Primary extragonadal yolk sac tumor originating from adrenal gland

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Extragonadal germ cell tumors are germ cell tumors with no evidence of a primary tumor within the gonads, most often located in the mediastinum or retroperitoneum. We present an extragonadal yolk sac tumor that presented as an adrenal carcinoma and required left adrenalectomy, nephrectomy, and significant IVC thrombectomy. This case, to our knowledge, is the first documented case of extragonadal yolk sac tumor originating from the adrenal gland.

Key Words: adrenal gland, extragonadal yolk sac tumor

Introduction

In this case we present a primary yolk sac tumor within the left adrenal gland with invasion into the left renal vein and IVC. Extragonadal germ cell tumors (GCTs) are germ cell tumors that have no evidence of a primary tumor in the testes or ovaries and are malignant in nature.[1] While primary GCT in the mediastinum are common, an extragonadal GCT located in the adrenal gland, is exceedingly rare.[2] This is the first documented case, to our knowledge, to describe a primary yolk sac tumor of the adrenal gland. This case provides an example of a rare presentation of primary extragonadal GCT while highlighting diagnostic criteria for distinguishing between primary extragonadal GCTs and GCTs with metastasis as well as their treatment modalities.

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Case report

A 76-year-old Sudanese male presented to our outpatient clinic for a large left adrenal mass. One year prior the mass was incidentally discovered during cardiac evaluation and was approximately 1 cm in size. Unfortunately, at that time the patient was lost to follow up. Later that year during work up secondary to a fall the mass was found to be 11.1 cm x 10 cm x 10.8 cm. At this time he underwent biopsy and laboratory testing to assess for pheochromocytoma, adrenal cortical carcinoma, hyperandrogenism, or Cushing's. This work up was negative for a functional adrenal tumor and his biopsy from the outside hospital was read as adrenal cortical carcinoma. The patient was referred to an Academic Urology clinic for what was believed to be T4 adrenal carcinoma with IVC thrombus and pulmonary metastatic disease. MRI was obtained and showed a heterogeneous mass suggestive of adrenocortical carcinoma with tumor thrombus extending into the left renal vein and infradiaphragmatic inferior vena cava, Figure 1. An appropriate discussion was had with the patient and his

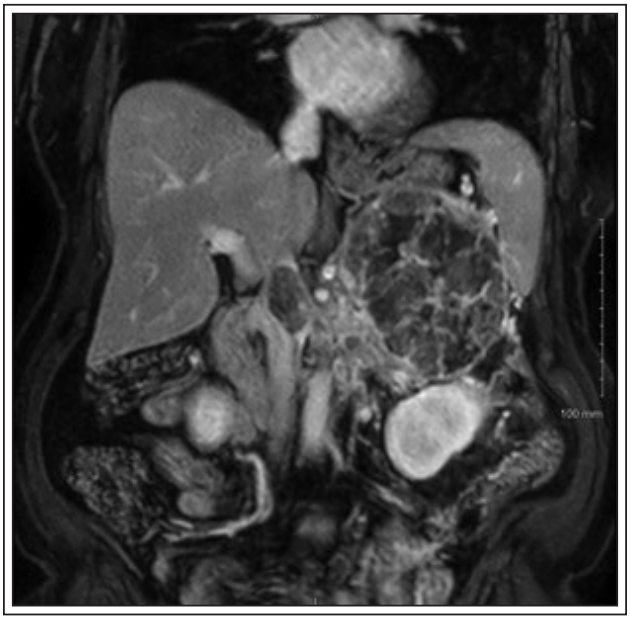

Figure 1. Heterogeneous left adrenal mass suggestive of adrenocortical carcinoma with tumor thrombus extending into left renal vein.

family in regards to the options, and surgical consent was obtained. The patient was taken to the operating room where left adrenalectomy, nephrectomy, local lymph node dissection, and IVC thrombectomy was performed. Postoperatively the patient did well, he was discharged on postoperative day 9 and pathology revealed a high grade malignant neoplasm consistent with extragonadal yolk sac tumor, with no residual adrenal parenchyma, and no invasion of into the renal parenchyma. AFP level on POD 9 when pathology was returned was 10,889 ng/mL. Testicular examination was normal and an ultrasound showed small bi-lateral varices without presence of a mass/primary source. The patient underwent four cycles of VIP chemotherapy with etoposide, ifosamide, and cisplatin. The patient is now 5 months from surgery and most recent AFP was 5.8 without evidence of local recurrence and shrinkage in his pulmonary disease.

Discussion

Extragonadal germ cell tumors (GCTs) are neoplasms that arise via malignant transformation of germ cells outside of the testes or ovaries. Approximately 2 to 5 percent of all germ cell tumors are extragonadal GCTs. They are classified in the same manner as gonadal GCTs (seminomas and nonseminomas) and have the same serological, histological and cytogenetic characteristics as their gonadal counterparts.[3] It is believed extragonadal GCTs originate from germ cell tissue which was erroneously placed during embryogenesis and subsequently undergoes malignant transformation. As germ cells develop from the ectoderm along the urogenital ridge most extracellular GCTs are found in the median axis within the mediastinum (50%-70%) and retroperitoneum (12%-40%).[4] However, extragonadal GCT's can be rarely be found in other locations.[5]

Presentation of extragonadal GCTs is usually secondary to mass effect and may symptoms often include abdominal pain, back pain, fever, cough, chest pain, and vena cava syndrome.[6,7] Diagnosis of extragonadal GCTs is made via tissue biopsy followed by scrotal ultrasound with no evidence of a primary tumor. Testicular biopsy is not recommended.[8] Tumor markers including alpha-fetoprotein (AFP), human chorionic gonadotropin (β-HCG) and lactate dehydrogenase (LDH) should be obtained to assist in pathological diagnosis and long term monitoring.

Few resources aid in recommendations and treatment of extragonadal GCT's, in general guidelines established for GCT are used with cisplatinum being the cornerstone of chemotherapy. Non-seminomatous tumors receive four cycles of VIP (etoposide, ifosfamide, and cisplatin) or BEP (bleomycin, etoposide, and cisplatin) followed by resection of residual tumor.[9] Seminomatous lesions are treated with three cycles of BEP with resection of tumor being optional and based on responsiveness of seminomas to chemotherapy.[10]

When extragonadal GCTs of seminomatous type are treated with platinum based chemotherapy patients have an excellent 5 year survival rate. Following BEP both retroperitoneal and mediastinal extragonadal GCTs have an overall survival rate of 88% at 5 years and a 5 year progression free survival rate of 87%.[11,12] In non-seminomatous extragonadal GCTs survival rates are significantly lower and associated with location. Following platinum-based induction chemotherapy in non-seminomatous extragonadal GCTs, with or without surgery, 49% of those with mediastinal involvement are alive at 19 months, compared to 63% being alive at 29 months in those with retroperitoneal involvement.[11]

Our case presents, to our knowledge, is the first documented presentation of extragonadal yolk sac tumor originating from the adrenal gland and included tumor thrombus into the left renal vein. However, with 2-5 percent of all germ cell tumors being extragonadal, rather than metastatic, it is important for a urologist to be familiar with the diagnosis and treatment of this disease. □

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