Two Cases of Neuroendocrine Tumor of the Testis

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Abstract

Testicular cancer is uncommon with an incidence of about 5.6 per 100,000 men per year among the U.S. population. Neuroendocrine tumor of the testis accounts for less than 1% of all testicular tumors. In this paper we present two cases of primary neuroendocrine tumor of the testis. The first one is a 17 year old male patient who presented with testicular pain and was found to have well differentiated histopathologic features (carcinoid tumor). The second case was a 40-year-old male patient who presented with painless scrotal swelling and was found to have moderately differentiated histopathologic features (atypical carcinoid).

Keywords: Carcinoid; Testicular carcinoid; Testicular neuroendocrine tumor; Testis

Introduction

Neuroendocrine tumors also called carcinoid tumors arise from enterochromaffin cells, which are found throughout the body. They commonly arise from the intestinal and respiratory epithelium (65% and 25%, respectively). For that, one should exclude the possibility of metastatic tumor to testis before labeling the case as a primary testicular carcinoid tumor. [1] In 1930, Cope described the first case of metastatic carcinoid tumor of the small bowel to the testis, [2] and Simon et al. Reported the first case of primary testicular carcinoid, since then, more than 60 carcinoid tumors have been reported. [3] The usual presentation of testicular carcinoid tumor is a painless mass or to be accidentally discovered during ultrasound for another reason. [4]

Cases Reports

Case 1

A 17-year-old male patient, not known to have previous illnesses, presented with right testicular pain associated with gradual swelling. He denied any weight loss, trauma, hematuria or systemic symptoms. Upon admission his Hemoglobin was 14.5 g/dl; white blood cell count $8.4 \times 10^3/\mu\text{L}$, and creatinine 0.9 mg/dl, Electrolytes and liver function test values were within normal. He had a body mass index of 18 kg/m². He underwent radical

orchiectomy, which microscopically revealed a well differentiated neuroendocrine tumor confined to the testis and epididymis with no lymph vascular invasion. No other teratomatous elements or germ cell components were identified. Immunohistochemistry showed the tumor cells to be positive for chromogranin and CK(MNF) while negative for inhibin, calretinin and melan-A. Ki-67 proliferative index is 3-4% (Figure 1). An extra testicular carcinoid tumor was ruled out and the tumor was staged as pT1NxMx. After two years of follow up with cross sectional imaging and tumor markers including chromogranin A and 5-Hydroxyindoleacetic Acid, no recurrence has been documented and the patient is doing well.

Case 2

A 40-year-old male patient, not known to have previous illnesses, who presented with a progressive, painless left testicular swelling, he denied any weight loss, trauma, hematuria or systemic symptoms. On physical examination a hard mass was palpable on the left testis. Upon admission his hemoglobin was 13.6 g/dl, white blood cell count $7.4 \times 10^3/\mu\text{L}$, and Creatinine 1.0, electrolytes and liver function test values were within normal range. He had a body mass index of 26 kg/m². He had ultrasound imaging which showed a mass of about 5cm associated with hydrocele (figure 2,3).

The patient underwent radical orchiectomy, which revealed

Figure 1: The histopathologic findings of case 1.

grossly a mass measuring 5X4cm and a hydrocele composed of hemorrhagic fluid. Microscopically, the mass was confined to the testis and epididymis with no lymph vascular invasion. The presence of 5MF/10HPF and a Ki-67 proliferative index of 15% indicate that this is an intermediate grade neuroendocrine neoplasm (atypical carcinoid). The tumor cells are positive for CD56 and synaptophysin. They are negative for SALL-4 and CD30 (see figure 4). The tumor was staged as pT1NxMx.

The patient is planned to be followed up on active surveillance with cross-sectional imaging and tumor markers including chromogranin A and 5-Hydroxyindoleacetic Acid which.

Discussion

Testicular cancer is uncommon with an incidence of about 5.6 per 100,000 men per year among the U.S. population [5]. Neuroendocrine tumor of the testis represents 1% of all testicular tumors [6]. The testicular carcinoid tumor can be primary, associated with teratoma or metastatic from another origin. Primary carcinoid tumor of the testis carries an excellent prognosis, [7,8]. The overall incidence of metastasis is about 11% [9] and they are thought to be as a differentiation of the pluripotential germ cell to argentaffinlike cells or the development of a simplified teratoma without other teratomatous elements. [10,11]. Although it should be the first concern to rule out that it is metastatic disease from another site, as the primary tumor cannot be distinguished from metastatic one using only the histopathologic findings. [12]. It also has been noted that the larger the tumor the more malignant it seems to be [12].

Figure 2: The Ultrasound showing the mass for case 2.

Figure 3: Ultrasound showing the mass and hydrocele for case 2.

Figure 4: The histopathologic findings of case 2.

According to the published cases, the right testis seems to have a higher chance of developing primary carcinoid of the testis [8,9]. Primary testicular carcinoid is difficult to diagnose pre-operatively as the currently available imaging modalities unable to differentiate it from other testicular tumors such as germ cell tumors and it is a rare tumor that routine laboratory markers are not cost effective to be done [13]. The most common presentation of testicular carcinoid tumors is painless testicular swelling, nevertheless association with hydrocele and pain has been noted in some cases [6]. It is also known that metastatic testicular carcinoid tumor could present with carcinoid syndrome with symptoms of hot, red flushing of the face; severe and debilitating diarrhea; and asthma attacks. These symptoms are usually the result of substances secreted by the tumor [14].

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