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AN INTRIGUING ANOMALY: SMALL CELL NEUROENDOCRINE CARCINOMA IN THE URINARY BLADDER

Pathology

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ABSTRACT

Small cell type neuroendocrine carcinoma of the bladder is a rare and aggressive tumor associated with poor prognosis. It often presents at an advanced stage and comprises less than 1% of bladder malignancies. We report a case of a 72-year-old male diagnosed with cystoscopic biopsy who later underwent a radical cystoprostatectomy, subsequently followed by adjuvant chemotherapy.

KEYWORDS

Bladder carcinoma, Small cell neuroendocrine carcinoma

INTRODUCTION

Fewer than 1% of nonurothelial cell carcinomas of the bladder in humans are small-cell neuroendocrine carcinomas (SmCC). [1] Cramer et al. originally described it in the 1990s, however, up until now, the only available data are cohort studies, case reports, or case series. [2] Because there is a dearth of literature on this entity, it poses a diagnostic hurdle to urologists and radiologists. SmCC occurs more commonly in the sixth and seventh decades of life and is more prevalent in males. It carries a worse prognosis than transitional cell carcinoma and is associated with more aggressive behaviors, with the median survival with and without therapy being 12–24 months and 4-5 months, respectively. [3]

CASE REPORT

A 72-year-old, chronic smoker, presented to the Surgical Outpatient department of this institute with complaints of hematuria for the last 6 months, which was associated with loss of weight and appetite. He was admitted and worked up. The USG findings revealed a lobulated heterogeneous polypoidal soft tissue mass lesion measuring 38x24 mm along the left postero-basal wall of the urinary bladder with mild vascularity on USG doppler, just encasing the left vesico-uretric junction within it. Similar findings were noted on Cystoscopy and a cystoscopic guided biopsy was taken. The histopathological examination of which revealed a tiny fragment lined by urothelial cells, with subepithelium containing a cluster of tumor cells. (Fig 1A,1B) These cells were small, having scant cytoplasm, hyperchromatic nuclei, and inconspicuous nucleoli, showing nuclear molding and prominent mitosis. On Immunohistochemistry, these cells were positive for NSE, and showed dot-like positivity for CK, while they were negative for LCA and Desmin. Overall features were strongly indicative of Small cell Neuroendocrine Carcinoma. Meanwhile, the patient underwent a CECT Abdomen and Pelvis, which showed an ill-defined focal heterogeneously enhancing polypoidal mass lesion measuring 1.8x2.9x1.6 cm with frond-like projections arising from the trigone. (Fig.1C,1D) No evidence of hydronephrosis or obvious lymphadenopathy was seen, however mild fat stranding was observed. The patient underwent a Radical Cystectomy. Grossly, the specimen of urinary bladder along with attached prostrate and B/L seminal vesicles measured 10x10x5 cm. A growth measuring 4x3.5x2 cm was identified involving the left lateral wall of the bladder. (Fig. 1E) Microscopic sections examined from the growth showed features of small cell neuroendocrine carcinoma, pT2bN0M0, with a morphology similar to that of cystoscopic biopsy and R_0 resection. (Fig.1F,1G,1H) The adjoining bladder mucosa showed Tumor-infiltrating Lymphocytes. No lymphovascular/perineural invasion was noted. The patient is currently asymptomatic and undergoing adjuvant cisplatin-based chemotherapy, with a close follow-up.

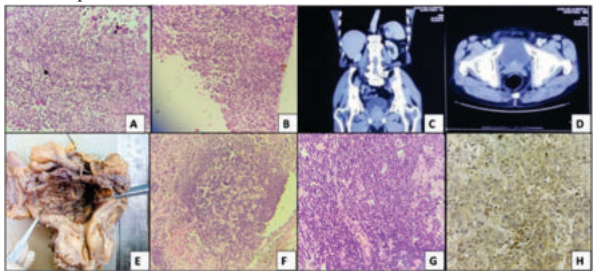

Figure- 1A, 1B: Photomicrograph of cystoscopic biopsy showing tumor cells; H & E stain; 400X magnification; 1C, 1D: CECT Images showing the lesion; 1E: Surgically resected suture-marked specimen; 1F: Photomicrograph showing the presence of tumor; H &E Stain, 200X magnification; 1G: Photomicrograph of tumor cells showing characteristic salt and pepper nuclear chromatin, HE Stain, 400X; 1H: Tumor cells showing positivity for NSE Immunohistochemistry, 400X magnification, DAB Chromogen

DISCUSSION

Neuroendocrine carcinomas are rare and aggressive malignant tumors, mainly occurring in the respiratory and digestive tract. This histological entity includes three subtypes, namely, neuroendocrine small-cell carcinomas, neuroendocrine large-cell carcinomas, and differentiated neuroendocrine tumors such as carcinoid tumors. Urinary neuroendocrine tumors are very rare and small-cell neuroendocrine tumors of the bladder represent only 0.5 to 1% of all bladder tumors. [4] Although most of the risk factors are unclear, smokers, people with chronic cystitis, and people who have bladder stones are more likely to develop these tumors. [5]Due to lacking typical clinical symptoms, early diagnosis is difficult to make. Gross hematuria, pelvic discomfort, and irritable voiding symptoms are the most common non-specific clinical signs, as was seen in our case. The diagnosis depends on histopathological recognition and reactivity for neuroendocrine markers such as Synaptophysin, chromogranin A, and neuron-specific enolase (NSE). [6] Immunohistochemistry aids in distinguishing them from various other differential diagnosis like high-grade urothelial carcinoma, lymphoma, carcinoid, and lymphoepithelial-like carcinoma from the lung. Usually, small cell carcinoma of the urinary bladder is admixed with urothelial carcinoma in 68% of the cases, [7] but in our present case there was no evidence of urothelial carcinoma and it was purely small cell neuroendocrine

carcinoma. Previous reports are suggestive of the notion that this tumor presents at a locally advanced stage, with up to 57% of cases showing lymph node involvement and distant metastasis in 28-50% of cases. [8] In our case, the radiological evaluation did not find evidence of metastatic disease at the time of diagnosis. The preferred local treatment for localized tumors is surgery, however, the risk of metastasis remains, worsening the survival rates and disease-free progression. As this is a rare subtype and only a few reported cases are available, no standard treatment regimen has been established. In localized disease, neo-adjuvant platinum-based chemotherapy with cystectomy has been shown to provide the best result in retrospective studies. [9] In conclusion, it is important to differentiate SmCC from other malignant neoplasms in the bladder in view of its aggressiveness and presentation at an advanced stage with frequent metastasis. Thus, owing to the rarity and poor prognosis of these tumors, their management requires a multidisciplinary approach, in order to improve the survival of these patients.

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