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Sarcomatoid Variant of Urothelial Carcinoma of the Bladder: Five Case Reports and a Review of the literature

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ABSTRACT

Sarcomatoid variant of urothelial carcinoma (SVUC) is an uncommon histological variant of urothelial carcinoma. Sarcomatoid carcinomas are aggressive malignancies with poor prognosis, which by definition are biphasic neoplasms with both an epithelial and a mesenchymal component. There is no consensus opinion on the best treatment modalities for this tumor. Herein on are reported 5 cases of SVUC with a brief review of the literature. The data was collected from the medical records in the Department of Urology, at the University Hospital Center of Rabat Morocco during the period from January 2014 to December 2017.

Introduction

Sarcomatoid variant of urothelial carcinoma is an uncommon histological variant with an epithelial component and a mesenchymal component. It accounts for less than 1% of all urothelial carcinomas [1]. Sarcomatoid carcinoma is the preferred terminology according to the World Health Organization classification for any biphasic tumor of urothelial origin, with or without heterologous elements, and now encompasses what has been previously referred to as carcinosarcoma [2]. In this article, five cases of SVUC are reported with a brief review of the literature on this aggressive malignancy.

Methods

Five cases of sarcomatoid variant of urothelial carcinoma were identified from the records of the department of Urology at the Ibn Sina University Hospital Center of Rabat from January 2014 to December 2017 and included in this retrospective study. All patients were treated and followed at the same institution. Age, gender, clinical presentation, pathological features, and follow-up were extracted from the medical charts.

Results

Five patients were included in this study. They were all male and their median age was 67 years (ranging from 60 to 74 years). All our patients had a smoking history. The initial presentation was gross hematuria in all cases; 3 patients also complained of lower urinary tract symptoms. Transurethral resection of bladder tumor (TURBT) was performed in all cases. Cystoscopic appearance revealed large papillary tumors in all cases. Histologicaly, the epithelial component was composed of high-grade urothelial carcinoma, whereas the sarcomatous component was composed of high-grade spindle cell (Figure 1). The muscle was invaded in all cases pT2. Computed tomography scan revealed metastatic disease to lymph node, lung and bones in three cases; two of them underwent cisplatin based chemotherapy while the third one could not have chemotherapy because of renal failure and died 2 months later. The two patients with localized disease underwent radical cystectomy with pelvic lymphadenectomy, and the final pathology result was pT3 N2 in both cases. These two patients had adjuvant chemotherapy. Two cancers progressed under chemotherapy and the patients died at 8 and 10 months respectively after the diagnosis. The evaluation could not be done for two patients.

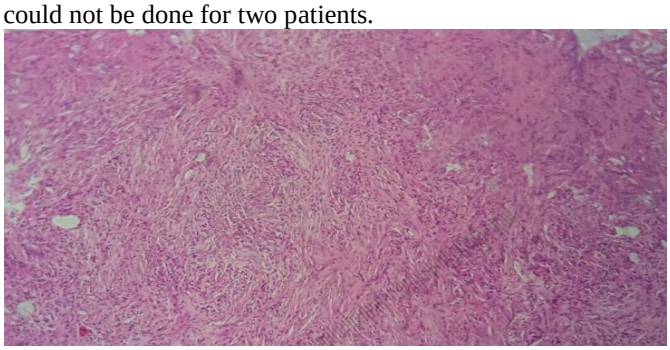

Figure 1 Low power magnification shows sarcomatoid variant of Urothelial carcinoma of the bladder

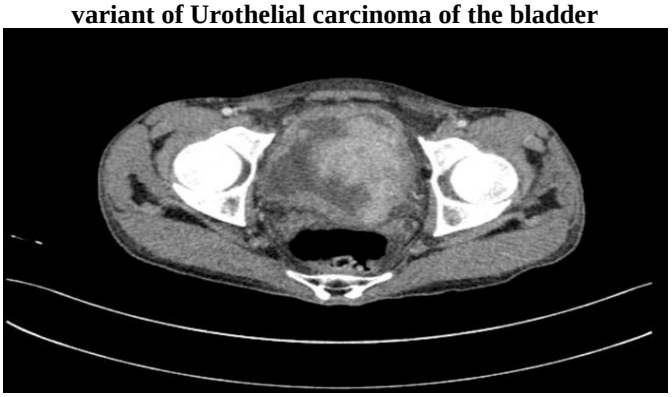

Figure 2. Computed tomography scan showing a large tumor of the urinary bladder

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Discussion

Sarcomatoid carcinoma is a fairly uncommon form of carcinoma whose malignant cells have histological, cytological, or molecular properties of both epithelial tumours and mesenchymal tumours [3].

SVUC of the urinary bladder predominantly affects elderly patients, with male predilection [4]. The mean age at presentation is 66 years, most commonly occurring in the sixth and seventh decades but with a wide age range from 30 to 91 years. There is a male predominance with a male-to-female ratio of 3:1 (ranging from 1.3 to 16:1) [3,5]. Commonly reported risk factors include tobacco smoking, previous urothelial carcinoma, recurrent cystitis, diabetes, neurogenic bladder, and bladder diverticulum [6]. Previous history of radiation or cyclophosphamide therapy has also been reported to be associated with this variant of urothelial carcinoma [7].

Patients with SVUC typically present with microscopic or gross hematuria; however, a variety of other symptoms can be encountered such as dysuria, nocturia, acute urinary retention, and lower abdominal pain [7].

On macroscopic examination, these tumors are typically a large intraluminal polypoid exophytic mass often with a hemorrhagic and/or ulcerated surface and variable amounts of necrosis. Average tumor size is 4.5 cm but ranges widely from 1 to 13 cm. The most common tumor sites within the bladder are the fundus and lateral wall, with the trigone being involved less frequently [5,7].

Microscopically, the epithelial component is most commonly composed of urothelial carcinoma, but adenocarcinoma, squamous cell carcinoma, or even rarely small cell carcinoma can be encountered [8]. Urothelial carcinoma in situ may be the only epithelial component present, or the urothelial carcinoma component may only be found after additional tumor sampling. The mesenchymal component is commonly composed of an undifferentiated high-grade spindle cell neoplasm, but heterologous differentiation includes osteosarcoma, chondrosarcoma, rhabdomyosarcoma, liposarcoma, and angiosarcoma, in decreasing frequency [8]. In a study of 28 cases by Sanfrancesco et al. [9] the sarcomatous component was found to consist predominantly of 4 histological patterns, including spindled not otherwise specified (most common), myxoid, pseudoangiosarcomatous, and malignant fibrous histiocytoma–like undifferentiated. Osteoid was the only heterologous differentiation seen in a small minority of the cases (3 of 28 cases). Interestingly, leiomyosarcoma is typically not seen as a component of sarcomatoid carcinomas [10]. In addition, lymphovascular invasion is frequently encountered, and commonly, there are nodal and visceral metastases at the time of diagnosis [2].

It has been stated that immunohistochemical staining had revealed that epithelial elements react with cytokeratins; whereas stromal elements react with specific markers which correspond with the type of mesenchymal differentiation [11].

SVUC can introduce a wide-ranging differential diagnosis. These would encompass other tumors and pseudotumors with spindle cell morphology including primary sarcomas of the bladder, most commonly leiomyosarcoma, inflammatory myofibroblastic tumor (IMT), urothelial carcinoma with reactive pseudosarcomatous stroma, and postoperative spindle cell tumor [3]. Although extremely rare, sarcomatoid carcinomas are said to be more common than a primary sarcoma of the urinary bladder [3].

Because of the rarity of sarcomatoid carcinomas, there is no standardized treatment, and patients are often treated in a similar fashion to patients with urothelial carcinoma [12]. Many authors suggest cystectomy should be performed even for early-stage disease (T1) to avoid the risk of tumor progression and the potential for incomplete tumor removal in a TURBT [5]. Radical cystectomy with pelvic lymphadenectomy seems to be the treatment option of choice for those who are candidates for surgery. However, despite radical surgery, local recurrence and/or metastasis have been reported [4]. Multimodal therapy, usually consisting of cystectomy followed by adjuvant chemotherapy and/or radiation, is the usual approach, yet the effectiveness of these treatments has not been well studied [13]. Some complete responses have been reported after neoadjuvant chemoradiation therapy [14]. Some other authors have found no survival benefit for patients receiving chemotherapy and/or radiation [9]. Atilgan et al. in his case report proposed that although sarcomatoid carcinoma is very aggressive and lethal tumor, disease-free survival times could be enhanced with radical combination therapies [15].

Compared with conventional urothelial carcinoma, SVUC has a poorer outcome, with 46% and 63% mortality rates within 1 and 5 years of diagnosis, respectively [4]. Black et al. stated that patients with SVUC have worse disease-specific and overall survival, even after adjusting for stage of tumor, in comparison with patients with high-grade pure urothelial cancer [16]. The patients often present with a high histological grade and advanced-stage disease, with tumor stage being a significant predictor of cancer-specific survival [5]. Sanfrancesco et al in their study, observed that the complexity of the assorted sarcomatoid patterns present within the tumor appeared to have a significant association with decreased survival [9]. Furthermore, they found that those tumors with chordoid and myxoid features were also significantly associated with reduced overall survival [9]. Other poor prognostic factors include positive surgical margins at the time of resection and metastatic disease at the time of presentation [8].

Conclusion

Sarcomatoid urothelial carcinoma is a rare type of urothelial carcinoma usually presenting at a higher grade and stage. It is associated with inferior outcome in comparison with conventional urothelial carcinoma. There is a need for a multicentre trial in order to identify a consensus opinion regarding the best treatment option.

Competing Interests

The authors declare that they have no competing interests.

Author Contributions

All authors contributed in the development of this publication and approved the final manuscript.

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