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Case Report Open Access

Long Surviving Patient with Metastatic Neuroendocrine Bladder Cancer: About a Case Report

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Abstract

Introduction: Primitive Neuroendocrine bladder tumors are very rare and with a poor prognosis.

Case report: We report the case of a white 57 year-old man diagnosed with a 6-centimetre primitive neuroendocrine urinary bladder carcinoma associated with a right-retroperitoneal metastasis. He underwent surgery: radical cystoprostatectomy and large lymph node dissection. The histopathology showed small neuroendocrine cells in the bladder and a prostate carcinoma (a low-risk cancer). At seven months post presentation, metastasis appeared at the bottom of the left kidney and in the retroperitoneum, the previous metastasis' place. The patient was treated with a Cisplatin and Etoposide chemotherapy regimen, every three weeks, for six cycles. He is watched for twenty-eight months.

Conclusion: Therefore, their management has to be collaborative.

Keywords: Bladder; Neuroendocrine; Rare tumor; Small cells

Introduction

Neuroendocrine tumors are a very rare entity. They represent about one percent of all tumors. Neuroendocrine tumors are, most of the time, located in the digestive tract (75%) and lungs (12%) but can be found everywhere [1]. Tumors of the urinary tract are more often bladder cancer, rarely prostate or urethra. Neuroendocrine urinary bladder tumors represent about 0, 5-1% of all bladder tumors [2,3]. In 50% of cases they are associated with carcinoma cells [4]. They also are associated with a more aggressive behavior and poorer prognosis than transitional cell bladder carcinoma.

Case Report

We report the case of a 57 year-old Caucasian man diagnosed with a 6-centimetre primitive urinary bladder neuroendocrine carcinoma associated with a right-retroperitoneal metastasis (Figure 1). The tumor was revealed by haematuria, but no dysuria. The patient didn't suffer from pain and had no abdominal mass. He had no medical past and had undergone no treatment. He smoked for 30 years but did not drink alcohol. Transurethral resection bladder tumor specimen showed pure neuroendocrine carcinoma, involving detrusor muscle. After discussing the case in genito-urinary round it was decided surgery for the patient. He underwent a radical cyst prostatectomy and large lymph node dissection without any post-operative complication. The histopathology showed pure small neuroendocrine cells in the bladder and a low-risk prostate adenocarcinoma with a Gleason score 6 (Figure 2). No adjuvant chemotherapy was given. The prostate cancer was treated by surgery. The patient underwent monitoring. At seven months post-presentation, metastasis appeared on Computerized Tomography (CT) scan, at the bottom of the left kidney and in the retroperitoneum, the previous metastasis' place. The patient had no symptom. Any new biopsies were done. The patient was treated with a Cisplatin and Etoposide chemotherapy regimen, every three weeks, for six cycles. The chemotherapy was well tolerated with minor side-effects, no more than grade 2. After six cycles of chemotherapy the patient was in complete remission as the CT scan showed. He is watched for twenty-eight months. He is still in remission.

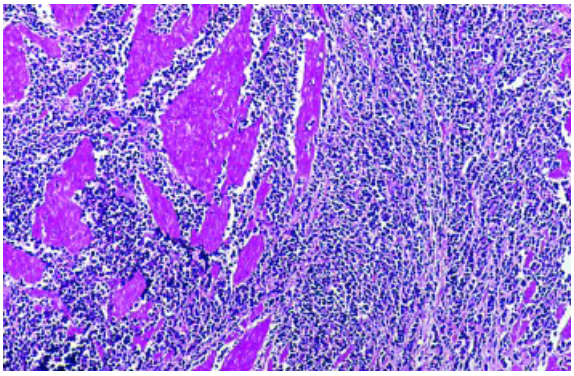

Figure 1: Small cell carcinoma.

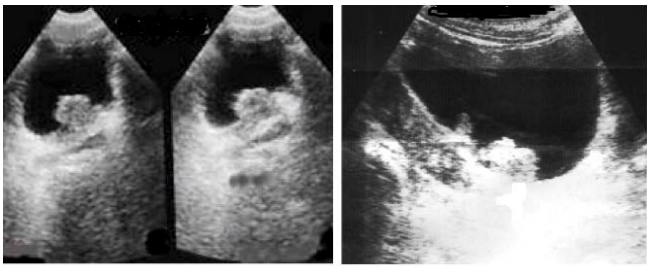

Figure 2: Ultra sound bladder cancer.

Discussion

Neuroendocrine urinary bladder tumors were first described by

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Cramer et al. in 1981[5]. It was the case of a 69-year-old man who showed symptoms of urinary bladder irritability and hematuria. The tumor shared histologic features with oat cell carcinoma of the lung including the presence of small numbers of neurosecretory type granules.These tumors are extremely rare, with an incidence of 0.35-0.70% of all bladder tumors. They have a more aggressive behaviour than transitional cell bladder carcinoma. Small cell carcinoma of the bladder comprises only 0.5% to 1.0% of primary bladder malignancies. Pure small cell carcinoma of the bladder is infrequent and is usually mixed with another histologic subtype (most commonly urothelial carcinoma). The bladder is the most common site for genitourinary extrapulmonary small cell carcinoma. Neuroendocrine tumors affect more men than women (sex ratio 2,5/1), in the seventh decade and are revealed by haematuria, pollakiuria, pain or paraneoplastic syndrome (Lambert-Eaton syndrome, Cushing syndrome, hypercalcemia), but less than in lung small cell carcinoma [6]. The histopathological characteristics are small cells with scant cytoplasm, ill-defined cell borders, finely granular nuclear chromatin and reduced or absent nucleoli. Cells are round or oval and the mitotic count is high. The cells express the Neurone Specific Enolase (NSE) marker, Chromogranine A and synaptophysin. The patho-physiology of these tumors is however still unclear. Since the tumors have a high spread potential, the authors recommend a CT scan and a bone scintigraphy. The presence of metastasis is a factor of poor prognosis [7]. Median survival is only nine months and 19% are alive after five years [4]. Cheng related the case of over nine years survival in a metastatic patient [4]. According two series, pure small cell histology was shown to have poorer outcome than mixed small cell histology [8,9]. Choong NWW et al. [10] reported, in their experience, that the overall survival rate at 5 years does not exceed 8%. At the time of presentation, 59% of patients have clinical stage> T2 and 56% show metastatic disease. In 50% of the patients, fatal progression occurs within 6 months. Local recurrence after radical surgery occurred in 50-70% of cases. Prognosis is dependent on performance status and extent of disease at diagnosis, whereas overexpression of p53, patient age, gender, and presenting symptoms do not appear to correlate with prognosis. Because of the rarity of these tumors, there is no standard treatment of the disease. It's an aggressive tumor. Multimodality treatments are recommended. Treatment is dependent on tumor stage at presentation and patient performance status.

In contrast with small cell lung cancer, more than half of the patients undergo surgery [10] due to the association of transitional cell carcinoma with small cell carcinoma. Surgery (cystectomy) and radiotherapy should be associated with chemotherapy [7]. Only radiotherapy cannot be effective. It could be palliative. Choong and coauthors recommend no adjuvant chemotherapy for stage I and II disease after reporting a 75% cure rate with radical cystectomy alone for stage II disease. Lohrisch and colleagues retrospectively reviewed 14 cases of bladder small cell carcinoma treated with chemoradiation (cisplatin was the chemoregimen used) for stage III or less disease. A 70% 2-year survival was observed and 44% 5-year overall survival [11].

Chemotherapy based on Cisplatin should be considered as the treatment of choice for patients with good performance status (0-1) and good renal function (glomerular filtration rate>60 ml/min). The treatment should be based on neuroendocrine regimens like Etoposide plus Cisplatin or the sequential protocol : Ifosfamide plus Doxorubicin at day 1 and Etoposide plus Cisplatin at day 21.

In unfit patients, Cisplatin should be substituted by carboplatin AUC 5 to 6. The results vary, depending on studies. A retrospective study of forty-six patients treated at the MD Anderson Cancer Centre has reported a five-year survival of 78% for patients receiving neoadjuvant chemotherapy followed by cystectomy, versus 36% for patients undergoing cystectomy alone [12].

Conclusion

Even if research has progressed, neuroendocrine bladder tumors still have bad prognosis. In the absence of prospective studies, the best treatment can't be established for certain. There are however recommendations. Combined-treatment seems to improve recurrence free and overall survival. Surgery with neoadjuvant or adjuvant chemotherapy could be the treatment of choice. Additional radiotherapy should be considered. The histology is imperative for diagnosis.

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