Because of the paucity of cases, little is known about the optimal treatment strategy and it has an extremely poor prognosis.^[3,4] Surgical resection has been recommended for the localized diseases and non-surgical treatments (Hormonal, radiation or chemotherapy) are generally associated with poor outcomes.^[5]

Herein we report a case of sarcomatoid carcinoma of prostate which occupied the entire true pelvis involving whole lower urinary tract and rectum and extending into the bulb of penis.

CASE REPORT

A 64-year-old man presented to his urologist with complaints of voiding symptoms and urinary retention. The initial prostate specific antigen (PSA) level was 9 ng/ml. He was treated with transurethral resection of prostate (TURP) outside our institute which showed high grade prostatic adeno-carcinoma (Gleason score 4 + 5 = 9). The staging workup subsequently showed a locally advanced prostate cancer. He was also started on hormonal therapy with leutinizing hormone releasing hormone (LHRH) analogue. Two months after initial surgery, he again went into painful urinary retention and per urethral catheterization failed, so a re-do TURP was done. The histopathology confirmed it to be adeno-carcinoma with sarcomatoid variant.

In the next six weeks, his voiding symptoms got worse again with gross hematuria culminating into urinary retention and obstructive uropathy, so a supra pubic catheter and bilateral percutaneous nephrostomy (PCN) tubes were placed and he was referred to us for further management. The PSA at

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this stage was 0.5 ng/ml. The patient also developed severe constipation with deep seated pelvic and perineal pain.

On physical examination, a stony hard mass was felt in suprapubic area through which suprapubic catheter was coming out. Another hard mass was felt in the perineum involving the bulb of penis. The digital rectal examination revealed a huge irregular stony hard mass with total occlusion of rectal lumen. An abdomino-pelvic magnetic resonance imaging (MRI) was done that showed an abnormal heterogeneous signal intensity mass lesion in the pelvis showing peripheral post contrast enhancement with central necrotic component measuring $13 \times 11 \times 14$ cm in antero-posterior (AP), transverse and cranio-caudal dimensions and extending down into the perineum where it measures $8.9 \times 3.8 \times 5.2$ cms involving the root of penis. Superiorly it was infiltrating into urinary bladder completely filling its lumen, anteriorly abutting the symphysis pubis, posteriorly involving rectum with loss of fat planes and laterally extending up to lateral pelvic wall [Figure 1]. No metastatic lesion was found on computerized tomography (CT) scan of chest and bone scans.

Because of extensive involvement of whole pelvis with severe urinary- and bowel-related symptoms, a total pelvic exenteration with pelvic lymphadenectomy along with urinary and fecal diversion with ileal conduit and an end colostomy formation was done. The pelvic tumor was completely resected in two pieces because of difficult dissection without any spillage and gross evidence of disease at the end of procedure. The residual preineal part of tumor was dealt with an elliptical perineal incision and it was removed from pelvic floor with a wide margin of normal tissue along with total penectomy. A margin of tissue was also resected from anterior abdominal wall along the cystostomy tract [Figure 2].

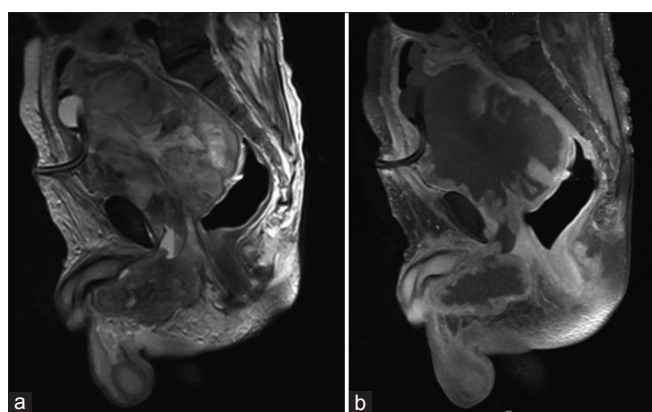


Figure 1: MRI sagittal view (a) Pre contrast (b) Post contrast. A large heterogenous mass with central necrosis that is infiltrating urinary bladder. Posteriorly abutting the rectum and inferiorly extending upto root of penis. It is Iso-intense on T1WI and showing peripheral enhancement on post contrast images. Note the prostate is totally replaced by the mass

The pelvic floor was repaired and re-enforced with a vicryl mesh. Hence, complete gross clearance was done with no palpable or visible disease.

The final histopathology revealed a mixture of carcinomatous and sarcomatous components comprising of diffuse sheets and aggregate of oval and spindle shaped cells arranged in short interlacing fascicles with areas of chondroid, osteogenic, and squamous differentiation. Cytokeratin AE1/AE3 and cytokeratin cam 5.2 immunohistochemical stains were performed on representative blocks, which showed focal positivity. PSA staining was negative. Two of recovered pelvic lymph nodes showed tumor involvement [Figure 3].

The patient made a wonderful recovery and is still alive 4 months after surgery with no evidence of disease recurrence and metastasis on subsequent imaging (CT scan chest, abdomen, and pelvis).

DISCUSSION

Prostatic adenocarcinoma is the most common malignant neoplasm in elderly population. Sarcomatoid carcinoma of prostate is a rare tumor and accounts for less than 0.1% of all prostatic malignancies.^[6] It shows biphasic growth pattern with poorly differentiated epithelial and mesenchymal components and this term has been used interchangeably with the term carcinosarcoma. However, both can be separately defined based upon the appearance of mesenchymal elements as homologous (undifferentiated spindle cell sarcoma) or heterologous (synonymous with carcinosarcoma) with differentiation of sarcomatous component along the lines of specific lineage such as bone or cartilage.^[7,8] Nonetheless, this

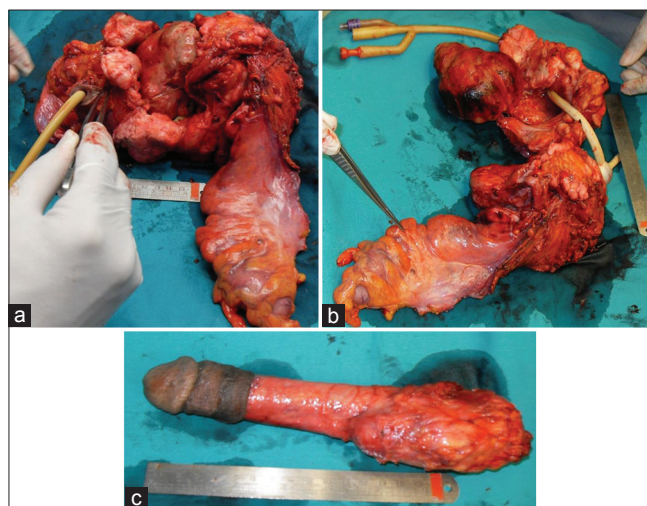


Figure 2: (a) Gross specimen after total pelvic exenteration. Note the main bulk of tumor and the excised tract along the suprapubic catheter. (b) Another view showing involvement of rectum by the tumor. (c) Total penectomy specimen with attached tumor removed from perineum

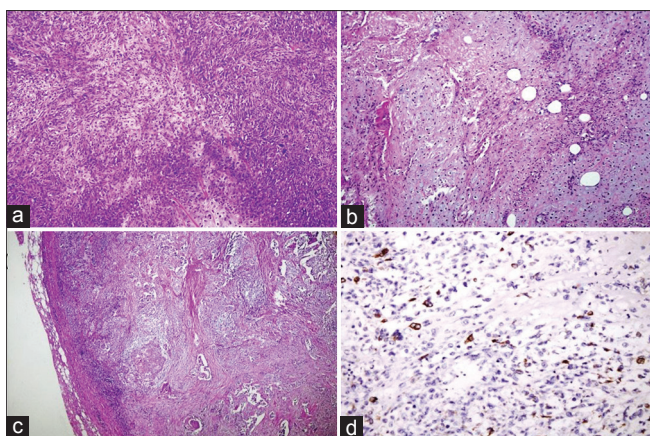


Figure 3: (a) ×4 magnification. H and E stained slide. Sarcomatoid carcinoma showing a predominant spindle cell population with pleomorphic cells arranged in interlacing fascicles. (b) ×10 magnification. H and E stained slide. Areas of chondroid differentiation. (c) ×4 magnification. H and E stained slide. Lymph node showing tumor metastasis. (d) ×10 magnification. Cytokeratin cam 5.2 immunostain showing positivity in scattered tumor cells

histological characteristic of sarcomatous component is of little clinical or prognostic significance^[9,10] and recent World Health Organization (WHO) classification uses the term sarcomatoid carcinoma to denote all these lesions.^[11]

There are many theories of its histogenesis. These include (1) simultaneous development of both sarcoma and carcinoma from different regions of same prostate, (2) dual differentiation of malignant precursor (totipotential) cells,^[12] (3) transformation of adenocarcinoma into sarcoma and vice versa,^[10,13] and (4) de-differentiation of tumor due to effect of hormonal or radiation treatment.^[8,13,14] However, loss-of-heterozygosity (LOH) analysis studies on tissue samples have shown that both the malignant epithelial and mesenchymal components are clonally related, supporting the hypothesis of a single malignant transformation process.^[3,11]

In our case, there was only high grade adenocarcinoma in previous TUR and subsequent pelvic exenteration specimen showed carcinomatous component mixed with the sarcomatous component. Moreover, our patient did not receive any radiation or hormonal therapy, therefore, we postulate the transformation of adenocarcinoma to sarcoma.

In majority of reported cases, there is history of prior acinar adenocarcinoma of prostate and time between original diagnosis to sarcomatoid carcinoma is between 6 months to 16 years;^[15] however, in our case the time interval was much shorter (3 months) explaining a very aggressive disease process.

The previous reported cases have shown epithelial component to be high grade adenocarcinoma. Morphologically, the

sarcomatoid component can comprise from 5-99% of total specimen^[15] and these are composed of undifferentiated spindled and pleomorphic cells arranged in the form of fascicles and sheets. Various subtypes of sarcoma can also be found including osteosarcoma (50%), chondrosarcoma (33%), and leiomyosarcoma (17%), and rarely rhabdomyosarcoma, malignant fibrous histiocytoma and fibrosarcoma.^[5] Our patient mainly had undifferentiated sheets of large spindle cells with few areas of osteogenic, chondroid, and squamous differentiation.

The majority of patients with this disease present with obstructive voiding symptoms. Dundore *et al.* from Mayo clinic in their largest series of 21 patients reported subsequent diagnosis of carcinosarcoma in 18 patients who had prior TURP secondary to obstructive symptoms.^[16] An equal number of patients suffering from sarcomatoid carcinoma have normal and higher PSA levels. The mean age of our patient (64 years) is identical to that reported in their series and cumulative literature, i.e., 66 years.^[16,17]

The disease shows an aggressive course with local and systematic spread. Because of extensive rapid growth, nearly half of the patients develop urinary and fecal obstruction.^[18] and severe pelvic or perineal pain due to involvement/compression of pelvic nerves.

Various therapeutic modalities such as radiotherapy, hormones, chemotherapy, and surgery have been used for these patients with disappointing results. Many investigators believe surgical removal to be the first line therapy with potential cure.^[7,16] These approaches depend upon the extent of disease and include radical surgery (Radical retro pubic prostatectomy, cystoprostatectomy or pelvic exenteration). Due to the severe local symptoms, i.e., gross hematuria, obstructive uropathy, sub acute bowel obstruction and sever pelvic and perineal pain, we opted for the pelvic exenteration and managed to get a complete gross clearance of entire pelvis without any visible or palpable disease.

Patients with sarcomatoid carcinoma have poor outcomes and mean survival period in the reported literature is extremely short (about 7 months).^[18] No parameters such as age, histologic subtype, percentage of necrosis, sarcoma grade or Gleason grade of adenocarcinoma and prior history of radiation or hormonal therapy has been found to be predictive of outcome.^[16]

CONCLUSION

Sarcomatoid carcinoma of prostate is a rare but highly aggressive disease with dismal prognosis. The role of neoadjuvant or adjuvant chemo-radiation is not defined and

patients should be managed with surgery and symptomatic treatment.

REFERENCES

1. Zizi-Sermpetzoglou A, Savvaidou V, Tepelenis N, Galariotis N, Olympitis M, Stamatou K. Sarcomatoid carcinoma of the prostate: A case report. *Int J Clin Exp Pathol* 2010;3:319-22.
2. Grignon DJ. Unusual subtypes of prostate cancer. *Mod Pathol* 2004;17:316-27.
3. Ray ME, Wojno KJ, Goldstein NS, Olson KB, Shah RB, Cooney KA. Clonality of sarcomatous and carcinomatous elements in sarcomatoid carcinoma of the prostate. *Urology* 2006;67:423.e5-423.e8.
4. Huan Y, Idrees M, Gribetz ME, Unger PD. Sarcomatoid carcinoma after radiation treatment of prostatic adenocarcinoma. *Ann Diagn Pathol* 2008;12:142-5.
5. Subramanian VS, Coburn M, Miles BJ. Carcinosarcoma of the prostate with multiple metastases: Case report and review of the literature. *Urol Oncol* 2005;23:181-3.
6. Melicow MM, Pelton TH, Fish GW. Sarcoma of the prostate. Review of the literature, table of classifications, and report of four cases. *J Urol* 1943;49:675-707.
7. Mazzucchelli R, Lopez-Beltran A, Cheng L, Scarpelli M, Kirkali Z, Montironi R. Rare and unusual histological variants of prostatic carcinoma: Clinical significance. *BJU Int* 2008;102:1369-74.
8. Mostofi FK, Price EB Jr. Tumors of the male genital system. In: *Atlas of Tumor Pathology. Series 2, Fascicle 8.* Washington DC: Armed Forces Institute of Pathology; 1973. p. 177-258.
9. Rogers CG, Parwani A, Tekes A, Schoenberg MP, Epstein JI. Carcinosarcoma of the prostate with urothelial and squamous components. *J Urol* 2005;173:439-40.
10. Quay SC, Proppe KH. Carcinosarcoma of the prostate: Case report and review of the literature. *J Urol*. 1981;125:436-8.
11. Eble JN, Sauter G, Epstein JI, Sesterhenn IA. editors. *The World Health Organization Classification of Tumors of the Urinary System and Male Genital Organs.* Vol. 160. Lyon, France: IARC Press; 2004. p. 209-11.
12. Kubosawa H, Matsuzaki O, Kondo Y, Takao M, Sato N. Carcinosarcoma of the prostate. *Acta Pathol Jpn* 1993;43:209-14.
13. Lauwers GY, Schevchuk M, Armenakas N, Reuter VE. Carcinosarcoma of the prostate. *Am J Surg Pathol* 1993;17:342-9.
14. Sak SD, Orhan D, Yaman O, Tulunay O, Ozdiler E. Carcinosarcoma of the prostate: A case report and a possible evidence on the role of hormonal therapy. *Urol Int* 1997;59:50-2.
15. Hansel DE, Herawi M, Montgomery E, Epstein JI. Spindle cell lesions of the adult prostate. *Mod Pathol* 2007;20:148-58.
16. Dundore PA, Chevillie JC, Nascimento AG, Farrow GM, Bostwick DG. Carcinosarcoma of the prostate. Report of 21 cases. *Cancer* 1995;76:1035-42.
17. McGee SM, Boorjian SA, Karnes RJ. Carcinosarcoma of the prostate replacing the entire lower genitourinary tract. *Urology* 2009;74:540-1.
18. Fukawa T, Numata K, Yamanaka M, Miyamoto T, Kurokawa Y, Kanayama HO, *et al.* Prostatic carcinosarcoma: A case report and review of literature. *Int J Urol* 2003;10:108-13.

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