1500 gram suprarenal mass: a case report

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Adrenocortical carcinoma can have a clinical presentation that mimics a primary renal tumor. We describe a case of a 47-year-old male who presented with flank pain, weight loss, and a 14 cm mass arising from the upper pole of the right kidney on imaging. Upon surgical resection he was found to have a 1500 gram stage II adrenocortical carcinoma. The clinical features, pathologic findings, grading criteria, and differential diagnosis of adrenocortical carcinoma are reviewed.

Key Words: adrenocortical carcinoma, adrenal tumor, Weiss criteria

Case report

A 47-year-old Caucasian male presented with a 6-month history of right flank discomfort, and an 11 pound weight loss. His past medical history was significant for obesity, hypertension, dyslipidemia, mild type II diabetes mellitus, alcohol use, and a 20-pack-year smoking history. Physical examination was consistent with a large right upper quadrant abdominal mass. A CT scan of the abdomen and pelvis revealed a 12 cm x 13 cm x 13 cm right upper pole renal mass. MRI confirmed a 12.8 cm x 14.0 cm x 11.4 cm mass arising from the upper pole of the right kidney with the compression of the inferior vena cava at the level of the right renal vein, Figures 1 and 2. There was no evidence of a venous thrombus. Additional findings included mild periaortic and peri-hilar lymphadenopathy. Metastatic workup, including chest x-ray and bone scan, was negative.

The patient was taken to the operating room for a right radical nephrectomy for a presumed large renal cell carcinoma. A right radical nephrectomy and a

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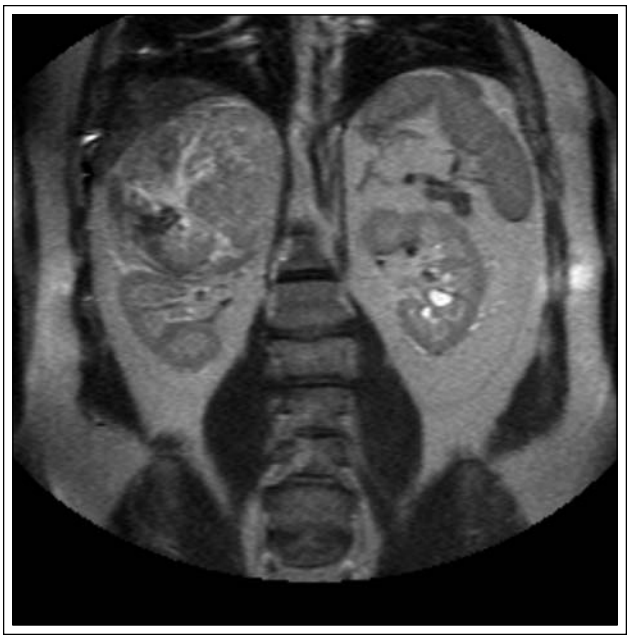

Figure 1. T2-weighted coronal MRI of abdomen revealed a large heterogeneous mass at the upper pole of the right kidney.

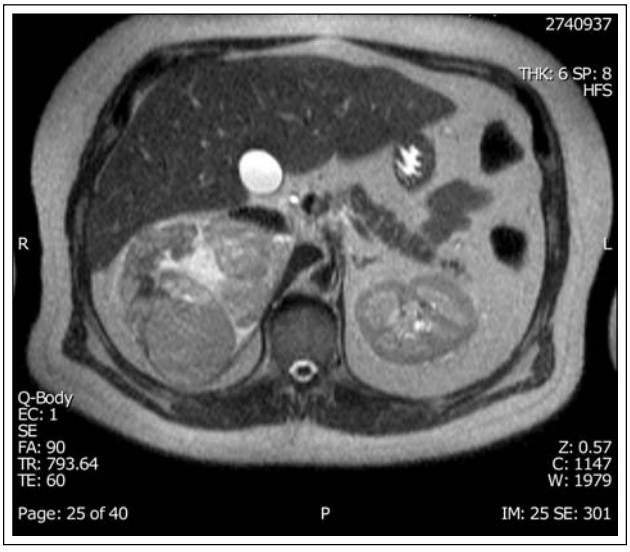

Figure 2. T2-weighted axial MRI of the abdomen revealed a large heterogeneous mass at the level of the right kidney compressing the inferior vena cava.

paracaval lymphadenectomy were performed. Intraoperative findings revealed a lesion that was not contained within the right kidney and was adherent to the liver capsule. The patient tolerated procedure well and was discharged from the hospital uneventfully on day 5. He has remained disease free 2 years after post-operative follow-up.

Pathologic findings

Gross pathologic examination revealed a 14 cm x 13 cm x 6 cm encapsulated suprarenal mass weighing 1500 grams, characterized by soft, yellow-red tissue of heterogeneous consistency, with traversing fibrous bands and widespread areas of hemorrhage and necrosis, Figure 3.

Representative sections were submitted in buffered formalin, embedded in paraffin, serially sectioned to 5 micrometers, and stained with hemotoxylin and eosin. Microscopic examination revealed a hypercellular lesion comprised of numerous cells containing eosinophilic cytoplasm with hyperchromatic nuclei, profound nuclear atypia, multi-lobulated nuclei, and nuclear pseudoinclusions, with large areas of hemorrhage, necrosis, and chronic inflammation, Figures 4 and 5. There were focal areas with hyaline globules, trabecular architecture, and oncocytic features, Figures 6-9. The mitotic rate was 8 per 50 high power fields, with many atypical mitotic figures, Figure 10. Invasion was not detected.

Discussion

The differential diagnosis of a suprarenal mass in an adult includes hepatocellular carcinoma, renal cell carcinoma, adrenal hyperplasia, adrenal adenoma, adrenocortical carcinoma, pheochromocytoma, ganglioneuroma, adrenal myelolipoma, angiomyelolipoma, malignant epithelial carcinoma, epithelioid angiosarcoma, neurinoma, primary adrenal lymphoma, composite adrenal tumors, and a malignancy metastatic to the adrenal gland.[1,2,5-8] Additionally, infectious sources may include *Mycobacterium tuberculosis* and histoplasmosis.[1]

There is no clinical or pathologic evidence that this tumor has metastasized from another site, and the gross and microscopic features are consistent with a primary adrenal tumor. The differential diagnosis of the most common primary adrenal tumors in an adult includes adrenal adenoma, pheochromocytoma, adrenal oncocytoma, adrenal lymphoma, adrenal myelolipoma, and adrenocortical carcinoma.

Adrenal adenomas are typically under 5 cm in diameter and do not exhibit mitoses. Pheochromocytoma is typically under 10 cm in diameter and contains ganglion cells or cells with granular cytoplasm arranged in a Zellenballen pattern. Adrenal oncocytoma is extremely rare and is composed exclusively of oncocytic cells—in this

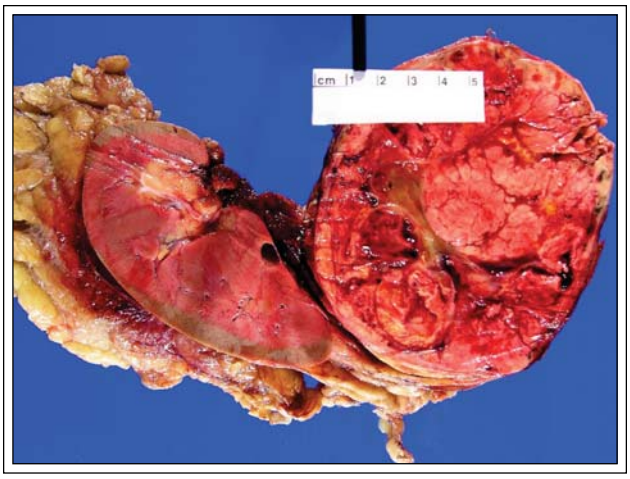

Figure 3. Gross nephrectomy specimen revealed a 14 cm x 13 cm x 6 cm encapsulated suprarenal mass.

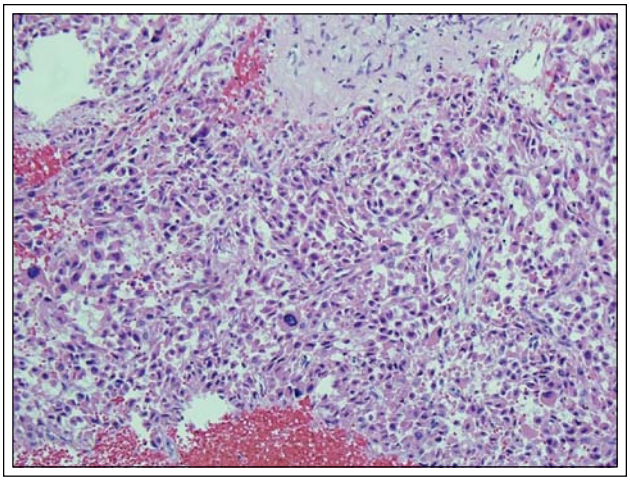

Figure 4. The tumor consists of cells with eosinophilic cytoplasm and profound nuclear atypia amid areas of necrosis and hemorrhage. 20x

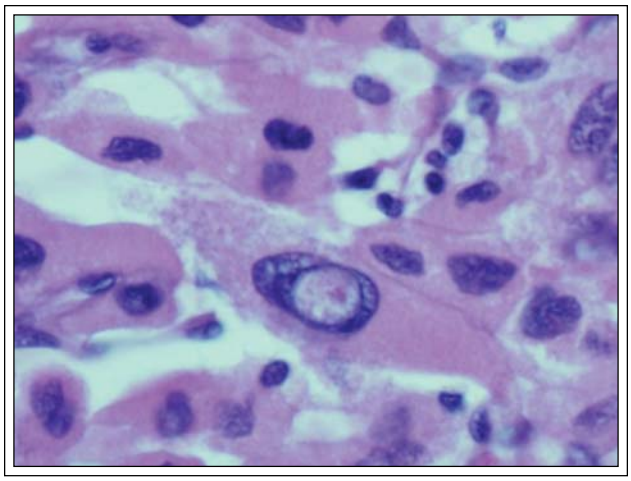

Figure 5. Nuclear pseudo-inclusions are may be observed in benign and malignant adrenal tumors, and represent an infolding of the cytoplasmic membrane into the nucleus.

case, oncocytic cells were a rare finding. Adrenal myelolipoma is a benign neoplasm comprised of hematopoetic elements interspersed with mature adipose tissue, and is not suggested by the histology of this case. The gross morphology of large size (over 100 grams) and the extent of confluent necrosis suggest that this mass is likely to be adrenocortical carcinoma or primary adrenal lymphoma; however, primary adrenal lymphoma is characterized by a predominance of lymphocytes, which were not characteristic of this tumor.

Tumors that meet at least 3 of the 9 Weiss Criteria for malignancy in adrenal neoplasms may be classified as adrenocortical carcinoma, Table 1.[4,10] The microscopic features detected in this tumor meet 6 of the 9 Weiss Criteria. Because this is a primary tumor in the adrenal

TABLE 1. **Weiss criteria for adrenal tumors**[4,10]

Weiss criteria definition	Feature	Detected in this case
Fuhrman's grade III or IV	High nuclear grade	Yes
> 5/50 high power fields	Mitotically active*	Yes
Abnormal spindles or chromosome distribution	Atypical mitotic figures*	Yes
> 75% of tumor, rather than clear cells	Eosinophilic tumor cytoplasm	Yes
> 1/3 tumor area	Diffuse growth pattern	Yes
Involving confluent nests of cells or larger areas	Necrosis	Yes
Vessel with smooth muscle in wall	Venous invasion*	No
Vascular space without smooth muscle	Sinusoidal invasion	No
Into/through capsule, with stromal reaction	Capsular invasion	No

*Found only in malignant tumors

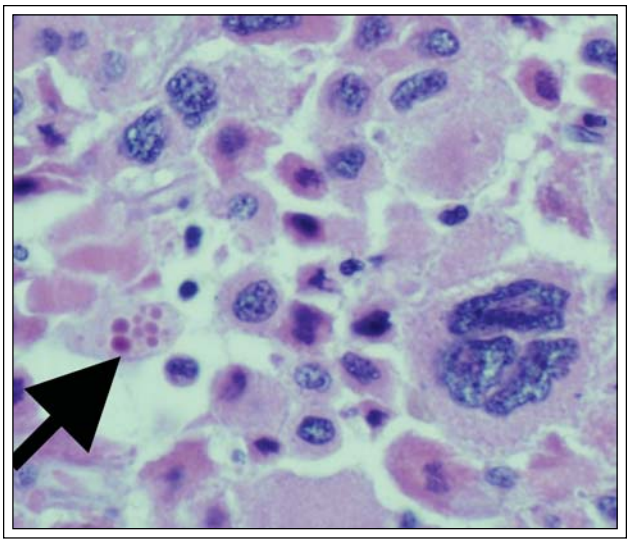

Figure 6. Ovoid, deeply eosinophilic intracytoplasmic hyaline globules are seen in cells of adrenal origin and in adrenal-derived tumors such as adrenocortical carcinoma and pheochromocytoma.

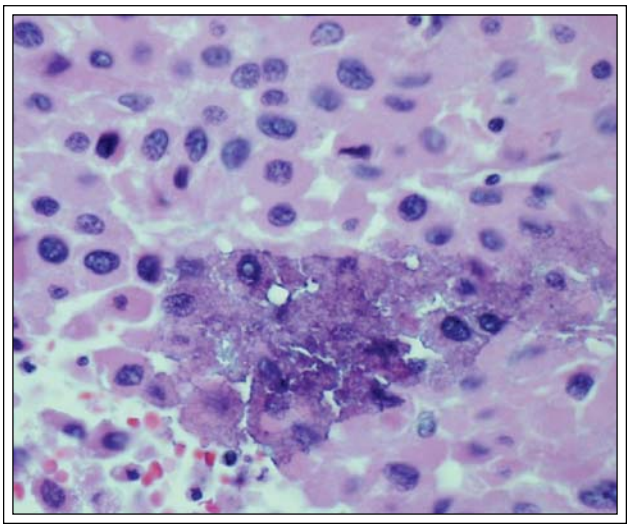

Figure 7. Foci of calcification are common in malignant adrenal tumors.

gland that is greater than 5 cm in diameter and there is no detectable invasion or extension, it fulfills classification criteria for stage II. Clinically, stage II tumors as large as 1800 grams have shown benign behavior, and the 5 year survival is 13%-57%, Table 2.[2]

The prognosis of adrenocortical carcinoma is generally poor, with a median survival of 18 months and an overall 5 year survival of 30%.[3] Survival is related to stage, and mitotic rate is an independent predictor of survival.[3] The only factor that improves survival is surgical resection.[2]

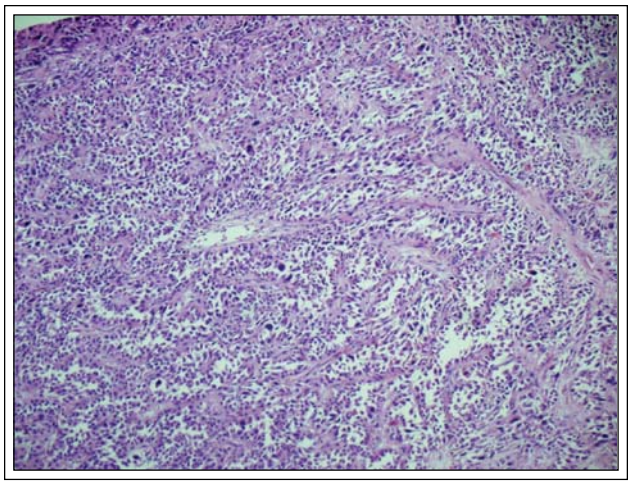

Figure 8. A trabecular architectural pattern can occur in adrenocortical carcinoma.

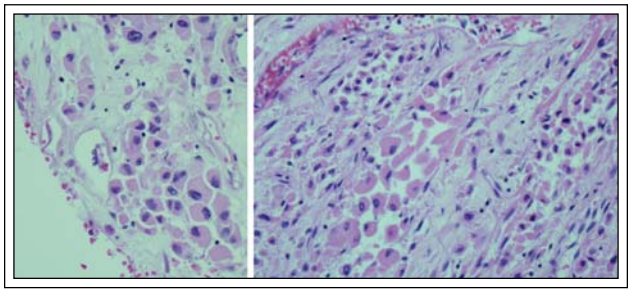

Figure 9. Focal areas of cells with oncocytic features can occur in adrenocortical carcinoma.

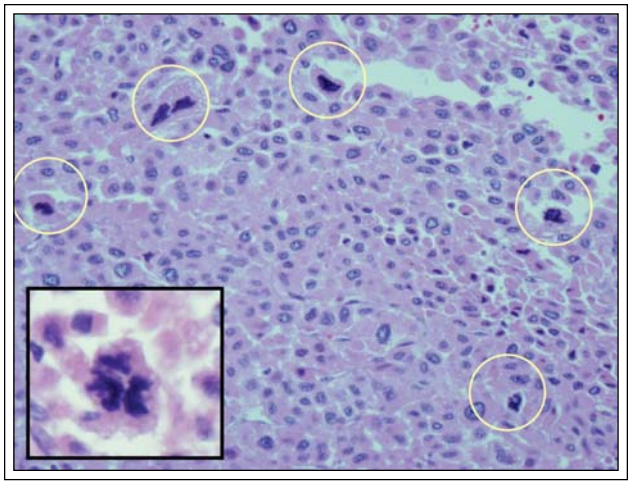

Figure 10. At least six mitotic figures are detectable in this 40x field. Cytologic borders are well-defined, typical of adrenocortical carcinoma. Lower left: atypical mitotic figure is highly suggestive of malignancy, 60x.

TABLE 2. **Staging/prognosis of adrenocortical carcinoma[2]**

Stage	Description	Percentage of patients at diagnosis	Percentage 5 year survival
I	primary, < 5 cm	21%	30%-45%
II	primary, > 5cm	20%	12.5%-57%
III	primary, limited extension into fat/ lymph nodes	20%	5%-18%
IV	primary, invasion into adjacent organs/spread to distant sites	39%	0%

Metastatic spread is discovered in approximately 20% of adrenocortical carcinoma patients at autopsy.[3] The mitotic rate and venous invasion correlate with metastasizing behavior.[3] The most common metastasis occurs in liver (48%), lung (45%), lymph node (29%), and bone (13%).[3] Local invasion occurs in kidneys (26%) or the inferior vena cava (15%).[3] Rarely, adrenocortical carcinoma will metastasize to pancreas, brain, diaphragm, small intestine, and thyroid.[3]

Adrenocortical carcinoma is rare, with an incidence of 1-2 cases per million; however, it accounts for up to 2% of cancer deaths, and survival figures have not significantly improved since 1978.[3] There is a bimodal age distribution with peak incidence in first and fifth decades.[3] Functional tumors are probably more common than non-functional, and cortisol hypersecretion is most common.[3] Androgen hypersecretion is more common in children than adults.[3] Estrogen and isolated mineralcorticoid hypersecretion are very rare.[3]

Clinical presentation is variable with the most common presentations being abdominal pain and weight loss.[1] A palpable mass indicates advances disease.[3] Other symptoms may include hypertension, weakness, anorexia, nausea, vomiting, severe flatulence, myalgias, and fevers.[3] Fevers may be due to tumor necrosis, hemorrhage or opportunistic infection.[3] Additionally, hypercalcemia of malignancy may be a presentation, as well as metastatic spread to other locations.[1] Virilizing features are more common in children, and cushinoid features are more common in adults.[9] Importantly, the patient in this case presented only with vague flank pain and mild weight loss with no endocrine abnormalities, despite the extensive tumor size.

Adrenocortical carcinoma requires consideration in a patient who presents with a suprarenal mass regardless of the clinical presentation. Even though adrenocortical carcinoma can present large in size, it may not exhibit extension or invasion, and can be a stage II tumor even up to 1500 grams as reported in this case. Prognosis is related to stage of the tumor, and the only factor that improves survival is surgical resection.[2] □

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