

Primary Signet-Ring Carcinoma in the Bladder Presenting as a Hypervascular Luminal Polypoid Mass¹

과혈관성 용종양 종괴로 발현한 방광의 원발성 반지세포암종: 1예 보고¹

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Primary signet-ring carcinoma is a very aggressive and rare variant of a primary urinary bladder cancer, accounting for less than 1% of cases. We reported on a 76-year-old patient with primary signet-ring carcinoma who occurred metastatic lymphadenopathy with extranodal invasion causing intraluminal tumor thrombi in the adjacent vein, and pulmonary metastasis over the course of three months. We demonstrated the computed tomography findings of primary signet-ring carcinoma of the bladder and correlated the imaging findings with the pathologic features. We reviewed the distinguishing imaging findings of the primary signet-ring carcinoma compared with urothelial cell carcinoma, the most common subtype of the bladder cancer.

Index terms

Signet-Ring Carcinoma
Bladder Cancer
Adenocarcinoma
Thromboembolism
Metastasis

INTRODUCTION

The common types of epithelial bladder tumors include urothelial carcinoma, squamous cell carcinoma, and adenocarcinoma in decreasing order of frequency (1). Among them, adenocarcinoma accounts for 0.5-2%, and is mostly the secondary type (1, 2). Primary adenocarcinoma mainly arises in the gastrointestinal tract, breast or gall bladder, and rarely in the urinary bladder (1, 3). In particular, signet-ring cell subtype in adenocarcinoma of the bladder is relatively rare and one of the most aggressive variant (4). We reported characteristic computed tomography findings of signet-ring cell carcinoma distinguishable from that of urothelial cell carcinoma in the bladder.

CASE REPORT

A 76-year-old woman with painless gross hematuria was

referred to our hospital. On pelvic ultrasonography, a 1 cm hyperechoic luminal polypoid mass was seen in the left-sided wall of the bladder. The mass showed greater arterial enhancement than the perilesional spared mucosal layer on enhanced abdominal and pelvic computed tomography (CT) (Fig. 1A). The lesion revealed a venous wash-out enhancing pattern (Fig. 1B). Additionally, there were no perivesical fatty infiltration or enlarged lymph nodes and no metastases in other organs. Pathologically, signet-ring carcinoma was confirmed by the transurethral resection of the bladder tumor. Perilesional proper muscle and lymphatics have been invaded by the malignant cells.

After three months, the mass grew remarkably with an irregular shape presenting as wall thickening of the bladder on dynamic CT (Fig. 2A). A venous phase image demonstrated inner necrosis-like low density portion of the wall with perivesical fat infiltration (Fig. 2B). The lesion extended to the trigone in-

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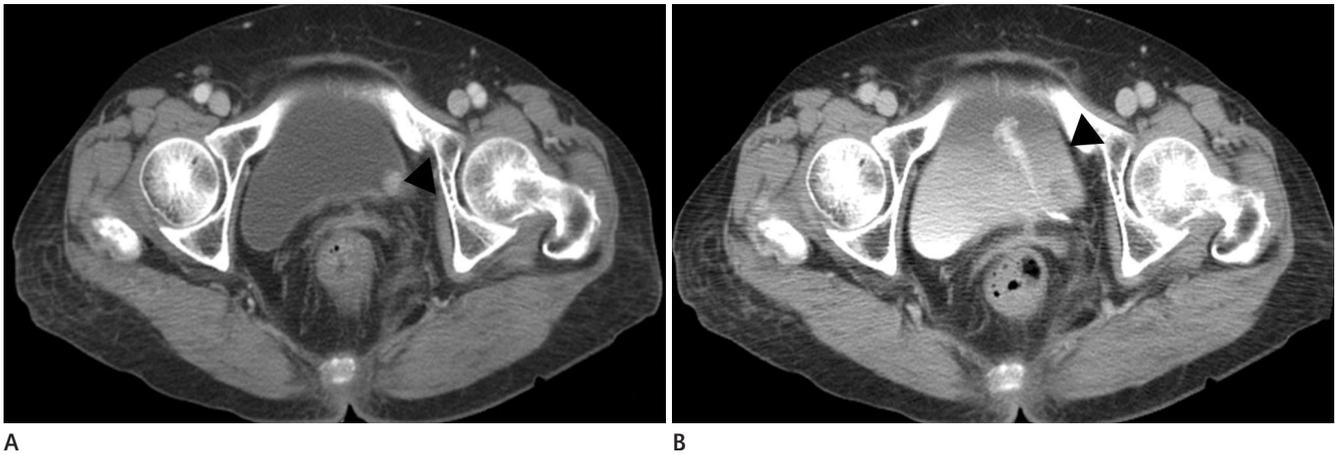


Fig. 1. Primary signet-ring carcinoma of the urinary bladder in a 76-year-old woman.
A. Initial dynamic contrast-enhanced axial CT shows an arterial enhancing intraluminal nodular mass in the left-sided posterolateral wall of the bladder.
B. Luminal mass in the bladder shows a venous wash-out pattern. There was no detectable perivesical fat stranding or enlarged pelvic lymph nodes.

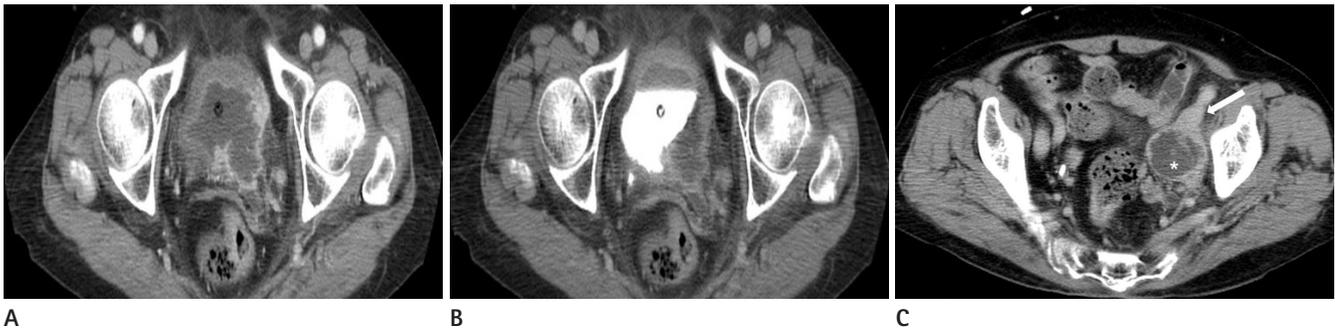


Fig. 2. Three-month follow-up dynamic contrast-enhanced CT.
A. An enhancing enlarged and extended mass with central low density correlates with necrosis on arterial phase image.
B. Mass shows a wash-out enhancing pattern with perivesical fat stranding on venous phase image.
C. Newly developed a 3.6 cm low density mass (asterisk) with peripheral rim-like enhancement and inner whirling enhancing pattern invades the left external iliac vein on venous phase image. Juxta-distal portion of the left external iliac vein to the mass involving site shows luminal dilatation, indicating venous stasis (arrow).

cluding left ureterovesical junction, resulting in hydronephrosis (Fig. 2B). In addition, a newly developed 3.6 cm poorly enhanced low density mass with inner venous whirling enhancing portion was discovered in left-sided pelvic cavity. It destructed adjacent left external iliac venous wall and formed luminal tumor thrombi, causing venous stasis (Fig. 2C). A new pulmonary nodule was noted in scanned right lower lobe. Three weeks later, the pulmonary nodule was double in size with hot-uptake on positron emission tomography CT images indicating metastasis. There were no evidence of other primary malignancy.

Radical cystectomy with an ileal conduit was performed for relieving the mass effect. A pathologic report identified the central low density area of the bladder tumor on CT images

as characteristic mucin pool of adenocarcinoma (Fig. 3). The low density mass in left sided pelvic cavity was the metastatic lymph node with extranodal invasion to adjacent left external iliac vein. After 7 days, a CT venography manifested thrombi in the left external iliac vein and other deep vein thrombosis of left lower extremity. The patient expired, owing to a pulmonary thromboembolism.

DISCUSSION

Histologically, six patterns are known as subgroups of adenocarcinoma: no specific type, enteric, mucinous, signet-ring cell, clear cell, and mixed (5). The primary signet-ring carcinoma, a rare type of the adenocarcinoma, secretes unevenly

distributed mucin within the intracellular space and nuclei (1, 2, 6). This tumor shows perivesical fat strandings or distant metastases more commonly than urothelial carcinoma at diagnosis (1). Therefore, diffuse bladder wall thickening and perivesical fat stranding are commonly seen on CT images, suggesting a transmural extension of the cancer (1, 7). The heterogeneously enhancing bladder wall thickening findings of the tumor show similar features to linitis plastica in other abdominal visceral adenocarcinoma (2). Further, the inner mucinous component of the tumor appears as a bizarre-shaped, central necrosis-like low density area, and a distinct margin on enhanced CT images (7). The primary adenocarcinoma have been reported with enlarged lymph nodes in 25% of cases, with peritoneal deposit in 38% of cases (2), and distant metastases or involving local organs in 28% of cases (8).

On the other hand, urothelial carcinoma generally presents as an intraluminal papillary, nodular mass or focal wall thickening at diagnosis. More than half of the lesions measured less than 2.5 cm (1). The perivesical spread in 10% of cases and cancer invasion of the adjacent organ or abdominal wall in 3% of cases are noted in the patients with urothelial carcinoma (9).

In our case, primary adenocarcinoma of the bladder belongs to the nonurachal type based on the origin and signet-ring carcinoma according to the histologic subgroup. Upon initial diagnosis, the lesion revealed a hypervascular polypoid mass similar to urothelial carcinoma. After three months, the mass demonstrated typical further aggressive features with remarkable growth, developing irregular wall thickening, and perivesical fat strandings considered as adenocarcinoma. Moreover the cancer metastasized to the left external iliac lymph node, hence causing venous invasion and a thromboembolism. No evidence of extravesical primary malignancy was found.

A diffuse wall thickening appearance on CT image can be seen in an inflammatory, infective, or fibrotic condition and mimic presence of the bladder adenocarcinoma (2). In addition, the signet-ring cell carcinoma can be demonstrated as an arterial enhancing luminal polypoid mass resembling urothelial cell carcinoma as in this case. The lesional biopsy can be performed for the differential diagnosis, but the initial pathologic report of the signet-ring cell type malignancy can be inconclusive (2). Therefore radiologists need to consider bladder adeno-

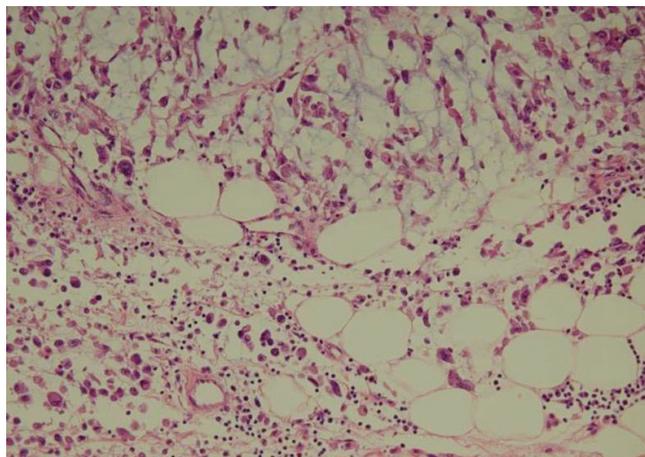


Fig. 3. Photomicrograph of the bladder mass. Center of the mass shows a redundant mucoid background with lower cellular density of the signet-ring cells (Hematoxylin-eosin stain, $\times 400$).

carcinoma with these CT findings and try to determine the lesional extension or metastatic lesions (2).

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과혈관성 용종양 종괴로 발현한 방광의 원발성 반지세포암종: 1예 보고¹

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원발성 반지세포암종은 방광암의 한 종류로서 공격적인 성향을 보이며, 발병 빈도가 약 1%로 매우 드물다. 이번 증례는 방광의 원발성 반지세포암종을 진단받은 환자로, 초기에 기존에 알려진 반지세포암종보다는 요로상피암종과 유사한 소견을 보였으며, 추적 검사에서 림프절 전이 및 림프절 외 파급으로 정맥벽을 침윤하여 종양성 혈전을 생성한 경우이다. 이에 저자들은 증례를 통하여 원발성 반지세포암종의 병리학적 소견과 연관되는 CT 소견을 정리하고, 방광암 중 가장 흔한 요로상피암종의 영상 소견과 비교하여 발생 기전 및 임상적 관점에 대한 이해를 넓히고자 한다.

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