Intradiverticular Sarcomatoid Carcinoma of The Bladder: An Overview Starting From A Peculiar Case.

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INTRODUCTION

Sarcomatoid carcinoma (SC) of the bladder is a rare and highly aggressive tumour, defined according to the World Health Organization (WHO) as a biphasic neoplasm exhibiting morphologic and/or immunohistochemical evidence of epithelial and mesenchymal differentiation.[1] SC of the urinary bladder is an uncommon neoplasm, accounting for 0.1 to 0.3% of all bladder malignancies and is more frequent during the seventh decade.[2-4] As standard protocol of the university hospital in which the case was reported, the patient was informed at the admission and signed an informed consent allowing data collection for research purposes. This case report is in accordance with the Helsinki Declaration, conforms with the Committee on Publication Ethics

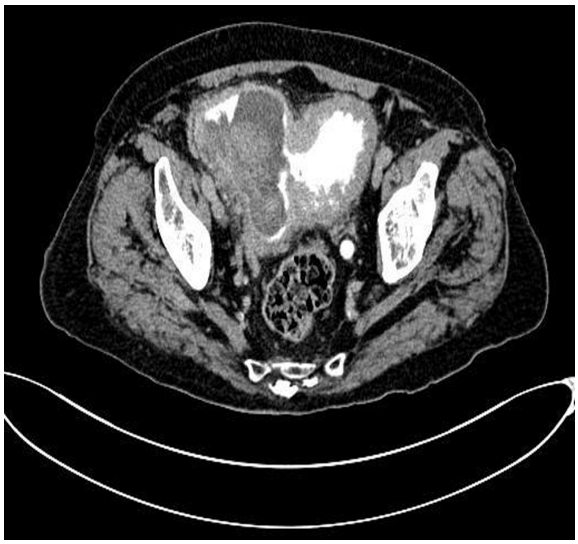

Figure 1. Suspicious lesion inside a large bladder diverticulum.

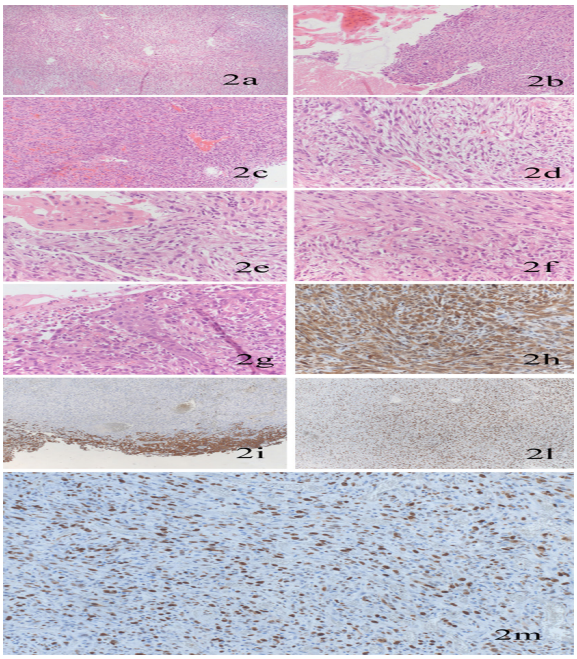

Figure 2. Pathology of the specimen: **a.** Hematoxylin-eosin (H-E), 63X; **b.** (H-E), 100X; **c.** (H-E), 100X; **d.** (H-E),160X; **e.** (H-E), 200X; **f.** (H-E), 200X; **g.** (H-E), 200X; h-i-l-m: Immunoperoxidase for: Smooth Muscle Actin (h), CK20 (i), p53 (l), Mib-1 (m) nuclear counterstaining with Mayer's hemalum.

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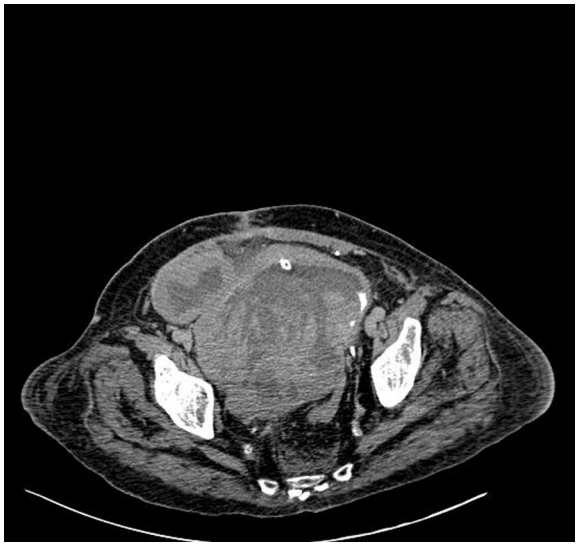

Figure 3. Abdomen CT showing a mass of heterogeneous appearance of 15x15 cm which occupies the pelvic cavity.

(COPE) guidelines (http://publicationethics.org/) and was approved by the Institutional Review Board (IRB) of the university hospital in which it was reported.

CASE REPORT

A 77-year-old man was referred to our hospital for macroscopic hematuria, dysuria, and pelvic pain. Cystoscopy showed a suspicious papillary lesion inside a large bladder diverticulum. Chest and abdomen computed tomography (CT) (**Figure 1**) showed no evidence of distant visceral metastases. Cystoscopy confirmed the presence of a polypoid area within a bladder diverticulum. A transurethral resection of the tumor (TUR-T) was performed and SC was histopathologically suspected. Since the patient refused cystectomy, we have opted for a less invasive surgery: bladder diverticulectomy. At gross examination, the specimen measured 13 x 10.5 x 7 cm. Surgical samples were formalin fixed, paraffin embedded and cut into 4 micron thick sections for the histological examination with haematoxylin-eosin stain and immunohistochemical procedures against Smooth Muscle Actin (SMA), cytokeratins 20, p53 and Ki-67 (DakoCytomation, Glostrup, Denmark). At the microscopic examination, the tumour had a biphasic appearance with an epithelial component represented by areas of squamous carcinoma with corneous pearls and areas of urothelial high grade carcinomas and a mesenchymal component with spindle, pleomorphic cells (**Figure 2a-2g**). Frequent mitoses and necrosis were evidenced. The tumour infiltrated the whole thickness of the diverticulum wall into the perivisceral fat. At immunohistochemistry, the spindle areas were positive for SMA (**Figure 2h**), while the epithelial (squamous and urothelial) ones displayed intense cytokeratin 20 staining (**Figure 2i**). P53 immunopositivity was found throughout the tumour (**Figure 2l**). High proliferation index (50%) was documented by immunohistochemistry against Ki-67 (**Figure 2m**). Based on the histological and immunohistochemical findings, SC of the urinary bladder was diagnosed. No postoperative radiotherapy or chemotherapy was performed. 40 days after, the patient's clinical status worsened with haematuria and abdominal pain and he died. Abdomen CT showed that pelvic cavity was largely occupied by a mass of heterogeneous appearance of 15x15 cm (**Figure 3**), with several satellite nodules in the context of intraperitoneal fat, anterior abdominal wall, left paramedian and left obturator sites, with various dimensions from 3 to 6 cm. The right ureter was encompassed by the sarcomatous mass.

DISCUSSION

SC of the urinary bladder is a rare neoplasm and histogenesis is a controversial issue.[5-8] Macroscopically, these tumors are usually large, polypoid or nodular. Most of the reported cases contain high-grade papillary/undifferentiated urothelial carcinoma. In addition, other subtypes with epithelial origins, such as small-cell carcinoma, squamous carcinoma and adenocarcinoma have been reported. The most common sarcomatous elements are chondrosarcoma, leiomyosarcoma, and malignant fibrous histiocytoma.[4] According to recent data, the most common location of SC is the lateral wall of the bladder.[2] In our case the sarcomatous element was a leiomyosarcoma and was located in a bladder diverticulum. Our patient presented with hematuria and pelvic pain. The only curative management of this kind of neoplasm could be early detection and aggressive surgery. The other preferred modalities of treatment include cystectomy or transurethral resection of the bladder (TURB) with or without radiation therapy and chemotherapy. Furthermore, chemotherapy and radiotherapy do not provide apparent survival advantages. Transurethral resection and partial cystectomy carry the risk of incomplete tumour resection. Neoadjuvant radiochemotherapy with radical cystectomy can provide 20 months of recurrence-free survival.[9] In a recent large retrospective study which analyzed 221 cases, the overall 5-year cancer-specific survival rate after radical cystectomy was 20.3%. The 1-, 5-, and 10-year survival rates for SC of the urinary bladder were 53.9%, 28.4%, and 25.8%, respectively.[2] Treatment of bladder SC should be aggressive and multimodal but optional treatment is not still encoded. As already reported,[10] SC has a poor prognosis despite of all treatment modalities and a median cancer-specific survival of approximately 14 months. In particular, cancer-specific survival was significantly better for those who underwent cystectomy instead of transurethral resection. On one hand, chemotherapy and radiotherapy do not provide apparent survival advantages;[11] on the other hand, neoadjuvant/adjuvant radiochemotherapy has been used in many cases, and there were complete responses after neoadjuvant treatment.[12] Considering our experience and the most updated literature, we solicit future multicentric large cohort analyses which may clarify the best evidence-based treatment of the reported condition.

CONFLICT OF INTEREST

None declared.

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