Case Report

Metastatic renal carcinoid: To skin, lungs, and pancreas

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ABSTRACT

Carcinoid tumors rarely originate in the urogenital system. We represent a unique case of primary renal carcinoid tumor which was initially diagnosed as renal cell cancer and 10 years later correctly diagnosed as metastatic renal carcinoid.

KEY WORDS: Pancreatic mass, renal carcinoid, renal cell cancer

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INTRODUCTION

Carcinoid cancers are relatively common in lungs. It is not unusual to have recurrence of lung carcinoid cancer and have hemoptysis. Carcinoid cancer to start in kidneys is relatively uncommon, and given historical similarity it can be confused with renal cell cancer. We represent a case of renal origin carcinoid cancer which was treated as renal cell cancer initially and later when it metastasis to lungs, real diagnosis was confirmed.

CASE REPORT

A 59-year-old male was referred to a pulmonary clinic for the evaluation of bilateral pulmonary nodules which had been progressively increasing in size over the past 2 years. He had presented earlier about 10 years ago with abdominal pain and was found to have a renal mass. He underwent right nephrectomy and was diagnosed with renal cell carcinoma (RCC), mixed papillary and granular cell type with focal sarcomatoid features. One year later, he had cutaneous lesion over right flank, and 2 years later, he had a chest wall mass. Both lesions were resected. Histological features were similar to renal biopsy of RCC. He was treated by his oncologist with sorafenib as metastatic RCC.

The pulmonary nodules were detected on surveillance computed tomography (CT) scans 8 years after his nephrectomy and were followed for 2 years [Figure 1]. Over the time, a left lower lobe (LLL) infrahilar mass doubled in size and measured 1.9 cm with mild standardized uptake value (SUV) uptake was observed [Figure 2]. An endobronchial lesion was found on bronchoscopy in the LLL which was biopsied. Pathology showed atypical cell with histological features consistent with neuroendocrine tumor. Immunohistochemistry stains were positive for synaptophysin, chromogranin, and cell adhesion molecule 5.2 [Figure 3]. It was negative for cytokeratin 7, PAX-2 (paired box gene 2), and PAX-8. There was discordance between lung biopsy specimen and previously diagnosed metastatic renal cancer. On review, renal mass showed areas of trabecular architecture more consistent with carcinoid. It had long parallel arrays that have been described in some cases of papillary RCC. All specimens were reexamined and stained. Results were more consistent with carcinoid rather than renal cell cancer. His serum chromogranin was four. Three months after the bronchoscopy, he was reevaluated for hemoptysis. His CT of the chest and abdomen showed increasing LLL and pancreatic mass [Figure 4]. Pancreatic mass also had moderate SUV uptake on preoperative surveillance positron emission tomography scan [Figure 5]. He had a left lower lobectomy and underwent partial

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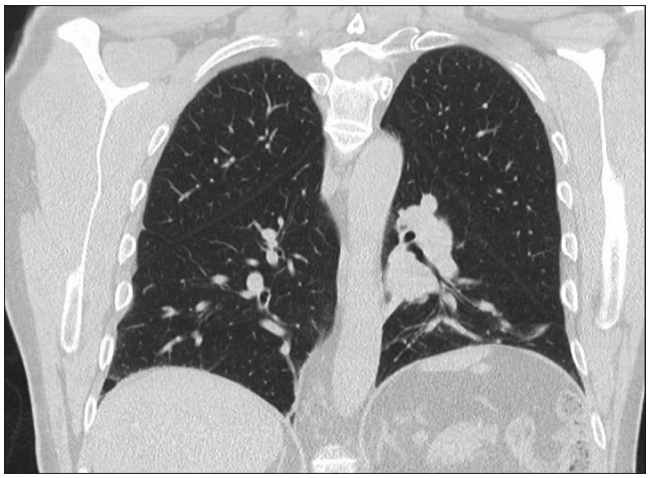

Figure 1: Coronal view of computed tomography of the chest showing a left lower lobe mass

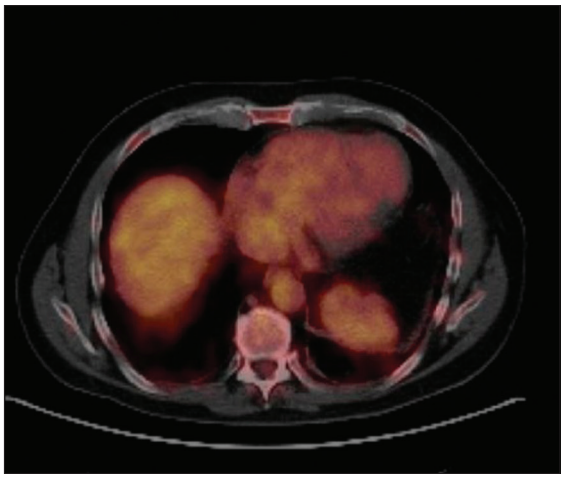

Figure 2: Positron emission tomography-computed tomography of the chest showing mild standardized uptake value uptake in left lower lobe mass

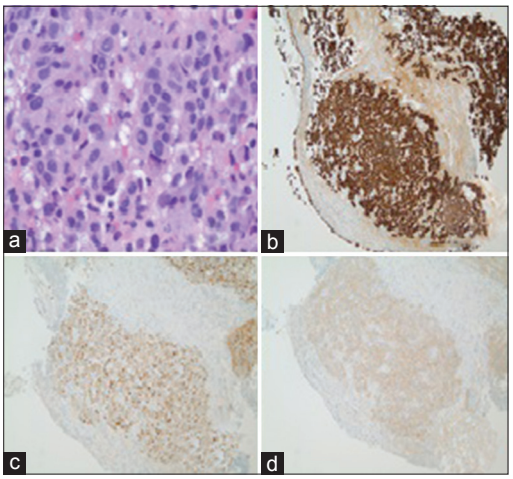

Figure 3: (a) Lung biopsy showing nests or trabeculae of medium-sized polygonal cells with lightly eosinophilic cytoplasm and small nuclei. (b-d) Tumors cells staining positive for cell adhesion molecule 5.2, chromogranin, and CD56

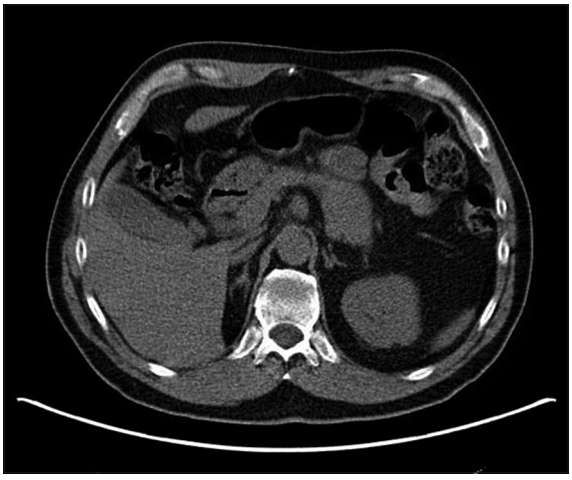

Figure 4: Computed tomography of the abdomen showing a mass in the tail of the pancreas

pancreatectomy to prevent pancreatic or bile duct obstruction. Both biopsies confirmed carcinoid metastasis with histological features similar to previous samples.

DISCUSSION

Renal carcinoid tumors (RCTs) are rare with only about 100 cases documented in literature since its first description in 1966 by Resnick *et al*.[1] This case had multiple recurrences in different parts of his body. The pathogenesis is unknown with several hypotheses supporting the notion that RCTs are derived from interspersed neuroendocrine cells associated with congenital and acquired abnormalities, i.e., horseshoe, polycystic kidney, and metaplasia of the pyelocaliceal urothelium induced by chronic inflammation.[2] The carcinoid cells have a histological appearance of uniform polygonal cells, with scant eosinophilic cytoplasm, round to elongated nuclei, and salt and pepper chromatin that are commonly arranged in trabecular pattern and can give the lesion a pseudopapillary appearance. This can lead to a misdiagnosis of papillary RCC and Wilms tumor. It has been reported in approximately 15% of cases.[3] Immunohistochemical staining is useful in confirming the diagnosis as these tumors are positive for chromogranin, synaptophysin, CD56, and other cytokeratin markers. Negative staining of PAX-2 and PAX-8 of the renal neuroendocrine cells suggests a nonnephrogenic origin and helps distinguish carcinoid from RCC.[4] Although RCT is less aggressive than RCC, 50% of cases are metastasized on initial evaluation. The largest case series consisting of 21 patients reported local and distant metastasis to liver, lymph nodes and one case of pulmonary involvement.[5] Metastasis rate is related to the tumor size, mitotic rate, and lack of confinement to the kidney.[6] They have been reported to occur in the breast, thyroid, uvea, bone, and spleen. There is, however, no documented case report of renal cell carcinoid tumor with metastasis to the pancreas and a concomitant pulmonary parenchyma and chest wall involvement.

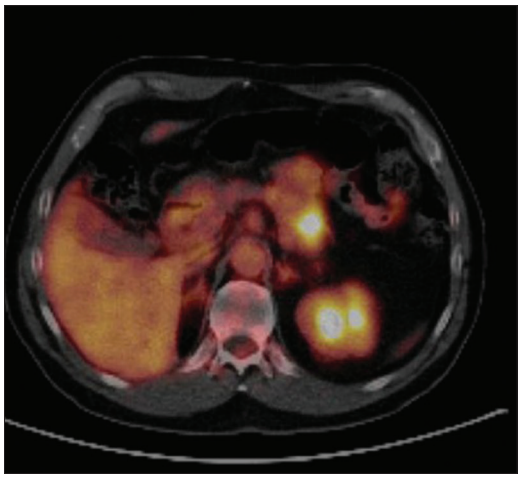

Figure 5: Positron emission tomography-computed tomography of the abdomen showing moderate standardized uptake value uptake in the pancreatic mass

CONCLUSION

Primary carcinoid tumor arising from the kidney can mimic primary papillary renal cancer histologically. Awareness of this entity and its pitfalls would improve accurate diagnosis and appropriate early intervention.

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Conflicts of interest
There are no conflicts of interest.

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