CASE REPORT
Urooncology

Doi: 10.4274/jus.galenos.2025.2025-2-14
J Urol Surg

Adrenal Heterotopia in the Middle Zone of the Kidney Resembling Renal Cell Carcinoma: A Case Report

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Abstract

A 49-year-old male was found to have two left renal masses suspicious for renal cell carcinoma during routine imaging for bilateral adrenal adenomas. The patient underwent unilateral synchronous multifocal partial nephrectomy. Pathology diagnosed a 1.7 cm upper pole lesion as a clear cell papillary tumor, while a 1.9 cm solid mass in the middle zone, was diagnosed as adrenal heterotopia. Although rare, ectopic adrenal tissue can occur in the kidney and should be considered in the differential diagnosis of renal cell cancer. Ectopic adrenal tissue should also be considered when conducting hormonal evaluation to avoid unnecessary adrenalectomy.

Keywords: Endourology, general urology, urooncology

Introduction

Adrenal heterotopia is found in less than 1% of the adult population, more commonly in men than women (1). It most commonly occurs when the adrenal cortex differentiates from the mesoderm in the fifth week of development (2). Adrenal tissue can separate along the migration path of the urogenital system, leading to ectopic adrenal tissue in the kidneys, liver, or gonads (3). Cases in the pancreas, gastric wall, and colon have also been reported, although less common (4). Because the adrenal medulla originates from the neuroectoderm, it is less likely to be near these structures during development, reducing the likelihood of resulting in adrenal ectopia (5). In this article, we present a rare case of adrenal heterotopia in the kidney coexisting with bilateral adrenal adenomas.

Case Presentation

A 49-year-old male was referred to urology due to an incidental finding of an upper pole renal mass during routine computed tomography (CT) imaging, without contrast. The CT was ordered for bilateral adrenal cortical adenomas measuring 2.8 cm and 3.8 cm. The adenomas had been under surveillance for 12 months. They have a medical history of chronic obstructive pulmonary disease, cholecystectomy, morbid obesity, smoking, and nonalcoholic fatty liver disease. Lab tests also revealed elevated hemoglobin levels, and the patient declined hormonal evaluation for the adrenal adenomas. Upon a subsequent MRI of the abdomen without and with contrast, two renal masses were identified in the left kidney. The upper pole renal mass was a 1.7 cm partially exophytic cystic lesion, classified as Bosniak 3, which remained stable in size compared to the previous year (Figure 1). The second renal mass was a 1.9 cm intraparenchymal lesion in the middle zone, hypointense on T2 weighting and demonstrating minimal enhancement on post-contrast sequences (Figure 2). Due to the concern about primary renal cell malignancy of both renal masses, the patient consented to undergo a synchronous, unilateral left robotic multifocal partial nephrectomy within 5 months without adrenalectomy. There were no surgical complications. Surgical findings included the 1.7 cm cystic lesion, and a homogeneous, tan-yellow 1.9 cm solid perihilar posterior renal mass. Pathology diagnosed the 1.7 cm upper pole lesion as a clear cell papillary renal tumor, while the 1.9 cm solid perihilar mass was diagnosed as adrenal cortical heterotopia, confirmed by positive immunohistochemical stains for calretinin and MART-1. There was no hemorrhage or necrosis in the perihilar mass. The patient reported no adverse events

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Received: 27.02.2025 **Accepted:** 30.04.2025 **Epub:** 09.05.2025

Cite this article as: Teixeira AJ, Weida C, Roy OP. Adrenal heterotopia in the middle zone of the kidney resembling renal cell carcinoma: a case report. J Urol Surg. [Epub Ahead of Print]

or complications at their two-week follow-up appointment. A six-month post-operative appointment and CT of the abdomen without and with contrast are scheduled. Patient consent was obtained for this study.

Discussion

Surgery was performed due to the risk of malignancy from both renal masses. Bosniak 3 cysts have an estimated risk of malignancy between 34% and 80% (6). The middle zone lesion was suspected of being renal cell cancer due to its location and appearance on imaging. The diagnosis of adrenal heterotopia in the middle zone of the kidney was unsuspected and serves as an important reminder to both clinicians and pathologists. Adrenal heterotopia in the kidneys is most commonly found in the upper pole due to proximity, but it should be included in the differential diagnosis of tumors in the middle zone as well (3;7). In this case, surgical intervention was recommended for the Bosniak 3 cyst and middle zone mass, but pre-operative biopsy or histological staining of the middle zone lesion may have reduced unnecessary additional excision and risk (7). Among the cases of adrenal heterotopia in the kidney, ectopic adrenal hyperplasia, carcinoma, and hormone production requiring surgical intervention are extremely rare (7). In this case, the status of the ectopic tissue was unknown; however, benign features included mild enhancement on imaging, lack of hemorrhage, necrosis, and atypical mitotic figures on histology, and a negative proliferation index.

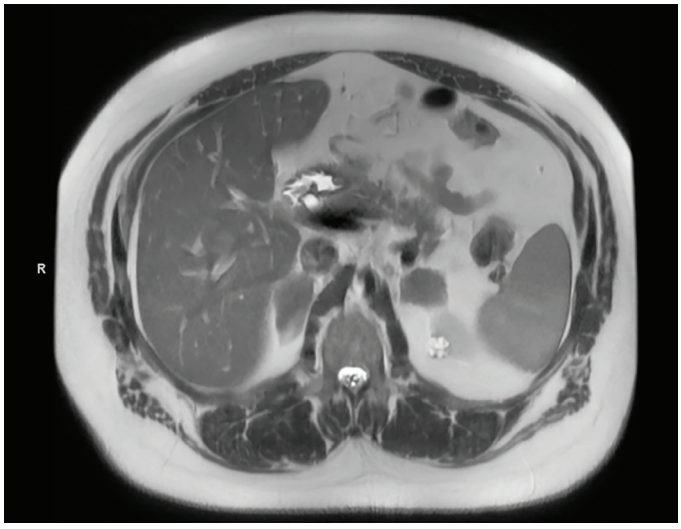

Figure 1. Left upper pole Lesion diagnosed as clear cell papillary renal cancer: MRI with and without contrast demonstrating the 1.7 cm left upper pole lesion diagnosed as clear cell papillary renal cancer

MRI: Magnetic resonance imaging

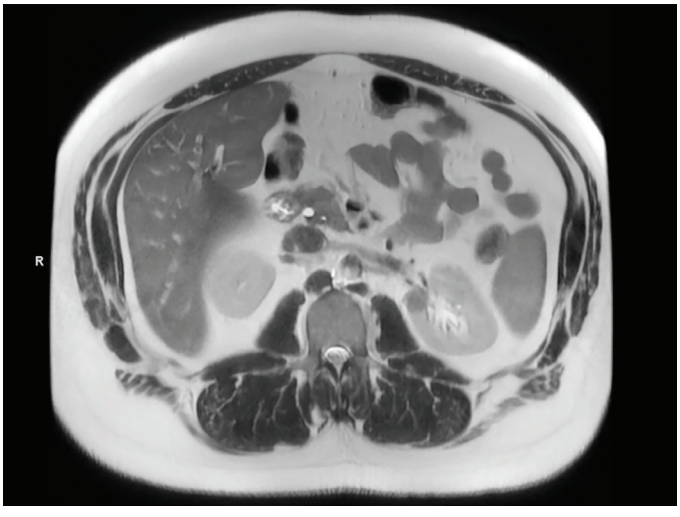

Figure 2. Midpole lesion diagnosed as adrenal heterotopia: MRI with and without contrast showing the left midpole lesion measured to be 1.9 cm. The location and appearance of the lesion was suspicious for clear cell papillary renal cancer; however, pathology diagnosed this lesion as adrenal heterotopia

MRI: Magnetic resonance imaging

The occurrence of bilateral adrenal adenomas and adrenal heterotopia is, to the best of our knowledge, not reported in the current literature. The adrenalectomy was not performed because the patient declined hormonal evaluation to determine whether the adenomas were functional. However, if hormonal testing revealed elevated levels of cortisol or aldosterone and the adrenal heterotopia was functional while the adenomas were non-functional, an unnecessary adrenalectomy may have occurred. Cases of Conn's and Cushing's syndrome, although very rare, caused by hormone-producing adrenal rests have been reported, so it is important to consider adrenal heterotopia to avoid unnecessary adrenalectomy and risk (8). This should especially be considered in cases with bilateral adenomas, which are less likely to be functional and lead to Cushing's syndrome than unilateral adenomas (9). In 2019, fewer than 40 cases caused by functional bilateral adrenal adenomas of Cushing's disease had been reported. Adrenal vein sampling may help determine the functionality of bilateral adrenal adenomas independently of coexisting ectopic adrenal tissue (9).

Conclusion

In conclusion, ectopic adrenal tissue can occur in the middle zone of the kidney and is a rare entity to consider in the differential diagnosis of renal cell cancer due to its appearance on imaging. Further, when conducting hormone testing to determine the functionality of adrenal adenomas, one may consider functional ectopic tissue, to avoid misinterpretation of hormone testing and unnecessary adrenalectomy.

Ethics

Informed Consent: Patient consent was obtained for this study.

Footnotes

Authorship Contributions

Surgical and Medical Practices: C.W., O.P.R., Concept: A.J.T., O.P.R., Design: A.J.T., O.P.R., Data Collection or Processing: A.J.T., Analysis or Interpretation: C.W., O.P.R., Literature Search: A.J.T., Writing: A.J.T., C.W., O.P.R.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study received no financial support.

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