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Case Report

Fibromyxoid nephrogenic adenoma protruding in a renal cortical cyst. A rare morphological variant in an outstanding location

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ABSTRACT

Nephrogenic adenoma (NA) is an unusual, benign lesion of the urinary tract, generally presenting in the bladder and with less frequency in the renal pelvis, urethra or ureter. It consists of tubules, microcysts and papillae lined by a single layer of low cuboidal epithelium without atypia. Recently, a fibromyxoid variant mimicking an infiltrating mucinous adenocarcinoma has been described. We report hereby the case of a 70-year-old female with a fibromixoid NA protruding in a renal cortical cyst. Only one case of NA in a renal cortical cyst has been found in the literature and it was of the classical type. The development of a NA in a renal cortical cyst lends support to the theory that the NA results from proliferation of secondarily implanted exfoliated renal epithelial cells.

1. Introduction

Nephrogenic adenoma (NA) is a rare benign lesion of the genitourinary tract that clinically presents as an incidental finding or associated to ureteral obstruction, hematuria or pain. It appears most commonly in middle-aged males and, although it can be seen anywhere in the urinary tract, it is most commonly located in the bladder (80%), urethra (15%), ureter (5%) and renal pelvis (<1%) [1,2]. Rare cases have also been reported in the mucosa of the bowel after cystectomy for urothelial carcinoma [3,5]. A case arising in a renal cortical cyst can be found in the Chinese literature [6].

Grossly, NA may appear as a nodular, papillary, sessile or pediculated polypoid lesion and several histopathological patterns can be observed singly or in combination. Classic NA consists of tubular structures lined by a single layer of cuboidal cells without cytological atypia or mitotic figures. Cystic dilatation of some tubules and degenerative-type atypia can be occasionally seen. The papillary variant consists of papillary projections covered by a single layer of cuboidal cells. Microcystic and vascular-like structures lined by compressed spindle cells resembling small vessels can be sometimes found [1–4,7,8]. Fibromyxoid NA is a newly recognized variant, first described by Hansel et al. in 2007 [9]. It is formed by epithelial spindle cells arranged without a defined pattern into a fibromyxoid extracellular matrix. To the best of our knowledge, only 8 cases of this variant have been reported up-to-date.

We present hereby a case of fibromyxoid NA protruding in a renal cortical cyst, an outstanding location for a rare morphological variant.

2. Clinical history

A 70-year-old woman was admitted to the hospital presenting intermittent macroscopic hematuria and left flank pain for the past year. The only noteworthy past medical history was a type II diabetes mellitus and hypercholesterolemia. There was no prior history of urinary tract infection, calculi or surgical procedures. Urine analysis showed abundant red blood cells. Urine cultures were sterile and urine cytology was negative for malignancy. Abdominal ultrasound revealed a mass measuring 35 mm in diameter, located in the lower pole of the left kidney. Two cortical cysts sited in the upper poles of both kidneys were also observed (58 mm in diameter the left and 52 mm the right). The patient underwent left laparoscopic radical nephrectomy. The postoperative course was uneventful and the patient is free of disease after a 15-month follow-up.

3. Materials and methods

Formalin-fixed and paraffin-embedded selected sections from the nephrectomy specimen were processed by routine methods. The sections were stained with H&E, alcian blue at pH 2.5, periodic acid-Schiff, Halle's colloidal iron and reticulin. Immunohistochemical study with cytokeratin 7 (CK7), PAX8, CD10, E-cadherin, racemase and vimentin was performed.

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4. Pathologic findings

Gross examination showed a yellowish-gray mass measuring 40 × 30 × 30 mm, which was located at the lower pole of the kidney, and a cyst at the upper pole that presented 2 whitish, firm polypoid excrescences bulging on its inner wall, 8 mm in diameter each, projecting into the lumen.

Histologically, the tumor of the lower pole was composed of medium-sized to large polygonal cells arranged in nests and solid sheets. They showed transparent, slightly reticulated cytoplasms, prominent cell membranes and nuclei with slightly irregular contours and small nucleoli (Fig. 1). Hale's colloidal iron stain was positive. Tumor cells were immunoreactive for CK7 and E-cadherin. CD10 and vimentin were negative. A diagnosis of chromophobe cell renal carcinoma was rendered.

The polypoid formations in the cyst consisted of a fibromyxoid matrix that contained compressed spindle cells and narrow small tubules resembling vascular structures (Figs. 2 and 3). Some tubules with opened round lumina lined by cuboidal or flattened cells could be focally found at the periphery of the lesion (Fig. 4). The cells showed oval or rounded, hyperchromatic nuclei and inconspicuous nucleoli. Mitotic figures were not seen. The cystic cavity was limited by a single layer of cuboidal bland epithelial cells (Fig. 5). Immunohistochemically, both spindle cells into the stroma and flat and cuboidal cells lining the pseudovascular and rounded tubules

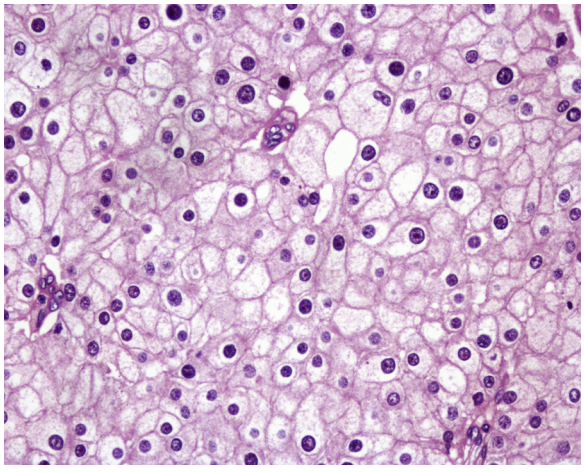

Fig. 1. High magnification view of the chromophobe renal cell carcinoma.

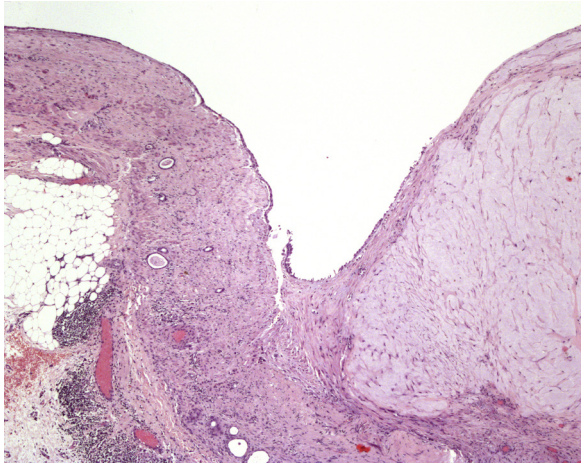

Fig. 2. Low power view showing a polypoid formation projecting into the lumen of a renal cortical cyst.

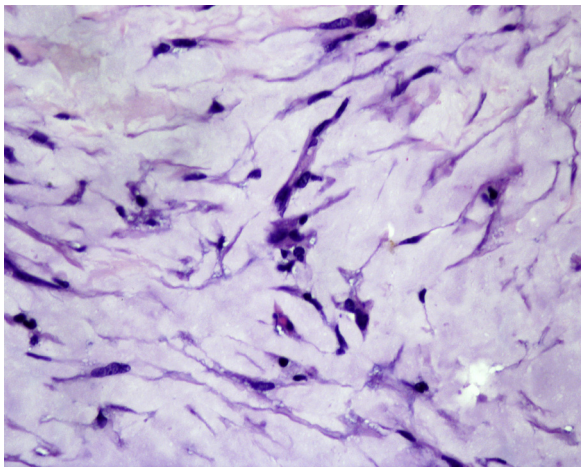

Fig. 3. Compressed spindled cells and narrow small tubules resembling vascular structures in a fibromyxoid matrix.

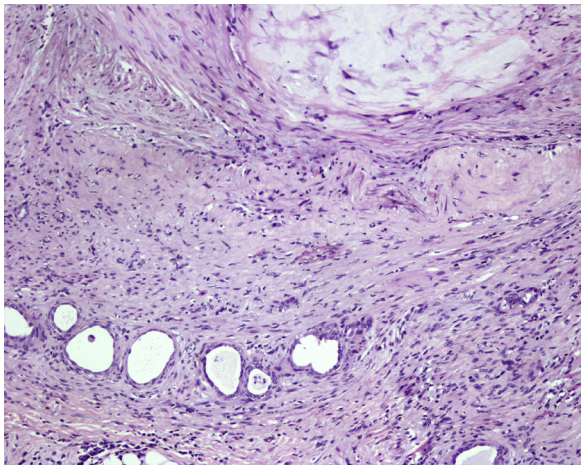

Fig. 4. A higher magnification demonstrating identifiable tubular structures typical of nephrogenic adenoma.

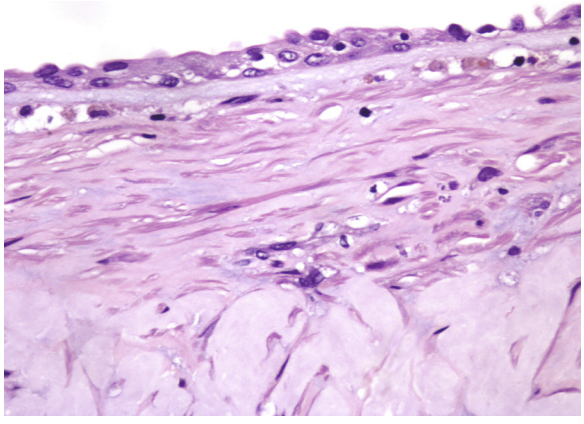

Fig. 5. The lesion presented a surface component of cuboidal bland epithelial cells.

expressed CK7 (Fig. 6), racemase (Fig. 7) and PAX8 (Fig. 8). The rest of the renal parenchyma showed nephrosclerosis.

5. Discussion

We describe a case of fibromyxoid NA protruding in a renal cortical cyst. Only 8 cases of this morphological variant have been