RESEARCH ARTICLE

PRIMARY SIGNET RING CELL CARCINOMA OF THE URINARY BLADDER – A CASE REPORT

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INTRODUCTION

Bladder cancer is the 9th most common cancer worldwide, with 357,000 cases recorded in 2002 [1]. Bladder cancer is the 13th most common cause of death, accounting for 145,000 deaths worldwide [1-2]. Histologically, 90% of bladder cancers are of urothelial origin, 5% are squamous cell carcinomas, and less than 2% are adenocarcinoma or other variants [3]. Adenocarcinoma of the bladder comprises only 0.5% to 2% of all epithelial bladder tumours and primary signet-ring cell carcinoma, a variant of adenocarcinoma, is extremely rare with a prevalence estimated at 0.24% of primary bladder cancers; no effective systemic chemotherapy has been established for this subtype [4,5]. Signet ring cell carcinoma can be of urachal origin and directly extend into the bladder. We report a case of primary signet ring cell carcinoma of bladder.

Case Report

An 65-year-old man presented with lower urinary tract obstructive/irritative urinary symptoms. The patient was evaluated at another center with cystoscopy and biopsy of a lesion from bladder neck region. The histopathological examination revealed an adenocarcinoma of the bladder. Urine cytology was negative for malignant cells. Ultrasound imaging revealed a mixed echoic polypoidal lesion on the anterior wall of the bladder. CT (Computed tomography) examination revealed an heterogeneously enhancing mass lesion (4.4×3.7×3.5 cm²) on the antero-superior wall of the bladder (Fig. 1a & b) (? Urachal in origin). A detailed imaging of the gastrointestinal tract was done to rule out primaries. Transurethral resection of the bladder tumor was done and rejected material sent for histopathological examination. Histopathological examination revealed signet ring cell carcinoma infiltrating into the lamina propria. The PAS (Periodic acid Schiff) stains highlighted the signet ring cells. Immunohistochemistry studies revealed cells that expressed Ck 20 strongly and Ck 7 focally (Fig 2 a&b). In view of the histopathological report and absence of lymph node/distant metastases on CT imaging, radical cystectomy with ileal conduit was done.

ABSTRACT

Primary signet ring cell carcinomas of the bladder are extremely rare tumors. These tumors generally present as high-grade, high-stage tumors and have a uniformly poor prognosis. We report a case of primary signet ring cell carcinoma of the bladder in a 65 year old male.

DISCUSSION

Pure primary signet-ring cell carcinomas of the bladder are very rare and only a few case reports exist of a mixed urothelial/signet-ring cell variant [6-8]. Saphir first described primary signet-ring cell carcinoma of the bladder in 1955, and less than 200 cases have been reported in the literature since then [7, 8]. Saphir [7] summarized that Signet-ring cell tumors were usually primary carcinomas of the gastro-intestinal tract, but could also occur as primary carcinomas of the urinary bladder.

Primary Adeno/signet-ring cell carcinomas identified in the bladder are rare and need to be differentiated from metastases from other primary sites, usually from the gastrointestinal tract. Clinical, imaging and immunohistochemical findings help in identifying the primary source of signet cell cancer. The immunohistochemical tests used to distinguish primary vesical adenocarcinoma from metastatic colonic adenocarcinoma includes cytokeratin (CK) 7, CK20, 34bE12, thrombomodulin, villin, CDX-2, and b-catenin [9]. CK7 and CK20 are reported to be positive in more than half of primary bladder adenocarcinomas [9]. The typical colonic adenocarcinoma staining profile, CK7 negative and CK20 positive, has been reported in 29% of primary vesical adenocarcinomas [9].

Fig. 1a, b CT (Computed tomography coronal and axial view) showing an heterogeneously enhancing mass lesion (4.4×3.7×3.5 cm²) on the antero-superior wall of the bladder
A combination of CK7 and CK20 by itself does not appear to differentiate primary bladder from colonic adenocarcinomas. Treatment of signet-ring variants of bladder cancer has not been well-defined due to the rarity of the tumor. Cystectomy remains the treatment of choice for localized disease. There is no systemic standard chemotherapy regimen for advanced disease, which in general is considered chemotherapy resistant [9].

CONCLUSIONS

Signet-ring cell cancers deriving from the bladder are rare entities and usually present with advanced incurable disease and associated poor outlook. As there is considerable overlap histologically and immunohistochemically between colonic and bladder adenocarcinoma, extensive clinical and radiologic workup is required for diagnostic accuracy.

References


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