Case Report

Neuroendocrine Carcinoma of Urinary Bladder: Misdiagnosed Neoplasm.

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Citation

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Abstract: Neuroendocrine carcinoma of the urinary bladder is an exceedingly rare pathology, comprising 0.35-0.70% of all bladder cancers. The presenting symptoms and gross morphology are identical to conventional urothelial carcinoma, but the prognosis is much worse with frequent metastasis. Neuroendocrine carcinoma has been subdivided into small cell and large cell carcinoma. Hematuria is the main symptom with some patients having associated paraneoplastic conditions. The cell of origin of neuroendocrine carcinoma is from a multipotential stem cell with the ability to differentiate to other types. We report a case of 76 yr old male who presented with history of hematuria was diagnosed on bladder biopsy as neuroendocrine carcinoma bladder (100%) with diffuse synaptophysin positivity, Ki 67 index of 85% and negative p63. The treatment of neuroendocrine carcinoma of the bladder is not standardised and the prognosis is poor; however, cisplatin-based chemotherapy regimens seem to have a better prognosis.

Key Words: Neuroendocrine carcinoma, urinary bladder

Introduction

Background

Neuroendocrine carcinoma of the urinary bladder is an exceedingly rare pathology, comprising 0.35-0.70% of all bladder cancers.[1] The presenting symptoms and gross morphology are identical to conventional urothelial carcinoma, but the prognosis is worse with frequent metastasis.[2] The main symptom of this malignancy is hematuria and some patients may have associated paraneoplastic conditions.[3]

Case Details:
A 76 years old male was presented with history of hematuria. On Ultrasound abdomen and pelvis a well-defined lobulated heteroechoic lesion arising from wall lateral to left vesico-uteric junction was noted with moderate left hydroureteronephrosis.
Discussion:
Neuroendocrine tumor arising from the genitourinary tract is uncommon. The bladder and prostate are the most common sites in genitourinary tract. The first patient of Small cell carcinoma bladder was described in 1981. They tend to have an aggressive behaviour than transitional cell carcinoma (TCC).[1]
It is more common in age group of sixth or seventh decade and in males with a history of smoking. The nonspecific risk factors are bladder calculi, bladder manipulation, and chronic cystitis.[2]
The most common presentation is painless gross hematuria (67%–100% in bladder SCC), with or without dysuria. The associated paraneoplastic syndromes can be hypercalcemia, Cushing syndrome and sensory neuropathy.[2]
Several hypotheses are proposed to explain the origin of Neuroendocrine tumor in the bladder. The most important ones are:
– Malignant transformation of bladder neuroendocrine cells
– Multipotent stem cells;
– Metaplastic changes of urothelium.[4]
Neuroendocrine carcinoma is further subdivided into small cell and large cell carcinoma, with large cell carcinoma being rare among them.[5]
Small cell carcinoma can present as a mixed tumor with one or more of the following: transitional cell carcinoma (TCC), adenocarcinoma and squamous cell carcinoma.4 Histologically, pure SCC tends to have a poorer outcome than mixed SCC of the urinary bladder.[2]
The small-cell carcinoma will express markers of both epithelial and neuroendocrine differentiation, the markers of neuroendocrine differentiation which are most commonly positive are neuron-specific enolase (NSE), chromogranin A, synaptophysin, CD 57, CD56, Epithelial markers which are present are cytokeratin 7 (CK7), epithelial membrane antigen (EMA), CAM5.2 and CK8/18. Cytokeratin CAM5.2 can be
positive in both urothelial carcinoma and small-cell carcinoma, but the pattern of staining helps to differentiate, that is, its punctate along the membrane in the first case, and perinuclear in the latter.[5]

The mean survival for all stages is 19.6 months, and the 5-year survival rate is only 8.1%. Bladder SCC has the same metastatic potential of TCC, with the most common sites being regional and distant lymph nodes, liver, and bone. Lung happens to be a site of late metastasis.[1]

Other differential diagnosis to be considered are high-grade urothelial carcinoma, lymphoma, lymphoepithelial-like carcinoma from the lung, metastases from another neuroendocrine tumor (lung), neuroendocrine carcinoma of the prostate infiltrating the bladder and rhabdomyosarcoma (in children).[5]

The 2015 guidelines of National Comprehensive Cancer Network’s recommend resection and chemotherapy with or without radiotherapy (similar to small cell lung carcinoma) for non-locally advanced tumor; radiotherapy and chemotherapy for loco regional advanced disease, and chemotherapy alone for metastatic disease.[5]

**Summary:**

Neuroendocrine carcinoma of bladder is a rare type of cancer accounting for 0.35%-0.7%. Patient usually presents with late stage disease. Our case was pure Neuroendocrine carcinoma without urothelial component, which carries a worse prognosis.

**References:**