Case Report

Primary Testicular Carcinoid Tumor presenting as Carcinoid Heart Disease

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ABSTRACT

Primary carcinoid tumors of the testis are very rare, and they seldom present with carcinoid syndrome. We report a hereto unreported instance, where a patient with a long-standing testicular mass presented with carcinoid heart disease, an uncommon form of carcinoid syndrome. He presented with symptoms of right heart failure, episodic facial flushing and was found to have severe right-sided valvular heart disease. His urinary 5-hydroxy indole acetic acid level was elevated. He underwent orchidectomy and the histopathology confirmed a testicular carcinoid tumor.

Key words: 5 Hydroxyl indole Acetic Acid, carcinoid heart disease, testicular carcinoid

INTRODUCTION

Carcinoid tumors are rare (0.23%), occurring mostly in the gastrointestinal tract (85%) while 10% occur in other sites.[1] Primary carcinoid tumors of the testis are very rare, and they account for <1% of all testicular tumors. Although reported in age groups ranging from 10 to 80, they present usually as a painless testicular mass in the fifth to seventh decades of age.[1] This case report is unique in that a testicular carcinoid presented with carcinoid heart disease.

CASE REPORT

A 49-year old male presented with history of pedal edema and breathlessness of 2 months duration. There was history of facial flushing, but no history of wheezing, diarrhea or abnormal blood pressure changes. There was no weight loss or abdominal symptoms. He did not have significant comorbid illnesses except for a long-standing right testicular swelling of 10 years duration. On examination, he had an early ejection systolic murmur in the pulmonary area and diastolic murmur in the tricuspid area. He had hepatomegaly secondary to the severe right heart failure. He had an enlarged, firm and nontender right testis while the left testis was normal. Lymph nodes including the supraclavicular lymph nodes were not palpable. His chest X-ray showed cardiomegaly [Figure 1a] with clear lung fields. Electrocardiogram showed right bundle branch block. Echocardiography revealed the presence of severe right heart disease in the form of dilated right heart chambers, dysplastic tricuspid, and pulmonary valves, with stenosis and regurgitation in both valves. Hence, we evaluated him for carcinoid syndrome. His 24-h urine 5-hydroxy indoleacetic acid (5-HIAA) was elevated, whereas other serum markers for testicular tumors were normal [Table 1]. Computed tomography of the abdomen showed hepatomegaly and infrarenal para-aortic lymphadenopathy, but there were no intestinal tumors. The right testis was enlarged (4.8 × 5.0 × 7.6 cm), vascular, showing heterogeneous density, necrosis, and calcifications [Figure 1b] along with minimal hydrocele. Combined together, these findings favored the diagnosis of a testicular carcinoid tumor. Under local cord block, sedationand continuous cardiac monitoring, he underwent right radical high inguinal orchidectomy [Figure 2]. The right testis measured 3 × 4.5 × 8 cms, with the cut section showing a well-circumscribed tumor with pale white to pale yellow surface and with areas of hemorrhage and necrosis. Histological analysis showed a well-circumscribed encapsulated tumor composed of cells arranged in an acini, insular, and trabecular pattern [Figure 2]. The cells were round to oval with round nuclei, granular chromatin,

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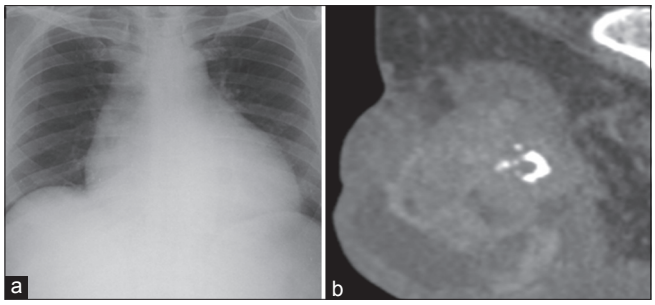

Figure 1: (a) Chest X-ray showing cardiomegaly and (b) CT showing heterogenous mass in the testis with calcification.

Table 1: Biochemical parameters of the patient

Test	Value	Normal range
24 hour urine 5-HIAA	29 mg	2-6 mg/24 h
Serum beta-hCG	0.1 mIU/ml	Up to 5.3 mIU/ml
Serum AFP	4 ng/ml	0-7 ng/ml
Serum LDH	191 IU/L	240-480 IU/L

5-HIAA = 5 hydroxyl indole acetic acid, hCG = Human chorionic gonadotropin, AFP = Alpha-feto protein, LDH = Lactate dehydrogenase

and eosinophilic granular cytoplasm with collagenous stroma. Immunohistochemistry staining for synaptophysin expression was positive. All these features confirmed a carcinoid tumor. We made a diagnosis of a primary testicular carcinoid with carcinoid heart syndrome. He was advised repeat biochemical evaluation during follow-up visits, but he lost to follow up.

DISCUSSION

Primary testicular carcinoid is exceedingly rare accounting for only 0.3-1% of all testicular tumors. Left testicular involvement has been reported commonly.[1] Carcinoid tumors can originate either primarily from the testis or from a mature teratoma or metastasize from an external source (in 10% cases).[2] Their membrane-bound neurosecretory granules secrete bioactive substances such as 5-hydroxy indoleacetic acid, bradykinin, chromogranin-A, substance-P, motilin, etc., These substances are responsible for the episodic flushing, diarrhea, bronchoconstriction, and right-sided valvular heart disease seen in carcinoid syndrome. Carcinoid syndrome occurs in only in about 10-20% of all carcinoid tumors of which <50% develop cardiac carcinoid syndrome.[2] This is due to the inactivation of these amines in the liver when released into the portal veins. For carcinoid syndrome to manifest, they should escape hepatic inactivation either by directly entering the systemic circulation as in bronchial carcinoids or by metastasizing to the liver as in small intestinal carcinoids. Carcinoid syndrome occurs in 1.3-3.1% of testicular carcinoid, and it does not require hepatic metastasis.[3] When released from the testis or ovary, these amines may escape the portal-hepatic system and drain directly into

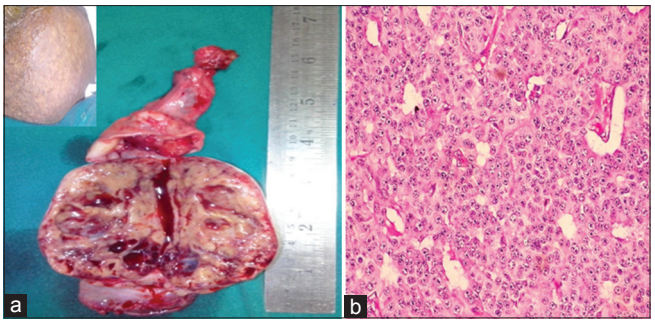

Figure 2: Macroscopy: coronal section of the testis with spermatic cord. Inset: Enlarged testis. (b) Microscopy: monomorphic cells in the nested trabecular patter

systemic circulation. Carcinoid heart disease occurs as a late complication of carcinoid syndrome because of the scarring and fibrosis of the tricuspid and pulmonary valves caused by the circulating amines, which reach the right heart through the systemic circulation.[4] The aortic and mitral valves usually escape due to the clearance of 5HT by the bronchial monoamine oxidase.

Surgical management with high inguinal orchidectomy has been curative for tumors confined to the testis. Somatostatin analogs provide symptomatic relief and improve prognosis. The long-term prognosis depends on the size of tumor, coexisting teratoma, and metastasis. Long-term follow-up with urinary 5-HIAA, serum chromogranin A, and CT or octreotide scintigraphy is mandatory.

To the best of our knowledge, this is the first report of a testicular carcinoid presenting with carcinoid heart disease. Moderately elevated urinary 5-HIAA levels and right testicular involvement were the other atypical features in our patient. It was not possible to ascertain the cause for the paraaortic lymphadenopathy, as the patient was not willing for further evaluation. The likelihood of tumor spread from the testes is high in our patient. In the background of a long-standing history and lack of evidence for intestinal tumor in the computed tomography, we made a diagnosis of a primary testicular carcinoid with carcinoid heart syndrome. In the largest case series of 29 cases with testicular carcinoid reported so far, the majority had presented as a palpable mass (21/29) while only two had carcinoid syndrome without any cardiac manifestations.[5] Low-grade carcinoid tumors are slow growing and usually present as painless testicular masses of variable duration. There are case reports of carcinoid tumor being diagnosed 10 years after the onset of testicular swelling.[6] In another instance, ultra sonogram revealed a nonpalpable testicular tumor that turned out to be a carcinoid.[7]

A careful physical examination of the structures of the scrotum and determination of its transillumination characteristics are important while evaluating chronic painless scrotal masses. Color Doppler ultrasonography is

helpful for the initial screening of all solid scrotal masses and in cases where there is a suspicion of noncommunicating hydrocele obscuring a testicular mass.

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