

Pleomorphic undifferentiated sarcoma of urinary bladder with calcified pulmonary metastasis: A rare entity

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ABSTRACT

We report the case of a 29-year-old male who presented to us with hematuria, dysuria and bilateral flank pain. On evaluation, the patient was found to have primary pleomorphic undifferentiated sarcoma of bladder with calcified pulmonary metastasis, confirmed with computerized tomography scan and immunohistochemistry.

Key words: Calcified pulmonary metastasis, immunohistochemistry, pleomorphic undifferentiated sarcoma of urinary bladder

INTRODUCTION

Pleomorphic undifferentiated sarcoma (also called as pleomorphic malignant fibrous histiocytoma) is an extremely rare tumor of the urinary tract in adults.^[1] In view of the rarity of its presentation, information about it is still limited. Herein we present a case report and literature review of primary pleomorphic undifferentiated sarcoma of urinary bladder. The intention of this article is to provide an update on the clinical characters, biologic behavior, and pathological and immunohistochemical characteristics of this rare tumor.

CASE REPORT

A 29-year-old male presented with history of hematuria, dysuria, and bilateral flank pain of 2 months duration. He had undergone right ureteroscopy and double J stent (DJ stent) insertion for right ureteric calculus and bladder biopsy for bladder tumor 1 month earlier at another hospital (operative details not available). Bladder biopsy had revealed necrotic

material. He had also undergone an open cysto-lithotomy 20 years earlier, i.e., at the age of nine years. He was of a low socio-economic status from a rural background, hailing from North Karnataka, where primary bladder calculus is still common. The patient did not give history of recurrent urolithiasis. So he was asymptomatic for the last 20 years.

General and systemic examinations were unremarkable. Per-rectal examination revealed a hard, nodular, bimanually palpable mass felt just above the prostate. Computerized tomography showed a heterogeneously enhancing mass with calcification, involving bladder base and lateral walls (70 mm × 52 mm) with bilateral hydro-ureteronephrosis (left > right) with right DJ stent *in situ* [Figure 1] and with calcified pulmonary metastases [Figure 2].

Cystoscopy revealed broad-based solid tumor, with necrotic material on the surface, occupying almost the whole of the urinary bladder except for the anterior wall and bladder dome. Both ureteric orifices were not seen. The lower end of the right DJ stent was almost completely engulfed by the tumor. The stent was seen just peeping out of the bladder mass on the right side and the right

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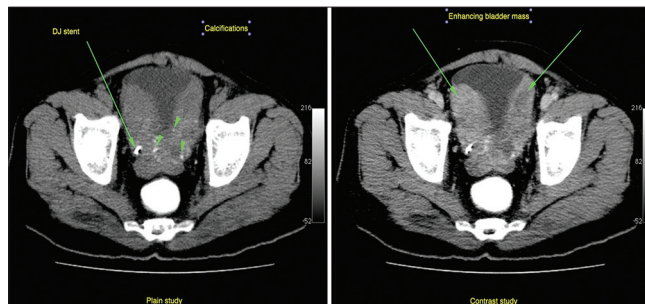


Figure 1: Computerized tomography scan showing the bladder mass with tumor calcification

ureteric orifice could not be visualized inspite of the presence of the DJ stent. Transurethral resection of a part of the tumor was performed. The biopsy was reported as sarcomatoid variant of urothelial carcinoma with heterologous (osteosarcoma) differentiation [Figure 3]. Immunohistochemistry [Table 1] supports the diagnosis

of undifferentiated pleomorphic sarcoma with osteoid formation.

Bone scan showed focal radio tracer concentration in the chest not conforming to any rib. Single-photon emission computed tomography showed uptake in lung parenchyma representing metastatic pulmonary lesions.

The patient was explained about the disease and the need for multimodality treatment. He refused any form of treatment and succumbed to his disease 3 months later.

DISCUSSION

The overwhelming majority of bladder tumors are epithelial in origin with mesenchymal tumors accounting for fewer than 5% of bladder tumors in adults. Essentially any mesenchymal tumor can occur in the adult bladder. These include smooth muscle neoplasms (leiomyoma and leiomyosarcoma), tumors of endothelial differentiation (hemangioma, hemangioendothelioma and angiosarcoma), paraganglioma, osteosarcoma, fibrosarcoma, solitary fibrous tumor,

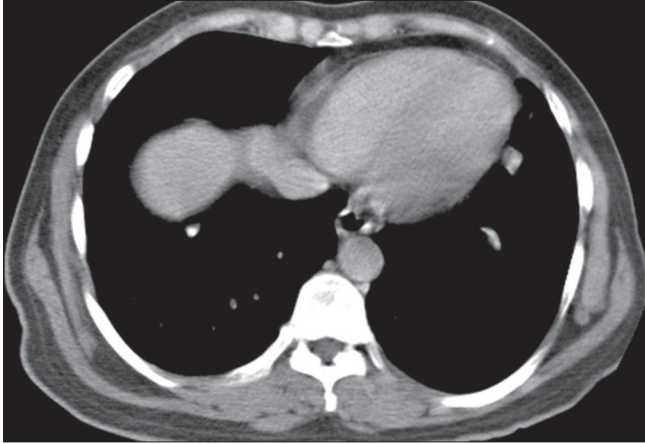


Figure 2: Computerized tomography scan chest showing calcified lung metastasis

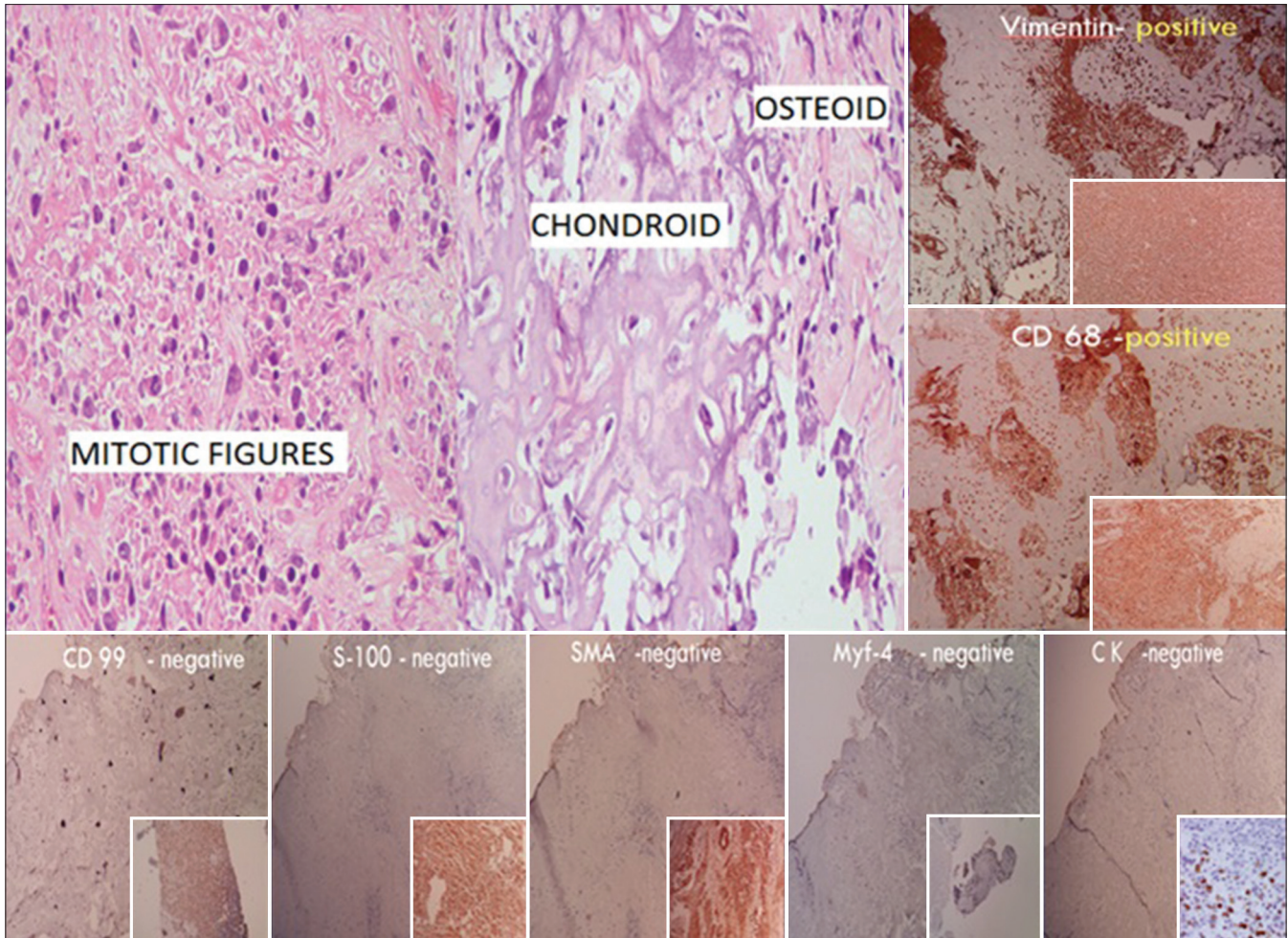


Figure 3: Histopathology showing infiltrating round to oval shaped cells with intervening lace-like osteoid and marked pleomorphism. Areas of abnormal cartilage seen. Large areas of necrosis noted. Abnormal mitosis noted (Immunohistochemistry showed tumor cells positive for vimentin and CD68. Negative for Pan cytokeratin (CK), CK20, CD99, SMA, S-100 and Myf-4)

Table 1: Immunohistochemistry markers

Markers	Carcino sarcoma	Leiomyo sarcoma	Rhabdomyo sarcoma	Neural tumors	Pleomorphic un differentiated sarcoma	Malignant melanoma	Ewing's sarcoma
Vimentin	+	+	+	+	+	+	+
CD68	-	-	-	-	+	-	-
S-100	-	-	-	-	-	+	-
SMA	-	+	+	-	-	-	-
Myf-4	-	-	+	-	-	-	-
CK	+	±	±	-	-	-	-
CD 99	-	-	-	-	-	-	+

SMA = Smooth muscle actin, CK = Cytokeratin, CD = Cluster of differentiation, Myf = Myogenin

alveolar soft part sarcoma, perivascular epithelioid cell neoplasm (PEComa), granular cell tumor, neurofibroma, lipoma, liposarcoma, and undifferentiated pleomorphic sarcoma (malignant fibrous histiocytoma).^[1]

Pleomorphic malignant fibrous histiocytoma (undifferentiated pleomorphic sarcoma) is a rare entity with only 29 cases reported in literature.^[2] It appears mostly in men (4:1) with a mean age of 60 years (20-84), and usually manifests with macroscopic hematuria or irritative urinary symptoms.^[2] There are no clear etiological factors that cause bladder sarcomas, although there is an association with pelvic radiation and systemic chemotherapy for other malignancies. Nimmanon *et al.* have described a case of malignant fibrous histiocytoma of the bladder occurring as a post radiation cancer after the treatment of a cervical carcinoma.^[3]

Immunohistochemistry is necessary for accurate pathological diagnosis of undifferentiated pleomorphic sarcoma. The transurethral resection biopsy done in this case was initially reported as sarcomatoid variant of urothelial carcinoma with heterologous (osteosarcoma) differentiation and immunohistochemistry was advised. Malignant fibrous histiocytoma must be separated from sarcomatoid urothelial carcinoma as well as reactive spindle cell proliferations of the bladder.^[4] The much more commonly encountered sarcomatoid urothelial carcinoma can be associated with a malignant epithelial component, and stains positively for the immunohistochemical markers of epithelial differentiation such as cytokeratin (CK).^[4] Immunohistochemically, MFH is nonreactive for cytokeratin. It is often reactive for vimentin, α -1-antichymotrypsin, and focally reactive for CD68 (Cluster of Differentiation 68).^[4] In our case, the tumor cells were positive for vimentin and CD68 and negative for PanCK (Pan Cytokeratin), CK20 (Cytokeratin 20), CD99, SMA (Smooth Muscle Actin), S-100 and Myf-4 (Myogenin) [Table 1].

Another interesting feature about this case was the presence of calcified lung metastasis. A large variety of neoplasms can produce calcified lung metastases. The sarcomas known to develop calcified lung metastases are

osteogenic sarcoma, chondrosarcoma, synovial sarcoma, and giant cell tumor.^[5] Among carcinomas, the papillary and mucinous adenocarcinomas are the histological types most likely to develop calcified lung metastases.^[5] The metastases of a number of other tumors have calcified after antineoplastic therapy. Calcification in metastases arises through a variety of mechanisms: Bone formation in tumor osteoid, calcification and ossification of tumor cartilage, dystrophic calcification and mucoid calcification.^[5] Since calcified lung metastases can strongly resemble granulomas or hamartomas, a reasonable suspicion of malignancy is necessary when evaluating calcified pulmonary nodules.^[5] In our case, histopathology did support osteosarcoma, but immunohistochemistry supported undifferentiated pleomorphic sarcoma with osteoid formation. Pulmonary metastasis was confirmed by bone scan, which showed radiotracer uptake. To the best of our knowledge this is the first case of bladder cancer to develop calcified lung metastasis. Kurian *et al.* have reported a case of urothelial bladder cancer with cavitary lung metastasis.^[6]

The rarity of the disease makes it difficult to assess the biologic behavior of these tumors. However, from the limited reports, it appears aggressive with high local recurrence rates and metastases.^[7] Radical surgery and systemic chemotherapy have been reported in the literature, but in view of the availability of only a few case reports therapeutic guidelines cannot be established. In a literature review of 20 cases of primary malignant fibrous histiocytoma, Gunia *et al.* observed an overall 1- and 2-year disease specific survival rates of only 47.8% and 31.9% respectively, after radical cystectomy and systemic chemotherapy.^[8] Hence the disease runs an aggressive course regardless of the therapeutic options employed. Our patient chose not to undergo any treatment.

CONCLUSION

Primary undifferentiated pleomorphic sarcoma of the urinary bladder is a rare and aggressive tumor. Immunohistochemistry helps in making an accurate pathological diagnosis.

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