

Case of a concurrent renal mass and extragonadal retroperitoneal teratoma

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AMBANI SN, JACOBS BL, PEREPLETCHIKOV AM, HREBINKO JR RL. Case of a concurrent renal mass and extragonadal retroperitoneal teratoma. The Canadian Journal of Urology. 2009;16(2):4607-4610.

The presentation of a synchronous renal cell carcinoma (RCC) and germ cell tumor (GCT) is rare. We report the case of a 57 year-old male who presented with a right renal mass and retroperitoneal lymphadenopathy. He underwent a successful right partial nephrectomy

and retroperitoneal lymph node dissection, and the subsequent pathology revealed a stage I clear cell RCC and a retroperitoneal teratoma with a component of benign prostatic tissue. We briefly discuss the rarity of this occurrence, the pathological features that helped support this diagnosis, and the likely etiologies of these synchronous lesions.

Key Words: renal cell carcinoma, extragonadal germ cell tumor, retroperitoneum, teratoma

Case

A 57-year-old male was referred to our clinic after a magnetic resonance image (MRI), which was done as

part of a work up for hypertension, revealed a 2 cm right renal mass abutting the collecting system and a 5 cm x 3 cm area of retroperitoneal lymphadenopathy; there was no other evidence of metastatic disease. Subsequently, a computed tomography (CT) with and without intravenous contrast and delayed images was done to better characterize these lesions, Figure 1. The patient was asymptomatic with no complaints of flank pain or gross hematuria. Other than hypertension, his past medical history was notable for coronary artery disease,

Accepted for publication December 2008

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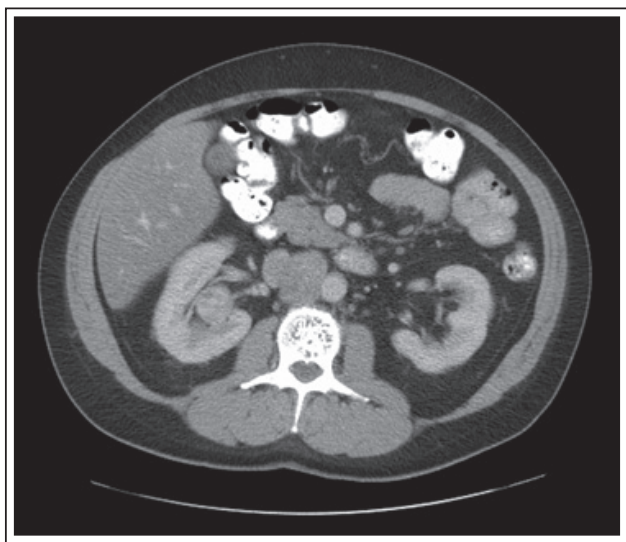


Figure 1. Computed tomography with intravenous contrast showing a centrally located right renal mass and interaortocaval lymphadenopathy.

diabetes mellitus, and membranous nephropathy, with a baseline creatinine of 2.0 mg/dl. Additional pertinent history included a prostate-specific antigen (PSA) of 0.5 ng/ml and a negative prostate biopsy. His family history was negative for renal, prostate, and testicular cancers, and he had a remote smoking history. On exam, his abdomen was soft with no palpable masses, and he had a slightly atrophic left testicle and normal right testicle without any palpable nodules. His digital rectal exam was normal.

The patient underwent an uneventful right partial nephrectomy with enucleation of the renal mass and a retroperitoneal lymph node dissection in which all visible retroperitoneal disease was removed. Postoperatively, the patient developed a perirenal urine collection that was drained percutaneously. In addition, a ureteral stent was placed, and the patient was discharged home in good condition.

Two weeks later at his follow up appointment, the pathology findings were reviewed. The renal lesion was pathological stage T1a, Fuhrman nuclear grade I/IV RCC conventional (clear cell) type with negative surgical margins.¹ The nodal retroperitoneal mass did not reveal any evidence of metastatic RCC and was most consistent with a primary extragonadal retroperitoneal teratoma.

The patient was referred to a medical oncologist, who obtained tumor markers: alpha-fetoprotein (AFP), β -human chorionic gonadotropin (β -HCG), and lactic dehydrogenase (LDH) were all within normal limits. A scrotal ultrasound was obtained, which was normal, and no further intervention was recommended at this time.

Discussion

The finding of a synchronous RCC and a primary extragonadal retroperitoneal GCT is unique and limited, having only been described once in the literature.² The use of immunohistochemical stains helped to support the diagnosis. On hematoxylin and eosin (H&E) stain, irregular glandular structures with discrete, crisp, sharp luminal borders were seen along with some glands with branching, focally cribriform lumina entrapped in primitive connective tissue, Figure 2a. The cellular component was composed of amphophilic glossy appearing cells without any particular differentiation, Figures 2b and 2c. No evidence of atypia was identified. Immunohistochemical staining showed patchy expression of PSA, Figure 3a, prostatic acid phosphatase (PSAP), androgen receptor (AR), Figure 3b, and alpha-methyl-acyl-CoA racemase; cytokeratin 903 and p63 stains highlighted scattered basal cells in some glands harboring benign prostatic tissue origin. The morphologic findings along with the immunohistochemical profile of these retroperitoneal structures were most compatible with a mature teratoma.

Several case reports describe patients with both RCC's and GCT's, although most of these reveal RCC's that develop subsequent to treatment of the GCT. Multiple studies have shown that there is an increased incidence of a second primary cancer (including kidney cancer) following testicular cancer, largely attributed to the effects of radiation therapy; these second primary tumors often develop several years

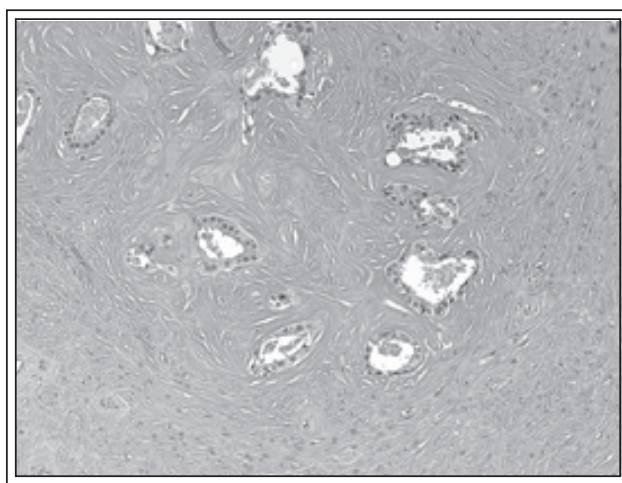


Figure 2a. Dense immature: mesenchymal connective tissue with extensive hyalinosis and entrapped well formed glandular structures, showing a haphazard architectural pattern. H&E, X100.

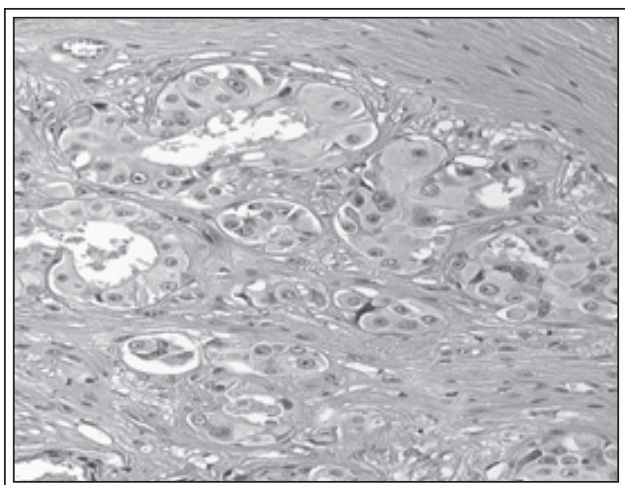


Figure 2b. Haphazardly arranged primitive glands with glossy cytoplasm, demonstrating probable sebaceous glandular differentiation. Note eosinophilic intracytoplasmic keratin containing globules. H&E, X200.

later.³⁻⁵ Fewer reports have described the finding of a synchronous RCC and GCT. Davis et al described two cases of synchronous RCC and GCT and concluded that a testicular tumor should be considered in an individual between 20-50 years old with a renal mass and generalized retroperitoneal adenopathy.⁶ White et al reported a concurrent testicular teratoma and RCC and suggested that genetic influences may influence

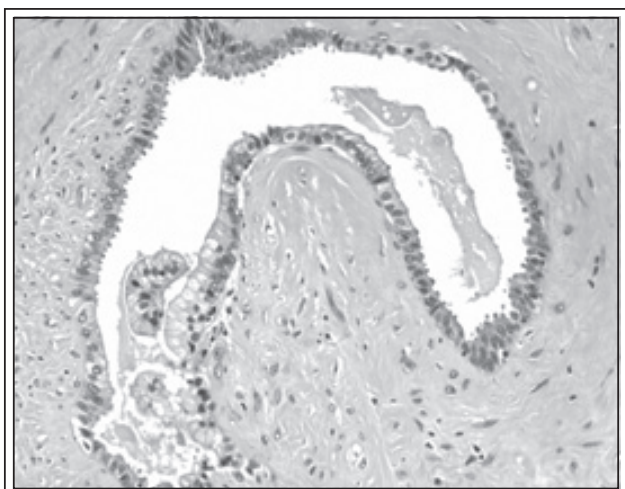


Figure 2c. Glandular structure lined by primitive epithelium with hobnailing and slightly amphophilic cytoplasmic features, surrounded by immature stromal elements. No atypia or mitotic activity is seen. H&E, X200.

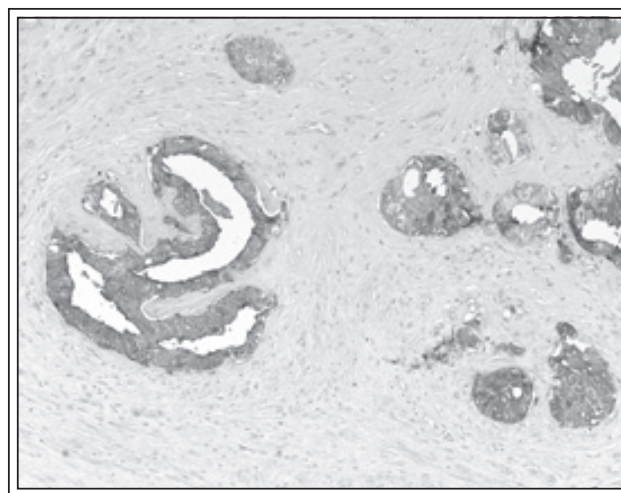


Figure 3a. Moderate to strong cytoplasmic PSA immunohistochemical staining focally present in teratomatous glands. PSA, X100.

the development of uro-genital tumors.⁷ In support of this, chromosomal studies have shown a loss of heterozygosity at 1p in 30% of renal cell cancers and 37% of testicular cancers.⁸ Dieckmann et al evaluated 23 patients who developed nontesticular malignancies in addition to testicular cancer, of which three occurred simultaneously, and argued that treatment related factors could not contribute to the pathogenesis of synchronous lesions and that genetic or environmental factors were involved.⁴ Common risk factors for RCC and GCT's do not appear to overlap, but it is conceivable that a sharing of pathogenic risk factors may affect both

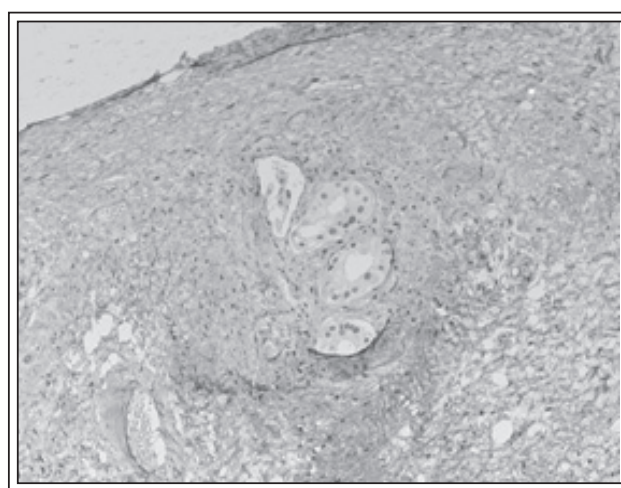


Figure 3b. Moderate nuclear androgen receptor (AR) immunoreactivity in some of the retroperitoneal glandular astructures. AR, X100.

organs during embryogenesis since, at that time, the kidney and testis are in close proximity.⁹

To our knowledge, this is the first case in the English literature to describe the synchronous presentation of an RCC and a primary extragonadal GCT. A prior report was described in the Japanese literature.² This case is further compounded by the presence of prostatic tissue in the teratoma, which is also exceedingly rare in males.¹⁰ Because of the presence of prostatic tissue, we contemplated whether the lymphadenopathy represented metastatic prostate cancer. Microscopically, however, the glands clearly had basal cells, supporting the diagnosis of benign prostate tissue. In addition, the patient's PSA of 0.5 ng/ml, negative digital rectal exam, and prior negative prostate biopsy made the diagnosis of metastatic prostate cancer unlikely.

In summary, the patient was found to have a 2 cm right renal lesion with focal retroperitoneal lymphadenopathy, a benign testicular exam other than a slightly atrophic left testis, and a history of a normal PSA and negative prostate biopsy on presentation. Although the renal lesion was small, it was fairly centrally located, and the lymphadenopathy was in a typical location for metastatic RCC. Before reviewing the pathology, at no point did we think the adenopathy was anything but metastatic RCC.

This case is an example of a patient presenting with synchronous RCC and primary extragonadal retroperitoneal teratoma with a component of benign prostatic tissue. Synchronous RCC's and GCT's are rare but do occur, and the reasons for their co-existence likely involve genetic and/or environmental factors. As stated in Davis et al,⁶ younger patients with a renal mass and concomitant generalized retroperitoneal lymphadenopathy should be evaluated for a testicular tumor as well. □

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