Metastatic Carcinoid Tumor Arising from Mature Kidney Teratoma

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Introduction

Primary carcinoid tumors and mature teratomas arising from the kidney are rare entities that compose only a small portion of genitourinary tumors, with carcinoid tumors of the kidney composing less than 1% of neuroendocrine tumors [3]. Rarer still is a combination of the two neoplasms, a primary carcinoid tumor arising from a mature teratoma. To date, only 7 prior cases of such a neoplasm have been published [1]. This case is even more unique as we believe this is the only case reported in the literature in which the mature teratoma has not only given rise to a primary carcinoid tumor, but that tumor also spread to lymph nodes and metastasized to the liver.

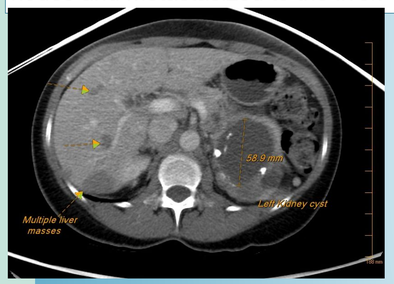
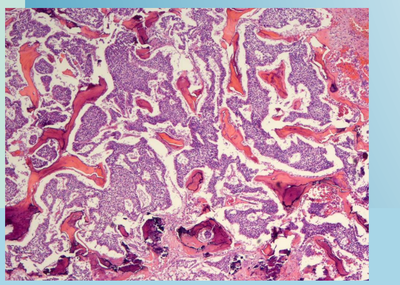
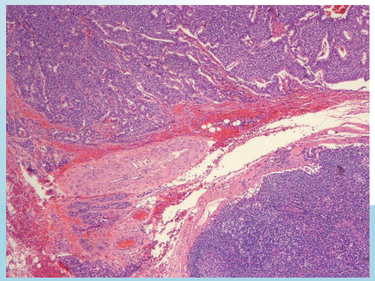


Figure 1. Initial CT Demonstrating multiple liver masses and left kidney cyst



Figure 2. Gross surgical specimen. Neuroendocrine carcinoid tumor (6.5 cm), associated with mature teratoma





Figures 3 & 4.H&E Stain of primary (left) and periaortic lymph node (right) both demonstrating similar hypercellularity and neoplastic changes

Case Report

The patient is a 62 year old African American female with a past medical history of anxiety, depression, a hysterectomy and salpingo-oophorectomy.

The patient presented with epigastric pain, bloating, and constipation. An US of the gallbladder was done which demonstrated multiple liver masses and a complex right renal cyst. A subsequent CT (Fig 1) identified numerous liver lesions and a Bosniak category 3 left complex cystic renal mass (which generally correlates to a 40-60% risk of malignancy) as well as retroperitoneal lymph node enlargement. A PET CT scan showed metabolic activity within the liver lesions, but no metabolic activity in the renal cyst or periaortic lymph nodes. She was referred to Urology for the renal mass, who advised following the patient closely with serial scans.

She underwent biopsies of the liver masses which demonstrated neuroendocrine tumor metastases of intermediate grade highly positive for synaptophysin and chromogranin, negative for CK7 and CK20, and a KI-67 of 7%. The origin of the tumor was still undetermined at this point and her urine Chromogranin A and 5-HIAA were within normal limits.

She was started on lanreotide, a somatostatin analog, per its approval for metastatic neuroendocrine tumors, and after 3 cycles underwent a repeat PET CT scan which revealed the numerous hepatic lesions with diminished radiotracer accumulation suggesting positive response to therapy and redemonstrated the stable calcified left renal cyst still without abnormal radiotracer accumulation.

Given the left periaortic lymphadenopathy and risk for malignancy of the left kidney, the patient underwent an open radical left nephrectomy with left periaortic lymph node dissection and an excisional liver biopsy. Pathology showed negative margins and was consistent with a 6.5 cm grade 2 neuroendocrine tumor (Fig. 2&3) with similar immunohistochemical characteristics to the previously biopsied liver mass. Two of the three peri-aortic lymph nodes also had metastatic foci (Fig 4) with neuroendocrine tumor marking. Specifically, positive chromogranin and synaptophysin,negative CK7 and a Ki-67 of 12%. Per pathology, the neuroendocrine tumor was arising from the mature teratoma, which also had renal parenchyma, mucinous, cuboidal, and bony elements present. The pathological TNM staging was pT1b N1 M1, correlating with Stage IV neuroendocrine carcinoma.

Discussion

This case is the first report in the literature of a primary carcinoid tumor arising from a mature teratoma of the kidney that has metastasized. There were 7 previous reports of similar neuroendocrine tumors from mature renal teratomas, but all of them remained local and were cured with definitive surgical excision[1]. The immunohistochemical characteristics of both the metastases and primary were very similar to each other, and also in line with previously described carcinoid tumors arising from mature renal teratomas[1].

There is minimal literature on very rare primary hepatic neuroendocrine tumors, but given that there were multiple liver masses, it is more consistent with metastases to the liver. While the patient previously had an ovarian cyst with elevated Ca125, on final pathology it was completely benign, so this was unlikely related to her neuroendocrine tumor. Per oncology the renal mass would likely not have responded to lanreotide given the lack of uptake on PET CT scan. It is uncertain if nephrectomy improves survival in this situation; however given the high malignant potential and unknown primary, nephrectomy was indicated.

Conclusions

The treatment for all previously described tumors of similar origin was purely surgical; however, this case is unique in that the patient presented with liver masses correlating with metastasis from this neuroendocrine tumor within this mature teratoma. Given that lanreotide was approved for advanced neuroendocrine tumors, and there were no targetable mutations, this was the first line treatment. The patient has remained stable on this treatment thus far and given that she is not a surgical candidate, this is her planned definitive treatment. The patient was generally in good condition and no discomfort was reported during 12 months of follow-up.

References

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