

## PRIMARY CARCINOID TUMOR OF TESTIS

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Carcinoid tumors are rather uncommon and about 85% of them occur in the appendix or ileocecal region. When they occur outside the GI tract, they are usually metastatic. A total of 43 cases of pure primary testicular carcinoid tumor have been reported.<sup>1</sup> We report a case of carcinoid tumor of the testis, the diagnosis of which was confused by the FNA diagnosis of Leydig cell tumor, another uncommon type of testicular neoplasm. Verification of the diagnosis was done by immunohistochemistry and electron microscopy. To our knowledge, this is the first case of primary carcinoid tumor of the testis reported from the Middle East.

### Case Report

A 26-year-old Saudi male student was referred to our hospital with the diagnosis of Leydig cell tumor of the left testis. The patient had presented to an outside hospital with a three-month history of a painless swelling of the left testis. Ultrasound examination revealed the presence of a localized area of suspicious hypoechoogenicity and FNA of the lesion was interpreted as a Leydig cell tumor. On examination, we found his left testis to be moderately enlarged in size and hard in consistency without tenderness. The other testis was normal and clinically there were no signs of abdominal metastases and no gynecomastia was found. A left radical orchiectomy was carried out through a groin incision. Tumor markers sent preoperatively (beta human chorionic gonadotrophin, alphafetoprotein, dehydroepiandrosteindione, androsteindione) all came back as normal. Histopathology of the testis was reported as carcinoid tumor of the testis and the margins were free of tumor. There was no history of symptoms of carcinoid syndrome. We performed quantitative estimation of 5-HIAA, gastrointestinal contrast study, chest x-ray and CT of the abdomen. These studies

were all normal and there was no evidence of other primary or metastatic carcinoid tumor.

Grossly, the tumor measured 4.5 cm in maximum dimension. It was well demarcated from the surrounding uninvolved testis, solid and yellow-tan in color (Figure 1). The spermatic cord and epididymis were normal.

Histologically, the tumor was well demarcated from the surrounding testicular parenchyma. The tumor cells were arranged in nests, tubules and trabeculae separated by delicate fibrovascular stroma (Figure 2). The cells were round or polygonal and had eosinophilic granular cytoplasm. The nuclei were round and uniform with evenly dispersed granular chromatin. Focal nuclear pleomorphism was noted. Occasional mitoses were encountered. Immunoreactivity for synaptophysin, chromogranin and cytokeratin was diffusely positive in the tumor cells (Figure 3).

Electron microscopic examination revealed numerous cytoplasmic, membrane-bound electron-dense granules, which were pleomorphic.

### Discussion

Carcinoid tumors are the most common tumors of the appendix and small intestine. They arise from the epithelium containing argentaffin or argyrophil cells. Up

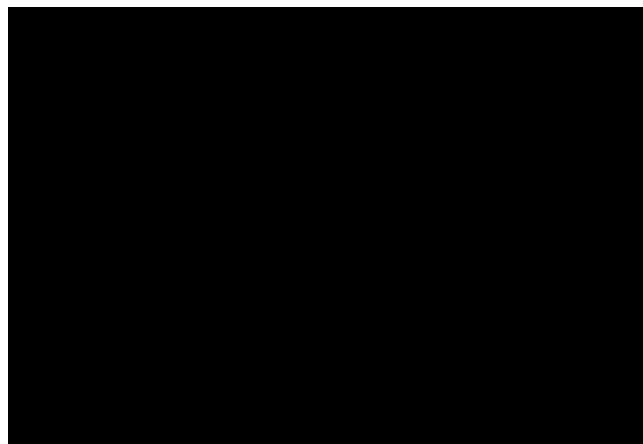


FIGURE 1. Gross specimen of the left testis shows a large mass surrounded by compressed parenchyma.

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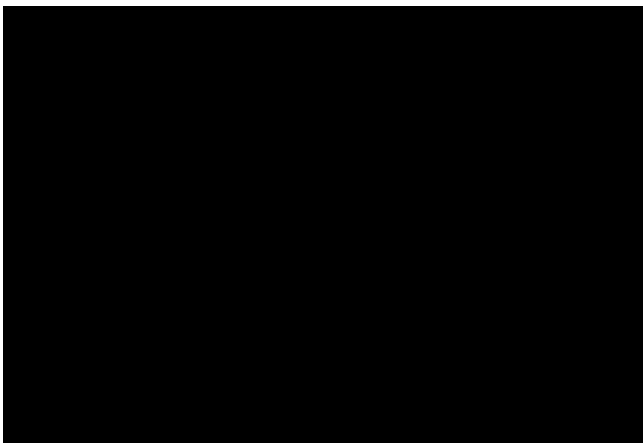


FIGURE 2. The tumor demonstrated nests, tubular and trabecular growth pattern separated by delicate fibrovascular septa (Hematoxylin and eosin, 100x).

to 15% of all carcinoids are found in the rectum, liver or genitourinary tract.<sup>2</sup> The majority of genitourinary carcinoids are associated with ovarian teratomas. Involvement of the testis by carcinoid tumor is uncommon and it comprises less than 1% of all testicular neoplasms. Fifty-seven cases of primary testicular carcinoma have been reported,<sup>1,5</sup> out of which 14 were associated with teratomatous lesions, while 43 were pure primary testicular carcinoids. The age of the patients ranged from 10 to 83 years. The tumor usually presented as a painless enlargement of the testis. The left testis was more frequently affected than the right one, while in one case the tumor was bilateral.<sup>4</sup> Some patients had a history of cryptorchidism.<sup>6,7</sup> Our case also presented with left-sided tumor, which was misdiagnosed to be a Leydig cell tumor. On the basis of the referring diagnosis, we tested the patient for the markers of Leydig cell tumor, which were all within normal range. He did not have gynecomastia, which is a feature in some of the Leydig cell tumor cases.

Pathologically, carcinoid tumor is recognized by its characteristic histology, showing nests of uniform cells with uniform nuclei and characteristic finely granular chromatin. Abrupt nuclear pleomorphism is present, which is supplemented by immunoreactivity of the tumor cells to neuroendocrine markers such as chromogranin and synaptophysin.

Ultrastructurally, carcinoid tumors have electron-dense neurosecretory granules varying in size from 75 to 500 nm and of variable morphology. They may be uniform and round or elliptical and asymmetrical.

Testicular carcinoids are divided into three categories: 1) primary in the testis; 2) primary arising in a testicular teratoma; and 3) metastatic. Due to lack of morphologic differences between primary and metastatic carcinoid tumors, it is necessary to try to exclude the presence of a

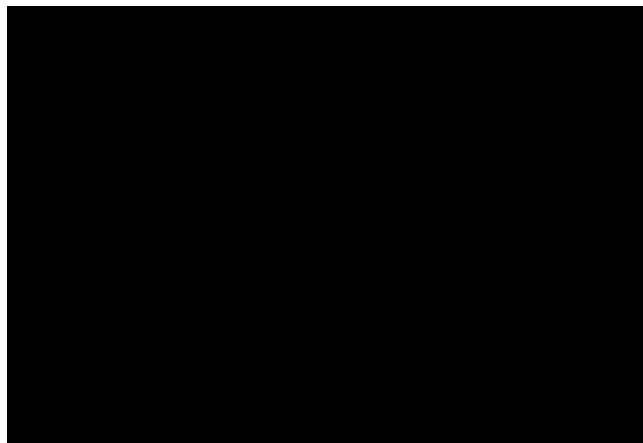


FIGURE 3. The tumor cells are strongly positive for chromogranin (LSAB, 250x).

primary tumor in another organ before making the diagnosis of primary testicular carcinoid tumor.<sup>1</sup> No other tumor was found in our patient despite extensive work-up, including 24-hour urinary 5-HIAA estimation, chest x-ray, GI contrast study and CT scan of the abdomen.

The histogenesis of apparently pure testicular carcinoid tumor is uncertain, but there are several possibilities.<sup>3</sup> The tumor might be part of a teratoma in which other elements are missing or have regressed, or it might represent monophyletic development of argentaffin cells in a teratoma. The treatment of choice for testicular carcinoid tumor is inguinal orchiectomy.<sup>8</sup> Until recently, testicular carcinoid tumor was considered to be a benign tumor.<sup>3,8</sup> However, some of these tumors develop metastases five to 19 years after the orchiectomy,<sup>3,9-11</sup> with an incidence of metastasis of about 11%. Long-term follow-up evaluation is clearly warranted for patients with this neoplasm. A simple algorithm,<sup>12</sup> which is based on urinary 5-HIAA estimation, abdominal CT scan and GI contrast studies, could make the basis for long-term follow-up. This uncommon condition is well described in the literature but is evidently the first such case reported from the Middle East.

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