JOURNAL OF CLINICAL AND DIAGNOSTIC RESEARCH

HOW TO CITE THIS ARTICLE:
MANNAN R , CHUFAL SS , MISRA V,SINGH PA. SARCOMATOID CARCINOMA OF PROSTATE WITH ALVEOLAR RHABDOMYOSARCOMATOUS PATTERN.Journal Of Clinical And Diagnostic Research [Serial Online] 2008 August: 2008 August:2: 1009-1012
Available From
Http://Www.Jcdr.Net/Back_Issues.Asp?Issn=0973-709x&Year=2008&Month=August &Volume=2&Issue=4&Page=1009-1012&Id:242

CASE REPORT

Sarcomatoid Carcinoma of Prostate with Alveolar Rhabdomyosarcomatous Pattern

MANNAN R , CHUFAL SS , MISRA V,SINGH PA

ABSTRACT

Sarcomatoid carcinoma (carcinosarcoma) is a rare type of prostatic cancer, composed of an admixture of malignant glandular and spindle cell elements. We report here a case of a 56 year old male who underwent prostatectomy for the symptoms of bladder outlet obstruction.. Histopathology showed small areas of poorly formed glands admixed with large areas of purely alveolar pattern and rossettes. A provisional diagnosis of sarcomatoid carcinoma with alveolar rhabdomyosarcomatous pattern (Gleason's score 5+4=9/10) was made. These two patterns led to a diagnostic dilemma between primary alveolar rhabdomyosarcoma of prostate, sarcomatoid carcinoma with rhabdomyosarcomatous pattern and mixed tumour of prostate. On immunohistochemistry, a positive staining for cytokeratin and negative staining for desmin, myogenin and myoD-1, led to a diagnosis of sarcomatoid carcinoma with rhabdomyosarcomatous pattern.The case was considered worth documentation, as it describes an alveolar rhabdomyosarcomatous pattern in a sarcomatoid carcinoma of the prostate. The correct diagnosis of such a case is important, as it may affect the treatment and prognosis of the patient.

Key Messages

[1] Sarcomatoid carcinoma should be ruled out in spindle cell tumours of the prostate
[2] Sarcomatoid carcinoma may show the pattern of Rhabdomyosarcoma
[3] Differentiation from primary rhabdomyosarcoma by immunohistochemistry is important as the two tumours have different prognoses and treatment.

Key Words: Sarcomatoid Adenocarcinoma, Prostate, Alveolar Rhabdomyosarcoma,

Corresponding Author :
Dr. Misra V ProfessorDepartment of Pathology
M.L.N. Medical College,Allahabad - 211 001 (INDIA)
Ph. (0532) - 2256087,Fax : (0532) - 2256274
E-mail : vatsmi@hotmail.com
vatsala.m@rediffmail.com

Introduction

Sarcomatoid carcinoma (carcinosarcoma) is a rare type of prostatic cancer, with approximately only 100 cases reported in literature. Tumours are most commonly composed of an admixture of both malignant glandular and spindle cell elements. These tumours are always associated with high grade prostatic adenocarcinoma that occurred previous to, or simultaneously with the sarcomatoid component[1],[2]. Sarcomatoid carcinoma is cytokeratin positive and negative for skeletal muscle markers[3].

Most of the primary rhabdomyosarcomas (RMS) of the prostate occur in the paediatric population[4] There are a very few prostatic rhabdomyosarcomas that have been reported in adults, ranging in age from 17 to 68 years old[5],[6] In younger patients, embryonal subtype is the predominant pattern, although; a single case of alveolar type rhabdomyosarcoma has been reported in an autopsy series from Japan[7]. The RMS is

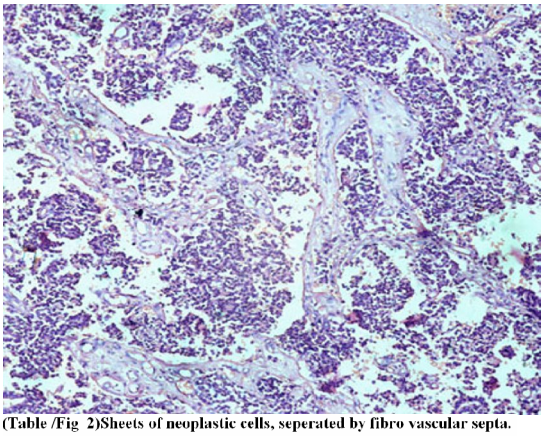
positive for skeletal muscle specific markers such as myogenin and myo-D1, but is negative for cytokeratin.

A case showing an alveolar rhabdomyosarcomatous pattern in sarcomatoid carcinoma of prostate in a 56 year old male is documented here to highlight the importance of differentiating it from primary RMS as well as mixed tumour of prostate (carcinoma and sarcoma).

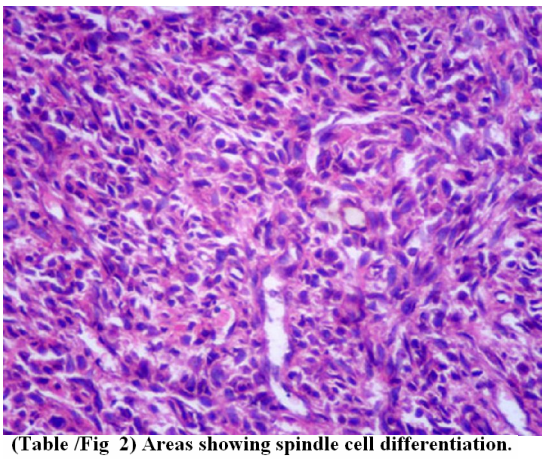
Case Report

A 56 year old male patient presented with re-emergence of symptoms of bladder outlet obstruction (BOO), following a successful trans-uretheral prostatectomy (TURP), two years back. At that time, histological diagnosis of benign prostatic hyperplasia was made at another centre. The patient was symptom free for a period of two years. Initially, re-emergence of symptoms of BOO was attributed to the hyperplasia of the prostate from the residual nodes retained after TURP. However, digital rectal examination and elevated prostatic spesific antigen (PSA) levels of 23.8ng/ml, alerted the surgeon incharge to an aggressive pathology. Ultrasonography showed hypertrophied middle and lateral lobes projecting into the lumen of the urinary bladder. Per-operatively, the plane of surgical separation could not be identified. So, bits and pieces of prostatic tissue were resected, and the specimen was sent for histopathological examination.

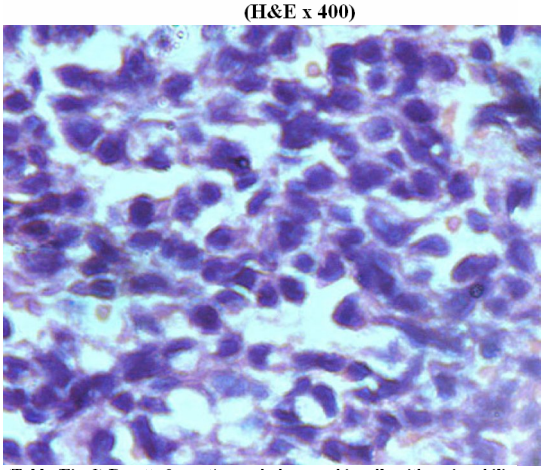
Sections processed showed sheets of neoplastic cells, seperated by fibro - vascular septa. The cells were pleomorphic, hyperchromatic with vesicular nuclei, and prominant nucleoli. A good number of cells with clear cytoplasm (hypernephroid type) were also seen. Two pieces showed spindle cell differentiation, rosette formation and pleomorphic cells with pink cytoplasm that were arranged in an alveolar pattern [Table/Fig 1], [Table/Fig 2], [Table/Fig 3].



(Table /Fig 2)Sheets of neoplastic cells, seperated by fibro vascular septa. (H &E x 100)



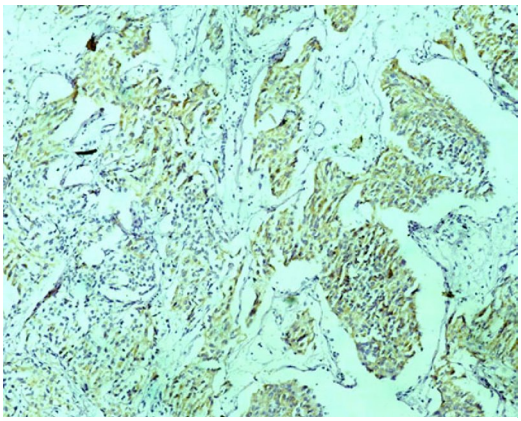
(Table /Fig 2) Areas showing spindle cell differentiation. (H&E x 400)



(Table /Fig 3) Rosette formation and pleomorphic cells with eosinophilic cytoplasm (H&E x400)

Sections processed from the bladder growth showed clear cells arranged in an alveolar pattern. A provisional diagnosis of sarcomatoid adenocarcinoma with rhabdomyosarcomatous pattern (Gleason's score 5+4=9/10) was made.

Immunohistochemistry showed strong cytokeratin positivity [Table/Fig 4]



(Table/Fig 4)Immunohistochemistry showed strong cytokeratin positivity. (x 100)

whereas desmin, myogenin and myo-D1 were negative. This confirmed the provisional diagnosis of sarcomatoid carcinoma with an alveolar rhabdomyosarcomatous pattern, and ruled out the possibility of primary rhabdomyosarcoma and mixed tumour of prostate.

Discussion

A majority of neoplasias of prostate are not difficult to diagnose, however, sometimes there is problem in differentiation of poorly differentiated adenocarcinoma having a sarcomatoid component, from other tumours having spindle cell morphology. They can be mixed tumour, primary mesenchymal tumour or benign stromal proliferation.

Spindle Cell Lesions of The Adult Prostate

Spindle lesions unique to the prostate

1. Stromal Nodules of Hyperplasia
2. STUMPs (Stromal tumors of uncertain malignant potential) and Stromal Sarcomas
3. Sarcomatoid Carcinoma of the Prostate
4. Sclerosing Adenosis

Spindle lesions not unique to the prostate

[1]. Leiomyoma/Leiomyosarcoma
[2]. Rhabdomyosarcoma
[3]. InflammatoryMyofibroblastic Tumor
[4]. Solitary Fibrous Tumor
[5]. Gastrointestinal Stromal Tumor
[6]. Miscellaneous Lesions-

A. Synovial Sarcoma
B. Osteogenic Sarcoma
C. Malignant Fibrous histiocytoma
D. Angiosarcoma
E. Neurofibroma
F. Malignant Peripheral Nerve Sheath tumor

In sarcomatoid carcinomas, the sarcomatoid component can vary from 5 to 99%; in which, cases having a predominant sarcomatoid component may be mistaken for a primary RMS prostate[1] Since sarcomatoid carcinoma and primary rhabdomyosarcoma have different aetiological profiles, histogenesis, prognosis and therapeutic interventions; it is of paramount importance to differentiate them. In contrast to the relatively good prognosis for treated rhabdomyosarcoma in children, the prognosis in adults is poor, with most dying of the disease in < 2 years, despite multimodality therapy[4] Patients with sarcomatoid carcinoma also have poor outcomes, with an actual risk of death of 20% within 1 year of diagnosis, and frequent widespread metastases to sites including bone, liver, and lung.

The sarcomatous component in sarcomatoid carcinoma demonstrates malignant features including hypercellularity, enlarged hyperchromatic nuclei, frequent mitoses and occasional necrosis. Bizarre tumour giant cells may be present within the sarcomatoid component. In approximately 10% of sarcomatoid carcinomas, the sarcomatoid component has the maximum mild atypia. In approximately 30% of cases, heterologous patterns of osteosarcoma, chondrosarcoma, or rhabdomyosarcoma are also present[1] In the present case, a rhabdomyosarcomatous pattern was evidenced by the presence of rossettes and a purely alveolar pattern, both of which are not described in literature. These patterns led to a diagnostic dilemma of whether it was a primary alveolar

rhabdomyosarcoma of prostate, or a sarcomatoid carcinoma with rhabdomyosarcomatous pattern. However, absence of the muscle specific markers led to a diagnosis of sarcomatoid carcinoma. Sarcomatoid carcinomas are usually positive for cytokeratin and epithelial membrane antigen[3] Expression of cytokeratin by the spindle cell component of sarcomatoid carcinoma suggests a common origin rather than a collision tumour composed of sarcoma and carcinoma[9] Thus, the neoplasm in question was diagnosed as sarcomatoid carcinoma having an alveolar rhabdomayosarcomatous pattern.

The diagnostic importance of recognizing them is important, as the treatment plans are different.Treatment for mesenchymal spindle cell lesions and especially rhabdomyosarcoma, is the intricate and organized blending of surgery, chemotherapy and radiotherapy. At present, patients are categorized according to their risk, which takes into account the location of the tumour and the histological and surgical results. For prostatic adenocarcinoma, there is a combination of treatments, such as surgery followed by radiation, or radiation paired with hormone therapy, which works best. So in adenocarcinomas, hormone therapy is used and conventional chemotherapy is reserved for the cases which are resistant to hormonal treatment. LHRH agonists such as bicalutamide and nilutamide are important hormonal agents used in prostatic carcinoma.

The present case is an adenocarcinoma, but two sections mainly showed spindle cell component [Table/Fig 2]. In the rest of the sections, an alveolar pattern with broad septa and small cells attached to these septa with rosette like arrangements [Table/Fig 1] mainly central area and [Table/Fig 3] left upper area) was quite similar to the rhabomyosarcomatous pattern. These may be poorly formed glands, but their appearance was more like that of a rosette, than glands. That's why a diagnosis of sarcomatoid adenocarcinoma with rhabdomyosarcomatous pattern (not differentiation) was made. The diagnosis was later confirmed by immunohistochemistry. The case was considered to highlight the varible presenting pattern of adenocarcinoma prostate that can be easily misdiagnosed (specially by postgraduates and young, less experienced pathologists), and to stress the role of IHC in resolving such diagnostic problems that could affect the treatment and prognosis of the patient.

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