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Metastatic Mucinous Adenocarcinoma and Carcinoid Tumor Arising From a Mature Cystic Teratoma of a Horseshoe Kidney

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ABSTRACT

Concurrent primary carcinoid tumor and primary mucinous adenocarcinoma arising within a mature cystic teratoma of a horseshoe kidney is a rare event and has been reported once in the literature. We present the first case where this tumor subtype has metastasized to lymph nodes. Treatment included open partial nephrectomy, lymph node dissection, and adjuvant chemotherapy with temozolomide and capecitabine. Due to the rare combination of tumor histologies, it is unlikely that this cohort will ever be able to be adequately studied to determine a standard of care. Thus extrapolation of treatment methods of similar tumors will continue to guide treatment.

Introduction

Mature cystic teratomas are typically benign tumors infrequently seen in the kidney. They can transform into malignant components, most commonly squamous cell carcinoma, followed by carcinoid and adenocarcinoma at near equal frequencies (0.17%–2%).[1] Horseshoe kidneys have an increased relative risk of developing renal malignancies. The presence of renal carcinoid tumor arising within a mature cystic teratoma has been reported in only 10 cases, of which five involved a horseshoe kidney (Table 1).[1–3] Only twice in the literature have there been reports of a third cancer—adenocarcinoma or clear cell renal cell carcinoma—presenting concurrently in a horseshoe kidney with mature cystic teratoma and carcinoid tumor.[1,3] All patients presented with organ-confined disease except for one patient who had liver metastasis at time of diagnosis.[1,2] This case-report represents the first documented instance of metastatic mucinous adenocarcinoma with signet ring features arising from a cystic mature teratoma in a horseshoe kidney with simultaneous low grade carcinoid tumor and the second to report a mucinous adenocarcinoma in a horseshoe kidney.

Case presentation

A 66 year old man presented with constant, dull, epigastric pain exacerbated by fatty food; diarrhea; and weight loss for twelve months prior to urologic evaluation. Two negative colonoscopies were completed 2.5 and 5 years prior to presentation. His past medical and surgical histories were significant for appendectomy and cholecystectomy. Social history was significant for a 40 pack-year smoking history and military service without chemical/dye exposures. No family history of genitourinary cancers. Physical exam revealed a large, palpable abdominal mass. Complete blood count, liver function, basic metabolic panel, and urinalysis were unremarkable. Computed tomography of abdomen and showed a 12 cm Bosniak IV renal cyst involving the right aspect and isthmus of a horseshoe kidney without collecting system involvement (Fig. 1). There was retroperitoneal lymphadenopathy present. Due to the highly malignant nature of solid renal masses >4 cm, the patient underwent a right partial nephrectomy and lymph node dissection without preoperative biopsy. The mass was enucleated without complication, and the patient was discharged home three days after an uneventful recovery.

Immunohistochemical evaluation of the mass demonstrated a mixed mucinous adenocarcinoma with signet ring feature and low grade neuroendocrine neoplasm (carcinoid) most likely arising from the cystic mature teratoma (Fig. 2). The lymph nodes were positive for mixed mucinous adenocarcinoma, presumed

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Table 1
Clinical characteristics of primary carcinoid tumor arising within a mature teratoma of the kidney.[1–3]

Author, year	Age, sex	Presenting symptoms	Horseshoe kidney association	Tumor components	Treatment	Follow-up
Kojiro et al, 1976	40, male	Epigastric pain, nausea	No	Mature Teratoma Carcinoid	Radical nephrectomy	Not available
Fetissof et al, 1984	65, male	Fever	Yes	Mature Teratoma Carcinoid	Radical nephrectomy	Not available
Lodding et al, 1997	23, male	Abdominal pain	Yes	Mature Teratoma Carcinoid	Radical nephrectomy	Alive with NED at 120 months
Yoo et al, 2002	30, female	Abdominal pain, fever	No	Mature Teratoma Carcinoid	Radical nephrectomy	Alive with NED at 3 months
McVey et al, 2002	39, male	Pruritus, weight loss	Yes	Mature Teratoma Carcinoid	Partial nephrectomy, alcohol injection of liver metastasis	Alive with disease, liver and spine metastasis at 72 months
Kim et al, 2004	39, female	Asymptomatic (incidental renal mass)	No	Mature Teratoma Carcinoid	Radical nephrectomy	Alive with NED at 6 months
Kurzer et al, 2005	58, female	Asymptomatic (incidental renal mass)	No	Mature Teratoma Carcinoid	Partial nephrectomy	Not available
Armah et al, 2007	35, female	Right flank pain, right costovertebral angle tenderness	No	Mature Teratoma Carcinoid	Partial nephrectomy	Alive with NED at 6 months after diagnosis
Armah et al, 2009	50, female	Low back and right hip pain	Yes	Mature Teratoma Carcinoid Adenocarcinoma	Partial nephrectomy	Alive with NED at 6 months
Sun et al, 2013	37, male	Asymptomatic (incidental renal mass)	Yes	Mature Teratoma Carcinoid Clear cell renal cell carcinoma	Partial nephrectomy	Alive with NED at 9 months
Current case	66, male	Lower abdominal pain, weight loss	Yes	Mature Teratoma Carcinoid Adenocarcinoma	Partial nephrectomy, adjuvant chemotherapy	Deceased 6 months postoperatively from complications of metastatic disease

Abbreviation: NED—No evidence of disease.

metastatic from the teratoma primary. The histologic diagnosis of carcinoid raised the concern for an occult gastrointestinal primary, and a carcinoembryonic antigen (CEA) level was obtained showing 13.2 ng/mL (normal <5 ng/mL). The patient was scheduled for a colonoscopy to search for an occult primary, but the procedure was canceled due to exacerbations of the patient's condition. However, the index of suspicion for a gastrointestinal primary was low due to his normal colonoscopy 2.5 years prior. Computed tomography of chest, abdomen and pelvis three months postoperatively showed the development of a hypodense para-aortic mass as well as new lung nodules concerning for progression of disease (Fig. 1). No biopsies were performed. After discussion of his case at a multidisciplinary tumor board, he initiated adjuvant chemotherapy with temozolomide and capecitabine for palliation. After two months of chemotherapy—seven months after initial presentation—he succumbed to cardiopulmonary arrest secondary to metastatic disease.

Discussion

Malignant transformation of mature cystic teratoma giving rise to carcinoid and mucinous adenocarcinoma in a horseshoe kidney is a rare event.[1] While lesions such as carcinoid are associated with a favorable clinical course, others like the one depicted in our case represent a more aggressive phenotype. Treatment of renal mucinous adenocarcinoma consists of nephrectomy with or without adjuvant chemotherapy. The chemotherapy regimen used in this case was based on a 2011 study by Strosberg et al which looked at first line chemotherapy for metastatic pancreatic endocrine carcinomas. Presence of mucinous adenocarcinoma with signet ring features prompted the use of capecitabine, whereas the neuroendocrine (carcinoid) component resulted in the addition of temozolomide. As shown in Stosberg's study, this regimen has demonstrated good clinical results with a median progression-free survival of 18 months.[4]

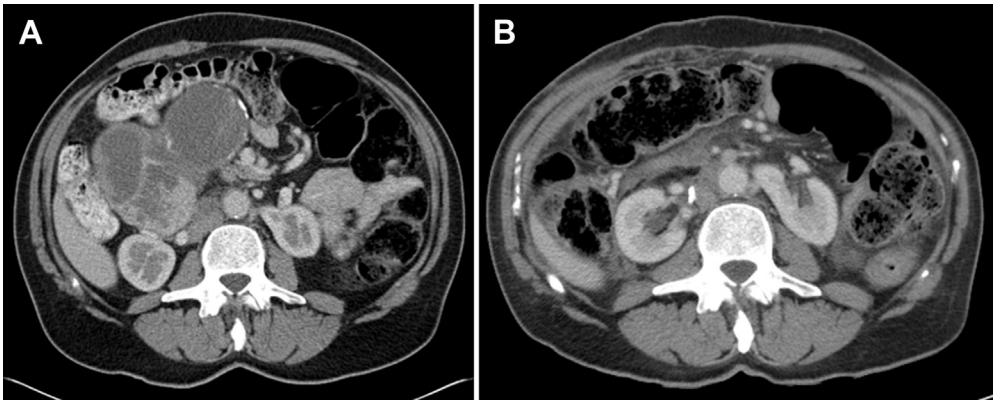

Figure 1. Preoperative (A) and three-month postoperative (B) comparison computed tomographic imaging. A new, approximately three centimeter hypodense para-aortic mass and new lung nodules (not shown) suggest locoregional and metastatic progression of disease.

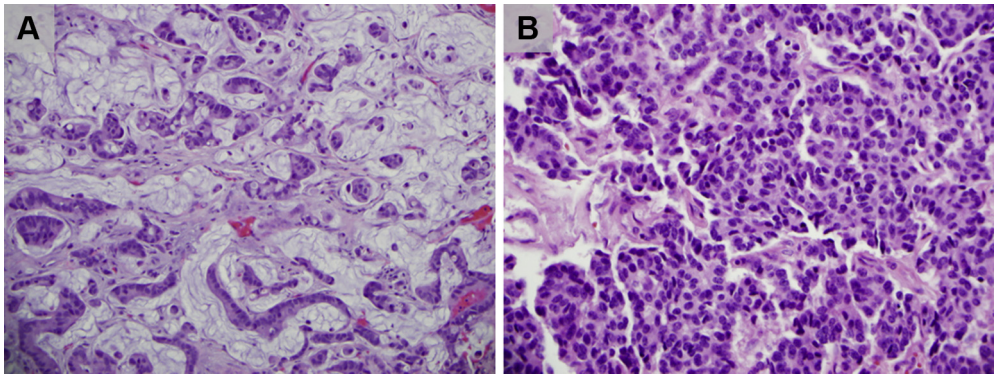

Figure 2. (A) Mucinous adenoarcinoma, H&E section, (B) Typical carcinoid tumor, H&E section.

The potential role of CEA as a biomarker of adenocarcinoma disease burden and therapeutic response in this case is indeterminate as only a single postoperative value was obtained. While its role has only been described in urological malignancies in a limited capacity, at least two cases of mucinous adenocarcinoma of the renal pelvis have shown elevated CEA levels.[5] In the first case, CEA levels were 103.94 ng/mL perioperatively and 151.83 ng/mL three months prior to her death from metastatic disease. In the second case, preoperative CEA level was 17.9 ng/mL and postoperative levels were 7.12 ng/mL at five days, 0.85 ng/mL at one month, and 0.20 ng/mL at nine months.[5] CEA's role as a potential marker for disease response for renal adenocarcinomas remains to be further validated. With a half-life of 3–5 days, a serum CEA level should be checked preoperatively or immediately postoperatively if adenocarcinoma is suspected.[5]

Conclusion

The unique histologic subtypes in the primary tumor and lymph node metastatic disease made management of this case very challenging. Malignant transformation of a renal teratoma is rare, particularly into two separate tumor types, and it is unlikely that there will be a significant number of horseshoe kidney tumors with this presentation to be able to better understand this cohort. Therefore, extrapolation of clinic data from similar tumor types will continue to dictate the treatment algorithm, such as the adjuvant chemotherapy used in this study being based on the therapy used for pancreatic mucinous adenocarcinoma. However, as next generation sequencing and comprehensive molecular characterization methods improve, it may become feasible to offer a more personalized treatment in the future.

Patient consent

The patient gave his permission to have his case presented and documented.

Conflict of interest statement

No author has any financial interest or conflict of interest to declare.

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