A case report of collecting duct carcinoma of the kidney coexistent with giant adrenal myelolipoma

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Abstract

Collecting duct carcinoma (CDC) of the kidney coexistent with giant adrenal myelolipoma is rare. We report a case of 70-year-old female who presented with a history of gross hematuria and back pain. On investigation, she had a mass in the left kidney and left adrenal gland, respectively. The patient underwent left radical nephrectomy with left adrenalectomy. Pathological findings revealed a renal CDC with partial aquamous metaplasia, in addition to an adrenal myelolipoma. Interleukin-2 was administered for 3 months after operation. Neither signs of recurrence nor metastasis has developed after a 4-year follow-up.

Introduction

Collecting duct carcinoma (CDC), also known as bellini duct carcinoma, is a rare but highly aggressive renal neoplasm arising from the distal portion of the nephron, and represents about 1% of all the renal cell carcinomas (RCCs).[1] Adrenal myelolipomas are relatively rare, non-functioning benign tumours composed of mature fatty and active hematopoietic elements. Their incidence ranges from 0.08% to 0.2%.[2] Simultaneous CDC and adrenal myelolipoma are uncommon. We present a case of CDC of the kidney coexistent with giant adrenal myelolipoma.

Case report

A 70-year-old woman presented with a 1-month history of gross hematuria and back pain. The patient did not have a history of lower urinary tract symptoms and fever. On physical examination, there were no positive findings. Urinalysis revealed a gross hematuria. Computed tomography (CT) scans of the abdomen showed a 4.5 × 5.0 cm heterogeneous mass with moderate enhancement in the upper part of left kidney and a 7.0 × 7.0 cm heterogeneous mass with fatty density inside in the left adrenal region (Fig. 1). No lymphadenopathy was noted. Intravenous pyelogram (IVP) showed the upper and middle renal calyx in left kidney did not develop and a mass was seen in the left adrenal gland area (Fig. 2). Cystoscopy revealed normal bladder mucosa with grossly bloody efflux from the left ureteral orifice. Laboratory investigations were unremarkable, with normal full-blood count, urea, electrolytes and liver function tests. Chest X-ray was negative for lung metastasis.

The patient chose to undergo open left radical nephrenctomy with left adrenalectomy. The surgery was performed with a reverse L incision on the left side of the abdomen below the rib (left subcostal incision). Her surgery and postoperative course were uneventful.

On sectioning the specimen, a 4.5 × 4.0 × 4.0 cm pale mass was mainly found in the upper part of kidney and a 6.0 × 6.0 × 6.5 greyish yellow mass in adrenal gland (Fig. 3). The tumour of kidney infiltrated the upper and middle renal calyxes. Histologic examination showed tumour cells of kidney displayed hobnail-like shape and diffuse tubular pattern with desmoplastic stromal reaction. The histological diagnosis of the tumour was left renal CDC involving the upper and middle renal calyxes and the renal pelvis. The pathological stage of the renal tumour was T1b, and the surgical margins were negative. The histological examination of the tumour in the left adrenal gland revealed the lesion consisted of lobules of mature adipocytes. The histological diagnosis of the tumour was myelolipoma (Fig.4). Immunohistochemical stains showed tumour cells diffusely positive for CK7,CK19, 34βE12 and vimentin and negative for E-cadherin, CK20, CD10 and P504S (Fig.5). The lymph nodes were negative. Interleukin-2 (IL-2) (2-Mu every other day by hypodermic injection) was administered for 3 months after operation. The patient has remained disease-free for the past 4 years.

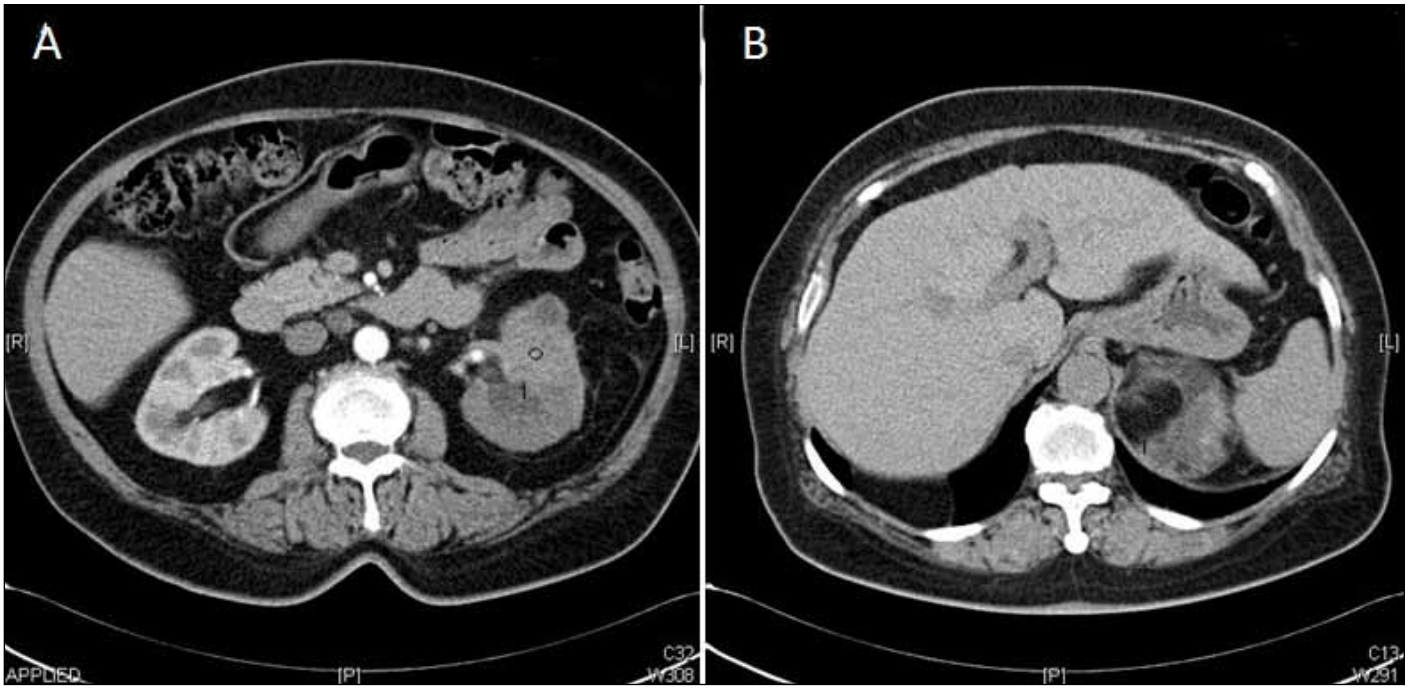

Fig. 1. Computed tomography (CT) scans revealed a mass in the kidney which presented moderate heterogeneous enhancement in enhancement CT scans (A) (oval mark) and a mass in the left adrenal gland area which presented a number of fat-density inside (B) (oval mark).

Discussion

Because about 80% to 85% of isolated renal masses represent RCCs, the detection of both adrenal and renal masses is generally considered metastatic RCC in the adrenal gland.[3]

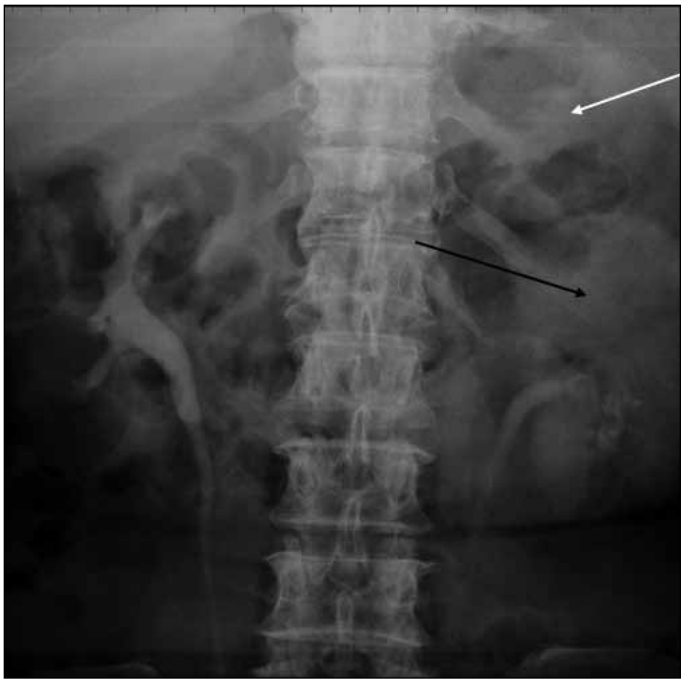

Fig. 2. Intravenous pyelogram showed the upper and middle renal calyx in left kidney did not develop (black arrow) and a mass was seen in the left adrenal gland area (white arrow).

Simultaneous renal CDC and adrenal myelolipoma at presentation are extremely rare. The histologic criterion for diagnosis of CDC, which is a rare tumour, was defined by Fleming and Lewi in 1986.[4] Because CDC is aggressive, it is easier to invade renal calyx and pelvis so that gross hematuria and back pain are more common than other subtypes of RCCs. CT scans help to differentiate the subtypes before surgery. Both the attenuation value and degree of enhancement of CDC are similar to papillary and chromophobe renal carcinomas.[5] But because of hemorrhage and necrosis, het-

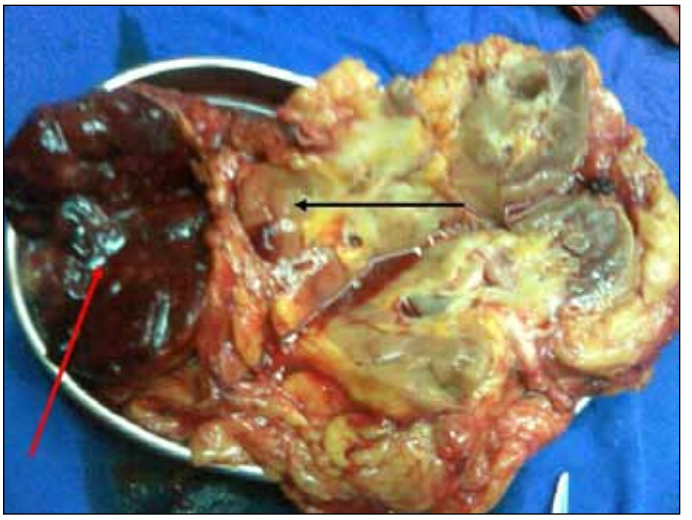

Fig. 3. The tumour of left kidney (black arrow) and the tumour of left adrenl gland (red arrow) in the specimen of left radical nephrenctomy with left adrenalectomy.

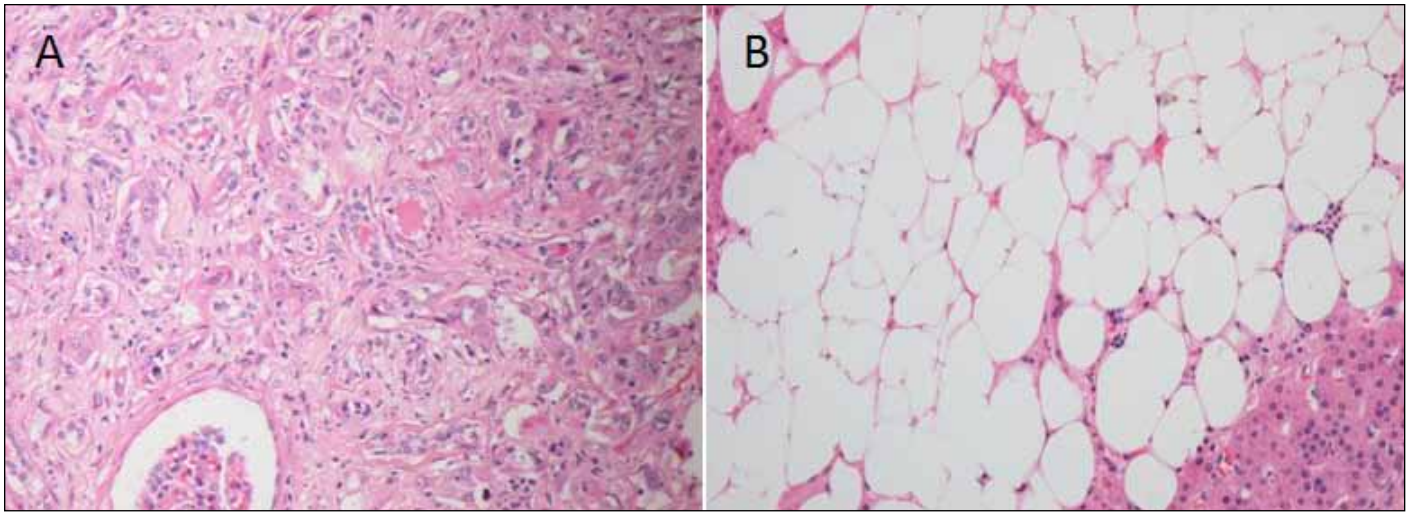

Fig. 4. Pathology of tumour in left kidney (A) (hematoxylin & eosin [H&E], original magnification ×100) and pathology of tumour in left adrenal gland (B) (H&E, original magnification ×100).

erogeneous or dominantly peripheral enhancement pattern were more common in CT scans, both of which are associated with a poor prognosis.[5] On the other hand, Medullary involvement and an infiltrative appearance are common findings on cross-sectional imaging and may suggest the diagnosis of CDC.[6] In this patient, CT scans revealed a nonhomogeneous mass which had an infiltrative growth pattern and density lower than renal parenchyma in left kidney. The lesion presented moderate heterogeneous enhancement in enhancement CT scans. The CT scan findings accorded with the description by Pickhardt.[6] The IVP appearance of CDC, similar to other RCCs, shows smooth convex filling defect or nonvisualization of involved calyxes.[6] However, the abnormality of IVP seems to occur earlier than other RCCs because of the medullary involvement and infiltrative pattern of tumour growth. Pathology is the gold standard for diagnosis of the disease. In structural terms, CDC is an aggressive epithelial malignancy with various morphologies; as such, urologists and pathologists sometimes confuse CDC with other RCCs and high-grade pelvic urothelial carcinomas, which invade the renal parenchyma.[7] As a result, immunohistochemical markers are used for differential diagnosis. Research shows that CK7 is positive in a small number of CDCs, 34βE12 and vimentin positive in some, while CKl9 positive in most. CK20, P504S, E-cadherin and CDl0 are usually negative.[8] Complete resection of the tumour, including radical nephrectomy and heminephrectomy, may lead to long-term disease-free survival. The only good prognostic factor is low pathological stage and Fuhrman grade.[9] Our patient underwent left radical nephrectomy with left adrenalectomy; at her 4-year follow-up, she was disease-free. Although immunotherapy based on IL-2 and interferon (IFN)-α does not appear to benefit all patients; it has shown efficacy in some cases.[10-11] The patient was administrated

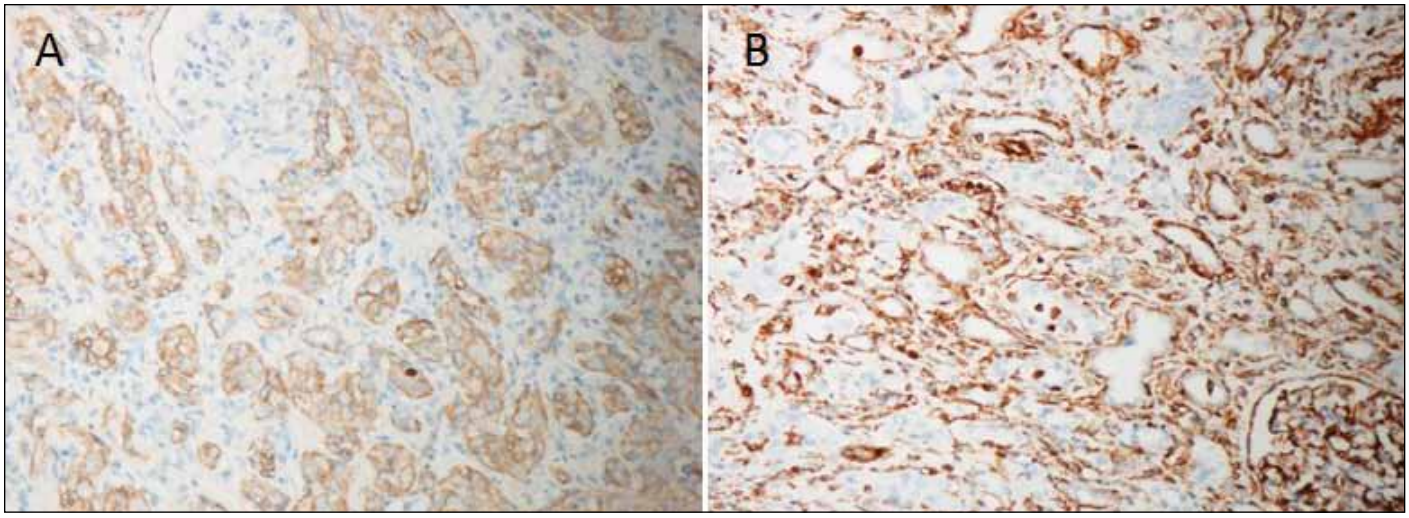

Fig. 5. Immunohistological examination of the tumour in kidney for cytokeratin (A) and immunohistological examination of the tumour in kidney for vimentin (B).

IL-2 for 3 months; this possibly improved her prognosis. Adrenal myelolipoma, which is an adrenal mass composed of lipoid and hematopoietic elements, was first described by Gierke in 1905, and later termed "myelolipoma" by Oberling. Myelolipomas account for about 8% of adrenal "incidentalomas."[12] Urologists may confuse giant myelolipoma with metastatic lesion of RCC in the case of CDC combined with giant myelolipoma, but it can be differentiated by fat-density; this supports the diagnosis of myelolipoma inside the tumour. It can be treated by adrenalectomy.

Conclusion

Simultaneous CDC and adrenal myelolipoma is rare. It could be misdiagnosed as RCC with metastatic lesion in adrenal gland. It could be successfully treated with radical nephrectomy with left adrenalectomy and immunotherapy after operation.

Competing interests: None declared.

This paper has been peer-reviewed.

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