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Metastatic primary testicular carcinoid tumor managed with radical orchiectomy, retroperitoneal lymph node dissection and inferior vena cava excision: Case report

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ABSTRACT

Metastatic primary testicular carcinoid tumor remains a very rare condition. We report the first case of metastatic primary testicular carcinoid tumor where along retroperitoneal lymph node dissection excision of the Inferior Vena Cava was also performed. The rarity is further emphasised by the presence of a contralateral testicular dermoid cyst. Given the features of the tumor were not in keeping with the traditional predictors of metastases (primary tumor >7.3 cm, poor differentiation and the presence of carcinoid syndrome) this case adds valuable addition to the relatively limited literature available on this rare condition.

1. Introduction

The first case of primary testicular carcinoid was reported by Simon et al. in 1954 as an element of a benign cystic teratoma[1] Primary testicular carcinoid remains a rare disease accounting for 0.23% of testicular tumors.[2] We report a case of a 22-year-old male with primary testicular carcinoid tumor with metastasis to the aorto-caval lymph nodes. He was also found to have a dermoid cyst in the contralateral testis.

2. Case presentation

A 22-year-old gentleman with no significant past medical history presented with a 1-year history of right testicular swelling and 1-week history of testicular pain. He denied any history of testicular trauma or previous infections. Scrotal examination detected an enlarged and mildly tender right testicular mass. Tumor markers were unremarkable (AFP 2, LDH 195, B HCG < 1). Ultrasonography (USS) of the scrotum revealed a right testicular solid heterogenous, hypoechoic mass with prominent internal vascularity measuring 44 × 29 × 29mm (Fig. 1). The left testis had an intra-testicular lesion which was heterogeneously echogenic however had no internal vascularity measuring 11 × 10 × 15mm.

Staging Computed tomography (CT) scan detected 2 enlarged (14 and 10 mm) aorto-caval lymph nodes. No solid organ metastasis was noted.

Patient underwent right radical inguinal orchiectomy. Histopathology revealed a well-differentiated neuro-endocrine tumor (carcinoid). There was rete testis and lymphovascular invasion noted. Immunohistochemistry showed positive staining with synaptophysin (Fig. 2). Ki-67 labelling index was 2% of tumor cells.

Following the diagnosis of the carcinoid tumor patient underwent a negative esophagogastroduodenoscopy and colonoscopy to rule out an extra testicular primary carcinoid. His serum chromogranin A level and urinary excretion of 5-hydroxyindoleacetic acid (5-HIAA) were within normal limits. Further staging whole body ^{68}Gallium-octreotate Positron emission tomography (PET) scan detected 2 enlarged (>10mm) aorto-caval and 1 enlarged precaval octreotate avid lymph node (Fig. 3a and b). No other octreotate avid lesions were noted. Left testicular lesion was not octreotate avid.

Patient underwent retroperitoneal lymph node dissection and

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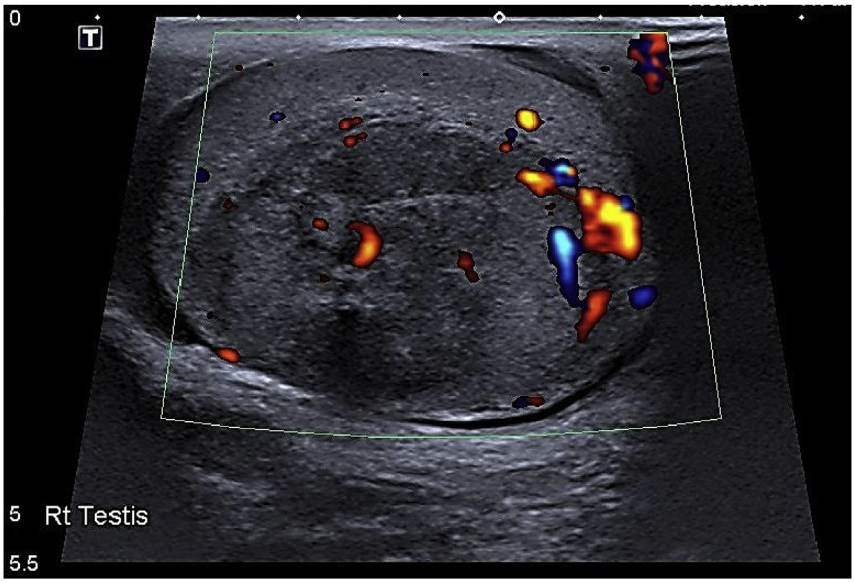


Fig. 1. USS of the right testis showing a solid heterogenous, hypoechoic mass with prominent internal vascularity.

left radical orchiectomy. At the time of the retroperitoneal lymph node dissection, the pre-caval nodes were adherent to the anterior inferior vena cava (IVC) causing compressive effect. En-bloc resection of IVC and pre-caval lymph nodes was performed and an interposition Polytetrafluoroethylene (PTFE) graft was used to replace the excised IVC.

Patient was discharged day 6 post op on long term anti-coagulation to maintain IVC graft patency.

The final histopathology revealed a left testicular 10 mm epidermoid cyst confined to testis. 3 out of the 33 lymph nodes excised were positive for well differentiated carcinoid tumor.

The patient remains well and active. His 3, 6 and 12 month post treatment ^{68}Ga-PET CT scan have shown no residual or recurrent disease.

3. Discussion

The pathophysiology of primary testicular carcinoids has not been well established. It has been described that testicular carcinoids usually occur on the background of teratoma giving the rationale that carcinoid might be a component of teratoma with regression of the remaining elements.[2,3] Twenty-five percent of primary testicular neuroendocrine tumors are associated with teratoma.[3]

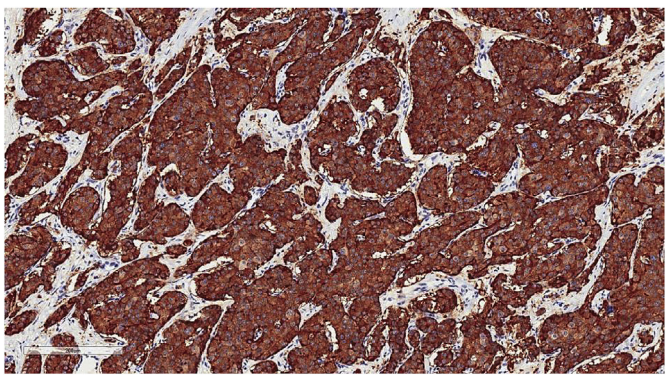
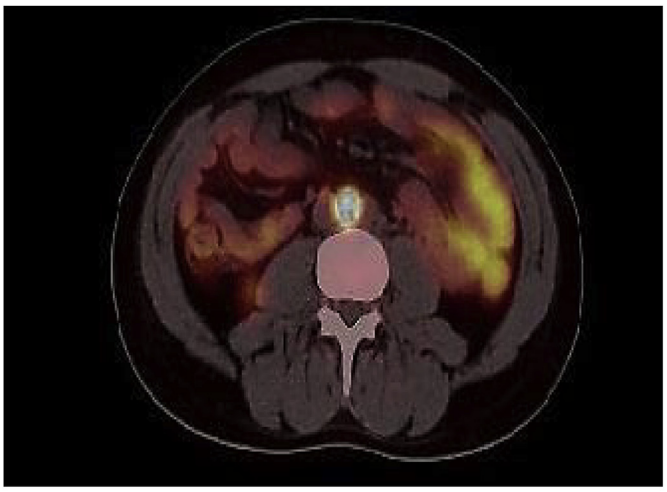


Fig. 2. Positive synaptophysin staining of right testis in primary carcinoid tumor.

Alternatively, it has also been hypothesized that Leydig cells may be the origin of testicular carcinoid tumor due to their neuroendocrine features. Mai et al. demonstrated transitional cells were present in primary testicular carcinoid tumor displaying features of both leydig cells and carcinoid tumor cells.[4] These were found to be absent in secondary testicular carcinoid tumor. There data supported that these cells probably arose from the same progenitor cells in cases of primary testicular carcinoid tumor.[4]

Most common presentation of testicular carcinoid tumor is painless testicular enlargement or palpable mass. Other presenting feature may include testicular pain or tenderness, hydrocele or very rarely cryptorchidism.[3] Carcinoid syndrome, which is a manifestation of carcinoid tumor, may be present in approximately 10% of patients.[3] Symptoms include episodic flushing, diarrhea and bronchospasm. This typically occurs when there is metastasis to liver or lungs due to the production and release of vasoactive substances such as serotonin in to systemic circulation. Patients presenting with symptoms of carcinoid syndrome and testicular swelling or mass should have serum chromogranin A and 24-h urinary 5-HIAA performed with elevated levels suggesting carcinoid pathology, higher burden of disease and increased likelihood of metastasis.[3] In the case of our patient he did not exhibit signs of carcinoid syndrome and did not have elevated serum serotonin or urinary 5-HIAA despite having metastatic disease.

Radical orchiectomy is the treatment of choice for testicular carcinoid. Once confirmed on histopathological assessment, testicular carcinoid should be investigated for an extra-testicular primary as 10% of testicular carcinoid tumors are secondary to metastasis from an extra-testicular primary.[2] There are no morphological or histological differences between primary and metastatic carcinoid and therefore the diagnosis of primary testicular is only made once an extra-testicular primary is ruled out. Staging scan begins with CT-scan to detect for any metastasis.



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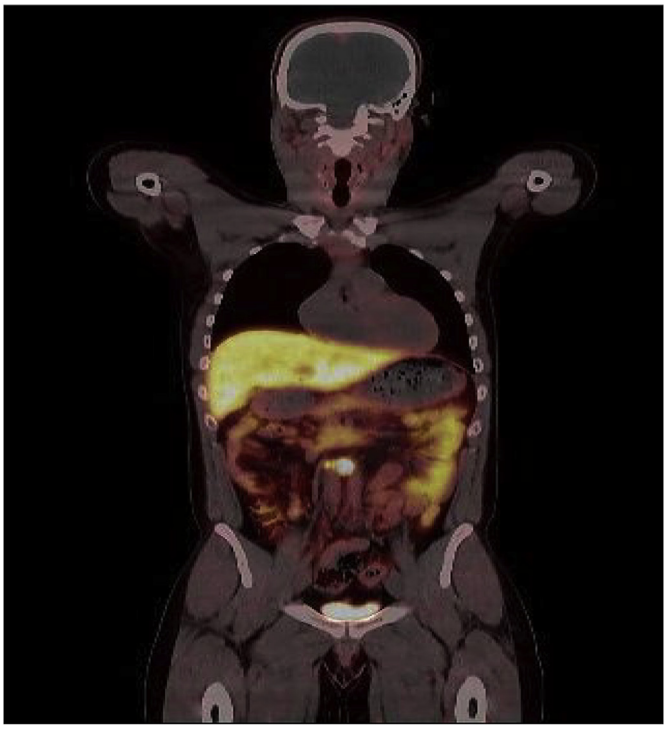

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Fig. 3. a and 3b. ^{68}Ga-PET scan demonstrating increased uptake in the aorto-caval lymph nodes indicating metastatic disease.

Somatostatin receptor-based imaging such as indium-111 labeled octreotide scintigraphy is also of great utility in looking for primaries and determining the extent of metastatic disease.[5] However, more recently there has been the introduction of the ^{68}Ga-labeled radioligands, which is a somatostatin receptor ligand and whose uptake is measured by PET scan. Comparison studies between ^{68}Ga-PET and standard imaging techniques (CT, OctreoScan) have universally demonstrated the superiority of ^{68}Ga-PET in detection of primary tumors and metastases.[5]

Testicular carcinoids rarely metastasize with the overall incidence between 11% and 17%.[3] No standard treatment has been established for metastatic carcinoid tumors. Chemotherapy is of minimal benefit and wherever possible, resection of metastatic deposits should be performed. Traditionally the size of primary tumor (>7.3 cm), poor differentiation of the tumors, and the presence of carcinoid syndrome are independent predictors for the development of metastases.[3] However it is important to note none of these features were present in our patient despite his metastatic disease.

Given the rarity of metastatic primary testicular carcinoid tumor there is no clear consensus on follow up. The unfavourable outcomes of these patients' necessitate regular review and close surveillance. This should include history, physical examination, 24-h urinary 5-HIAA and surveillance imaging ideally with ^{68}Ga-PET to detect early recurrences.[2,3]

Conflict of interest

All authors declare no conflict of interest.

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