Scholars Journal of Medical Case Reports

Sch J Med Case Rep 2016; 4(8):629-631 ©Scholars Academic and Scientific Publishers (SAS Publishers) (An International Publisher for Academic and Scientific Resources) ISSN 2347-6559 (Online) ISSN 2347-9507 (Print)

DOI: 10.36347/sjmcr.2016.v04i08.020

Sarcomatoid urothelial carcinoma of urinary bladder with heterologous smooth muscle elements-a case report and review of literature

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Abstract: Sarcomatoid urothelial carcinoma is a biphasic malignant neoplasm exhibiting morphologic and or immunohistochemical evidences of both epithelial and mesenchymal differentiation. This uncommon vesical neoplasm is more frequently diagnosed in males with a peak frequency in sixth to eighth decades. Radiotherapy or Cyclophosphamide has been reported as specific risk factors. These tumors are associated with poor prognosis and multimodality therapy is recommended. After surgical resection of the tumor of the urinary bladder, chemotherapy and chemoradiotherapy is recommended. Here we report a case of sarcomatoid urothelial carcinoma with heterologous smooth muscle elements in a 70 year old male.

Keywords: Urothelial carcinoma, sarcomatoid, heterologous, smooth muscle.

INTRODUCTION

Sarcomatoid urothelial carcinoma represents an aggressive form of urothelial carcinoma with a broad spectrum of histological features [1]. It is a very rare neoplasm and it accounts for about 0.3% of urothelial tumors [2]. Sarcomatoid urothelial carcinoma was originally termed as carcinosarcoma because of combination of epithelial and mesenchymal elements [3]. More recently the term sarcomatoid carcinoma has been applied to this neoplasm as the sarcomatoid features appears to be derived from de-differentiation of carcinomatous component [4].

CASE REPORT

70 year old male patient presented with hematuria and painful micturition for last two months. CT scan of abdomen revealed a large well defined lobulated heterogenous enhancing mass lesion in anterior wall and roof of urinary bladder with transmural invasion of wall of bladder measuring 6x4x4 cm.No evidence of adjacent organ invasion or metastasis. Cystoscopic biopsy was reported as poorly differentiated urothelial carcinoma.

Patient underwent radical cystoprostactectomy and extended pelvic lymph node dissection. The post operative course was uneventful.

Grossly it was a large polypoidal growth, fleshy and filling almost the entire lumen of the bladder. It was seen infiltrating the muscular wall without breaching it.



Fig-1: Cystoprostactectomy specimen showing a polypoidal growth filling the lumen of bladder

Microscopic examination revealed a biphasic neoplasm with both epithelial and high grade spindle cell areas. Epithelial areas showed cells arranged in nests and sheets separated by thin fibrovascular septae, and spindle cell areas showed markedly pleomorphic spindle cells admixed with inflammatory infiltrate. Immunohistochemically the neoplastic cells were positive for both Cytokeratin and Vimentin and focal areas showed SMA positivity.

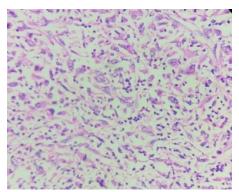


Fig-2: H&E stain shows high grade spindle cells

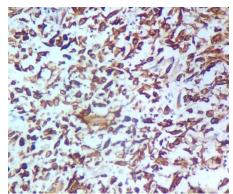


Fig-3: Pan CK positivity in spindle cells

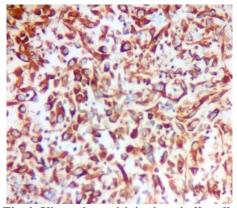


Fig-4: Vimentin positivity in spindle cells

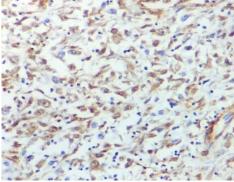


Fig-5: SMA positivity in spindle cells

Final report issued was Sarcomatoid urothelial carcinoma with heterologous smooth muscle areas (SMA Positive)

DISCUSSION

Sarcomatoid carcinoma refers to all biphasic malignant neoplasms exhibiting evidence of epithelial and mesenchymal differentiation with presence or absence of heterologous elements [1, 5].

Originally described by Dent in 1955, sarcomatoid urothelial carcinoma was termed as carcinosarcoma because of combination of epithelial and mesenchymal component [3, 4]. The current WHO employs the term sarcomatoid carcinoma with or without heterologous elements. There is disagreement regarding the histogenesis of this tumor, but molecular genetic studies have reported that both components in this biphasic neoplasm are monoclonal [6].

Macroscopic appearance of the sarcomatoid variant of urothelial carcinoma is large polypoid intraluminal mass.

Microscopically, the tumor is composed of epithelial and mesenchymal components. Epithelial component is represented by urothelial carcinoma, squamous cell carcinoma, adenocarcinoma and small cell carcinoma. Mesenchymal component is represented by high grade spindle cell neoplasms which can be malignant fibrous histiocytoma or leiomyosarcoma. Heterologous differentiation in the form of rhabdomyosarcoma, osteosarcoma and chondrosarcoma can occur, of these osteosarcoma is the commonest [5-7]. As per WHO, smooth muscle elements are also considered as heterologous elements [8].

Immunohistochemically, the carcinomatous component will be positive for epithelial markers like EMA and pan-keratin, and positive for vimentin in approximately 80% to 90% of tumors. The sarcomatous component reacts with vimentin and variably stains for keratin AE1/AE3, keratin CAM 5.2, and EMA. It may express specific markers for specific mesenchymal differentiation if present [5, 6].

This uncommon vesical neoplasm is more frequently diagnosed in males with a peak frequency in sixth to eighth decades. Radiotherapy Cyclophosphamide has been reported as specific risk factors. sarcomatoid carcinoma can arise in any portion of the urothelium. It is most frequently found on the lateral walls of the bladder and is rare in extravesical sites. Most patients with sarcomatoid carcinoma of the urinary bladder had high-histological grade and advanced stage disease at the time of presentation. It shows an unfavorable prognosis worse than high-grade urothelial carcinoma [7].

CONCLUSION

Sarcomatoid urothelial carcinoma of urinary bladder is rare and aggressive neoplasm that has similar clinical presentation as that of conventional high grade urothelial carcinoma, but has worse prognosis. Most of

the cases are at an advanced stage at the time of presentation. Radical surgery along with chemoradiation improves the prognosis of the tumor.

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