Adrenal-Renal Fusion with Adrenal Cortical Adenoma and Ectopic Adrenal Tissue, Presenting as Suspected Renal Mass: A Case Report

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Abstract

Adrenal-renal fusion with adrenal cortical adenoma is a rare anomaly with only a few cases described in the literature. Imaging-based identification of this anomaly remains a diagnostic challenge, making it difficult to differentiate upper pole renal malignancy from adrenal cortical adenoma. We describe a case of a 62-year-old woman with an upper pole cystic renal mass on imaging, who underwent robotic partial nephrectomy. Intraoperatively the renal mass was found to be an adrenal-renal fusion anomaly, with ectopic adrenal tissue. Adrenal-renal infusion of an adrenal cortical adenoma was confirmed on final pathology. Due to lack of imaging-based diagnosis, this condition should be considered in the differential for upper pole renal masses.

Key Words

Adrenal gland neoplasm • Adrenal incidentaloma • Renal neoplasm • Adenoma

Introduction

The incidence of localized renal masses has been increasing, partly due to the increased use of abdominal imaging [1]. Outcomes regarding a small renal mass are variable, and many patients choose operative intervention [2, 3].

Although rare, adrenal-renal fusion has been described in previous case reports and can be associated with ectopic adrenal tissue [4]. There is no standard preoperative imaging that can accurately predict this diagnosis. We describe a rare case of renal cystic mass on preoperative imaging, found to be adrenal-renal fusion of an adrenal cortical adenoma with ectopic adrenal tissue.

Case Report

A 62-year-old female presented with an asymptomatic left upper pole renal mass found on workup for microscopic hematuria. Diagnostic evaluation with contrast enhanced CT abdomen revealed an enhancing 2.2 cm hypodense, cystic lesion in the left renal upper pole. An MRI was obtained for better characterization given the location and cystic nature. The MRI revealed 2.5 cm exophytic lesion in the upper pole of the left kidney, consistent with a Bosniak 3 cystic mass (fig. 1). The patient chose to undergo surgical excision.

Robotic partial nephrectomy was started in a standard manner, with dissection of the renal hilum. After renal exposure, there was no distinct renal mass. Instead there was an adrenal mass overlying the anterior aspect of the left upper pole, at the site of suspected renal mass. A separate 1 cm exophytic lesion was noted on the kidney anterior mid-pole, which was not appreciated on pre-
operative imaging. Intraoperative ultrasound was used to confirm these findings and rule out occult renal lesion. The findings were discussed with the patient’s family intraoperatively, and decision was made to proceed with adrenalectomy.

We began the adrenalectomy by selectively placing clips along the adrenal vein. During further dissection, we were unable to identify the avascular plane between the adrenal gland and upper pole of the kidney. The mass was noted to be infiltrating and invading into the upper pole of the kidney tissue. Subsequently, to ensure complete excision, we performed a left upper pole partial nephrectomy with a margin of normal renal parenchyma to ensure negative margin. The isolated anterior mid-pole left kidney lesion was removed as well. Total warm ischemia time was 22 minutes.

The final pathology reveals adrenal gland parenchyma with a nodular proliferation composed predominantly of clear cells with intermixed foci of more oxyphilic cells. The majority of cells demonstrate abundant clear cytoplasm and varying amounts of lipid vacuolization, cytologically resembling cortical fasciculate. In some areas, cells are seen in direct contact with renal parenchyma including glomeruli and tubules, indicating the lesion arose from a renal-adrenal fusion or intrarenal ectopic adrenal tissue (fig. 2). No significant atypia, increased mitotic activity, or necrosis is present to suggest a malignant etiology. Immunohistochemical stains were performed. Renal cell carcinoma is negative in tumor cells and highlights benign proximal tubular cells. S100 is negative in tumor cells and does not stain sustentacular cells. Chromogranin is non-reactive. Inhibin shows diffuse cytoplasmic reactivity in tumor cells, confirming adrenal cortical adenoma. The adrenal gland mass, renal parenchymal margin, and separate renal mass excision demonstrate adrenal cortical adenoma arising in the setting of renal-adrenal fusion with intrarenal ectopic adrenal tissue.

Discussion

Adrenal-renal fusion is a rare finding that can be classified into congenital or acquired in origin. The congenital form was originally described by Rokitansky in 1849 [5]. It has been hypothesized that the congenital form arises from failure of adrenal capsule formation by the retroperitoneal mesenchyme tissue during development, which leads to a lack of a physical barrier separating the organs [6]. Secondary changes leading to adrenal-renal fusion are post-inflammatory fibrous reactions that extend to the underlying renal parenchyma. The true incidence is unknown as most cases are described on autopsy or nephrectomy specimens.

Previous case reports of adrenal-renal fusion have been identified on imaging. In one circumstance, the relationship of adrenal-renal fusion due to an adrenal cortical adenoma was identified on CT scan [7]. In another scenario, MRI was able to provide enhanced imaging characteristics suggestive of the diagnosis [8]. Despite these reports, it is difficult to distinguish this lesion from...
potential renal or adrenal malignancies. Patel et al. [9] described this in the setting of an infiltrating mass on imaging studies, noting the adherence of adrenal tissue to the renal parenchyma through fusion gives an impression of infiltration into the tissue by a malignant lesion. In our case, the mass appeared as an exophytic, cystic enhancing lesion arising from the upper pole of the left kidney on MRI imaging. Due to this equivocal finding, renal cell carcinoma could not be excluded. Our findings enhance these previous reports, highlighting the difficulty of identifying adrenal-renal fusion preoperatively and the diagnostic dilemma therein.

There have been a small number of cases of adrenal adenoma following adrenal-renal fusion [4, 7, 8]. To our knowledge, only one other reported case has been described with the presence of both adrenal-renal fusion and ectopic adrenal tissue giving rise to a non-functional adrenocortical adenoma within the renal parenchyma [9]. Ectopic adrenal tissue is thought to be a distinct anomaly from renal-adrenal fusion; arising from fragmentation of adrenal tissue during embryologic migration and is associated with implantation in visceral tissues [4]. This tissue may become hyperplastic or neoplastic however is often an incidental finding without clinical significance. Potentially, certain genetic or embryologic factors may have led to the development of adrenal-renal fusion and ectopic adrenal tissue with our patient; however, these findings may be spurious in nature. The clinical significance is unclear but the combined benign pathology is reassuring. Although imaging characteristics could not differentiate this lesion from potential malignancy, physicians should keep adrenal adenoma with adrenal-renal fusion in the differential when a lesion of the upper pole of the kidney is found.

In conclusion, adrenal-renal fusion with adrenal cortical adenoma is a rare entity, but should remain in the differential for an upper pole renal mass with cystic enhancement. Enhanced awareness of this rare pathology may help to elucidate additional cases and identify risk factors for preoperative identification.

**Statement of Ethics**

The patient provided a consent for this case report and agreed to the publication of details and figures related to the case.

**References**

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