Neuroendocrine Tumor (Carcinoid Tumor) Arising in Postpubertal Testicular Teratoma In A 32 Year Old Male: Case Report & Literature Review

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INTRODUCTION:

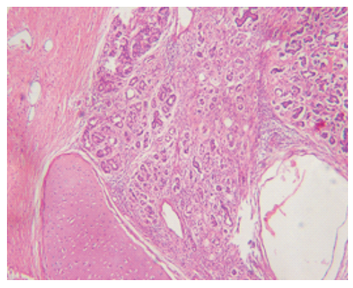
Primary carcinoid tumors are found commonly in the gastrointestinal tract, respiratory, hepatobiliary and genitourinary systems. A significant number of carcinoid tumors occurring in teratomas of the ovary have been reported. However, primary testicular carcinoid tumors arising within teratomas are extremely rare. Pure testicular teratoma accounts for 2.7% to 3% of all germ cell tumors in adult where it behaves as a malignant neoplasm. While testicular carcinoid tumours account for less than 1% of all testicular neoplasms.We herein, report a case of primary carcinoid tumor arising in postpubertal testicular teratoma.

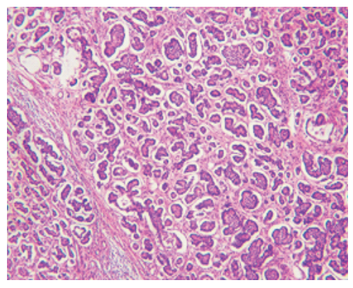
CASE PRESENTATION:

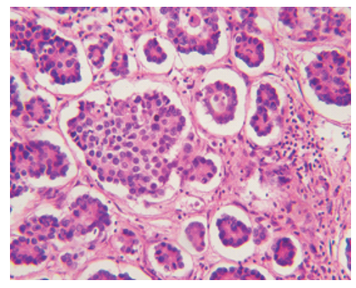
A 32-year old male presented with an abdominal swelling. There was history of undescended left testis. On examination, a firm mass was found in para aortic region of abdomen. Beta human chorionic gonadotropin [β-HCG], Alfa-fetoprotein [AFP] and Lactate dehydrogenase (LDH) levels were found to be elevated.

Macroscopically, a grey white well circumscribed tumor of 2.5x 2.0x1.8cm within testicular parenchyma was identified.

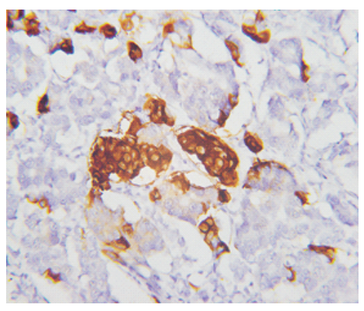
Figure 1: Grossly tumor shows solid and cystic cut surface.



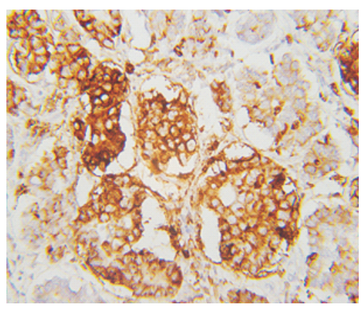




Histological examination revealed a neoplasm showing organoid arrangement of round blue cells with salt and pepper chromatin, rosette formation and occasional mitoses. Features of mature teratoma were seen in the background.
The tumor was classified as pT1 Nx Mx as per the American Joint Committee on Cancer (AJCC) TNM staging for testicular cancers.



(A)Synaptophysin



(B) Chromogranin- A

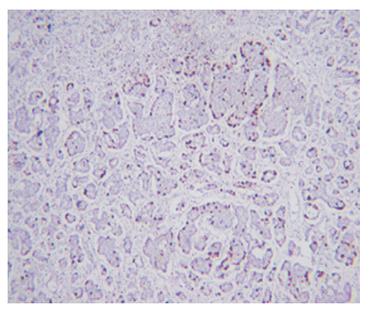


Figure 3: (C) Ki-67 Proliferation index

Immunohistochemistry revealed positivity for Synaptophysin and Chromogranin-A with a Ki-67 index of 3-20% which confirmed a well differentiated neuroendocrine tumor (Grade 2) arising in postpubertal teratoma.

DISCUSSION:

Teratomas are neoplasms comprising of tissues derived from more than one germ cell layer. These can be classified, on the basis of location and cellular differentiation into gonadal, extra- gonadal subtypes and mature and immature forms respectively. 3–6% of benign teratomas can undergo malignant transformation.

Malignant transformation of the somatic component of testicular teratomas is rare among which squamous cell carcinoma accounts for 70-88% of cases. Other reported somatic malignancies include: Adeno-squamous carcinoma, sebaceous carcinoma, malignant melanoma and rarely sarcomas. Neuroendocrine /carcinoid tumor arising within a mature teratoma is extremely rare. Our case belongs to a primary carcinoid tumour arising in a testicular teratoma.

Carcinoid tumors are neuroendocrine tumors which arise from enterochromaffin/ Kulchitsky cells. Neuroendocrine tumor of the testis accounts for less than 1% of all testicular neoplasms and occur either as a primary testicular neuroendocrine tumor; or metastatic neuroendocrine tumor to the testis. The primary testicular neuroendocrine tumor of testis can be pure or associated with teratoma. More than 60 cases of neuroendocrine tumor of testis have been reported to date with ages ranging from 10years to 83 years, with highest incidence in fifth and sixth decade. Most patients present with unilateral painless testicular mass. 16% of patients present with symptoms of carcinoid tumor syndrome. 11% of primary testicular carcinoid tumors are associated with metastasis.

The pathogenesis of primary testicular carcinoid is controversial. They are probably derived from interspersed neuroendocrine cells associated with abnormalities such as metaplasia induced by chronic inflammation, entrapped neural crests or pancreatic tissue during embryogenesis, activation of gene sequences common to neuroendocrine programed cells in multipotent stem cells, or concurrent congenital abnormalities.The origin of the testicular carcinoid tumour is also said to be a one-sided development of the mature teratoma or differentiation of toti-potential germ cells in the testis to argentaffin-like cells.

Carcinoid tumours produce vasoactive products which can cause carcinoid syndrome, characterized by sweating, flushing, wheezing, diarrhea, abdominal pain, cardiac valvular fibrosis and pellagra dermatosis. Carcinoid syndrome occurs in 8% of patients with a carcinoid tumour, especially in cases with metastasis, but our patient did not have any symptoms of carcinoid syndrome.

These are graded on basis of Ki-67 index and/or mitotic count. Our case showed Ki-67 index of 2-30 %. This makes the final diagnosis of well differentiated neuroendocrine tumor, Grade 2, arising in postpubertal teratoma. All previous reported cases had Ki-67 index of < 1 %.

Our case is Stage 1 as is localized to testis while previously reported case is with distant lymph node metastasis.

Carcinoid tumors have excellent prognosis for Stage I disease with 5 years survival rate of 93%. But it decreases to 20-30% for distant metastatic disease. The association with teratoma has a prognostic value depending on the age and stage. Radical orchidectomy is the treatment of choice. Carcinoid tumors in general have a very poor response to chemotherapy and radiotherapy. Symptomatic treatment should be given to the patients with carcinoid syndrome. Octreotide, a somatostatin analogue inhibits the release of hormones and neurotransmitters and thus causing symptomatic improvement.

Long-term follow-up is indicated because of tumor metastatic potential. Regular physical examination, serum hormones level, abdominal CT scan, and gastrointestinal contrast can be exercised for follow-up evaluation.

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CONCLUSION:

In conclusion, primary carcinoid tumor arising in postpubertal teratoma is very rare assosciated with adverse prognosis. Earlier diagnosis followed by surgical intervension with long term follow-up can lead to better outcome in patients with this rare malignancy.

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