Case Report

Primary testicular carcinoid

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ABSTRACT

Carcinoid tumors of testis are very rare and account for <1% of all testicular neoplasms. Clinically, they can be diagnosed only when there is a metastatic spread or presence of carcinoid syndrome. Mostly, they are diagnosed on histopathology. We report a case of primary carcinoid tumor of testis in a 34-year-old male without associated carcinoid syndrome. Patient underwent radical orchidectomy and is doing well on follow-up. Testicular carcinoid tumors, though rare, should be considered in differential diagnosis when evaluating a testicular tumor. They carry a good prognosis; however, long-term follow-up is necessitated due to potential for delayed metastasis.

Key words: Carcinoid, orchidectomy, testis

INTRODUCTION

Gastrointestinal tract is the most common site for carcinoid tumors, especially the appendix or ileocecal region (85%), while other sites are the lung, liver, and genitourinary tract (15%).[1] Carcinoid tumors of testis are very rare and account for <1% of all testicular neoplasms.[2] Carcinoid tumors of the testis can arise as a primary or a metastatic tumor, with primary tumor being the most common or as a component of a teratoma.[3] We report a case of primary carcinoid tumor of testis along with a review of literature on this entity.

CASE REPORT

A 34-year-old male patient was presented with a painless right scrotal swelling for duration of 6 months. He did not have any complaints of diarrhea, flushing, difficulty in breathing, or abdominal pain (symptoms related to carcinoid syndrome). On examination, his left testis was normal and right testis was enlarged, firm and nontender. Ultrasonography revealed a heterogeneous hypoechoic lesion with no calcification. The beta human chorionic gonadotropin and alpha-fetoprotein levels were within normal limits. With a clinical diagnosis of testicular tumor, the patient underwent radical orchidectomy.

Grossly, the orchidectomy specimen showed a well-defined yellow tan tumor measuring 4.0 cm × 3.2 cm with few hemorrhagic areas.

On microscopic examination, the tumor revealed relatively uniform population of cells arranged in nests, trabeculae, insular, and glandular pattern in a background of fibrous stroma. These cells had abundant granular eosinophilic cytoplasm, round to oval nuclei with a "salt and pepper chromatin" [Figure 1]. No elements of teratoma or other germ cell components were identified. On immunohistochemistry, the neuroendocrine markers chromogranin and synaptophysin along with cytokeratin were strongly positive [Figure 2a-c] supporting the diagnosis of carcinoid tumor. Other immunomarkers such as CD117, PLAP, vimentin, and inhibin were negative [Figure 2c-e]. Computed tomography (CT) chest, abdomen and pelvis were normal postoperatively. Patient is on follow-up since 3 months and is doing well.

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DISCUSSION

Carcinoid tumors of testis are a rare entity and a preoperative diagnosis is almost never suspected. Clinically, they can be diagnosed only when there is a metastatic spread or presence of carcinoid syndrome. Mostly, they are diagnosed on histopathology.[4] It becomes necessary to exclude the presence of a primary tumor in another organ before confirming the diagnosis of a primary testicular carcinoid tumor since there are no morphological differences between primary and metastatic carcinoid tumors. Carcinoid tumor of testis commonly presents with a painless scrotal swelling. Age of presentation ranges from 10 to 83 years. It may be associated with other symptoms such as painful testis, hydrocele, undescended testis, and symptoms of carcinoid syndrome. Being quite rare, only 7 cases of associated carcinoid syndrome (diarrhea, flushing, and bronchospasm) have been reported in literature.[5] Testicular carcinoid tumors are composed of neuroendocrine cells. According to a recent report, they are actually a phenotypic expression of testicular teratomas, thus of germ cell in origin. Various biologically active products are secreted by them, most common being 5-hydroxyindolacetic acid (5-HIAA), which is a metabolite of serotonin. Earlier considered to be benign, the metastatic potential of carcinoid tumors is now widely recognized. Metastases to liver, lung, abdominal wall, vertebrae, skin, and heart have been reported. Carcinoid syndrome may become apparent as a result of the systemic release of 5-HIAA, when the tumor is metastasized outside of the portal venous system.[6]

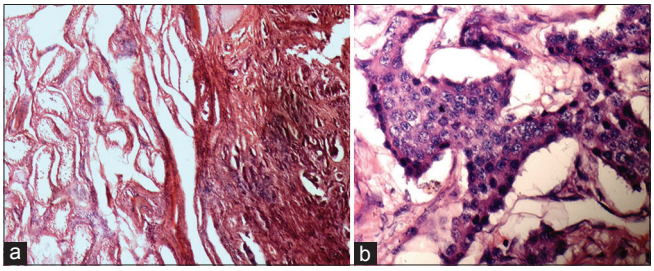

Figure 1: (a) Photomicrograph showing carcinoid tumor and surrounding normal testicular tissue (H and E, ×40). (b) nests and trabeculae of tumor cells displaying characteristic salt and pepper chromatin (H and E, x400)

Testicular carcinoid tumors are divided into three sub-groups: Pure primary testicular carcinoid, primary testicular carcinoid associated with a teratoma (mixed carcinoid), and carcinoid metastases to the testis. More than 75% of the reported cases were pure primary testicular carcinoid tumor. A pure primary carcinoid and a carcinoid metastasis cannot be distinguished by immunohistochemistry techniques; therefore a distant primary must be excluded. Since gastrointestinal carcinoids are usually > 2 cm before they metastasize, CT is used to identify the primary gastrointestinal and endobronchial tumors. Octreotide scintigraphy is approximately 80% sensitive and a useful modality for tumor localization and when combined with single-photon emission CT, it becomes as a most reliable staging procedure. The selection of patients with carcinoids that are likely to respond favorably to somatostatin analog treatment is also facilitated by the use of octreotide scintigraphy.[7]

The treatment of choice for carcinoid testicular tumor is radical orchidectomy.[5]

The reported cases till date suggest an excellent prognosis following orchidectomy in primary testicular tumors. Since these tumors are very rare, data regarding prognostic indicators are lacking. However, on reviewing literature, it is suggested that size of tumor, invasion, and evidence

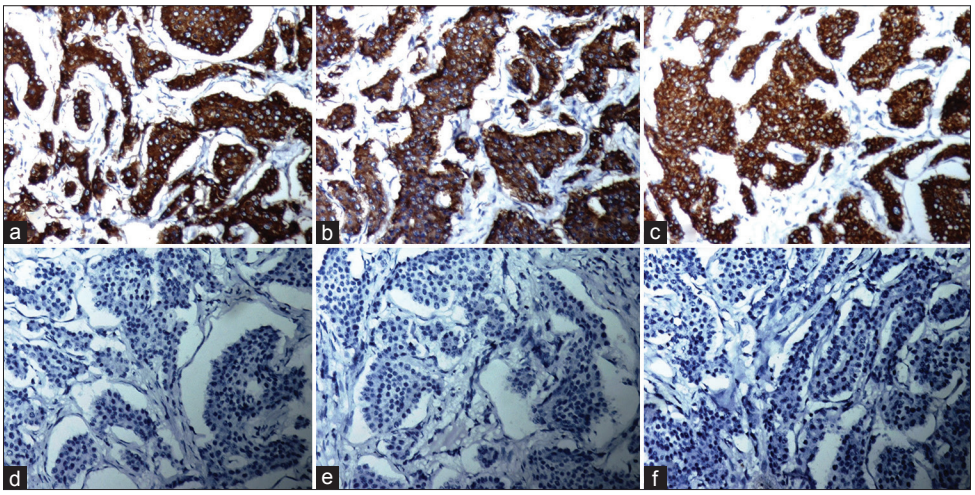

Figure 2: Immunohistochemical expression of tumor cells ([a] synaptophysin +, [b] chromogranin +, [c] cytokeratin +, [d-f] CD117, inhibin, and PLAP: Negative)

of the carcinoid syndrome are associated with metastasis and a worse prognosis.[8] Long-term biochemical and radiological follow-up is essential due to potential for delayed metastases.[4] It has been suggested to check urine 5-HIAA, review the history, and perform a physical examination every 3 months for 1-year and then yearly thereafter.[5]

CONCLUSION

Testicular carcinoid tumors, though rare, should be considered in differential diagnosis when evaluating a testicular tumor. They carry a good prognosis; however, long term follow-up is necessitated due to potential for delayed metastasis.

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Conflicts of interest
There are no conflicts of interest.

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