Mucinous Cystadenoma Arising From Renal Pelvis
A Report of 2 Cases

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Keywords: renal pelvis, mucinous cystadenoma, pelvic neoplasms

Urol J. 2008;5:197-9.
www.uj.unrc.ir

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Received July 2007
Accepted April 2008

INTRODUCTION

Epithelial tumors originating from the renal pelvis are uncommon, and adenocarcinomas account for less than 1% of malignancies which arise from renal pelvic epithelium.[1] Primary mucinous epithelial tumors occurring in the kidney are presumed to originate from the multipotential renal pelvic epithelium. Most reported cases are malignant.[2] Only isolated cases have been reported where the epithelial lining of the tumor was benign.[3] We report 2 cases of mucinous cystadenoma arising from the renal pelvis and discuss the histogenesis of this rare lesion.

CASE REPORTS

Case 1

A 35-year-old woman presented with complaints of a lump of 2 years duration in the left upper abdomen and intermittent pain in the abdomen. An irregular lump was palpable in the left hypogastrium. The clinical diagnosis was hydronephrosis. Left nephrectomy was done and the specimen showed grossly distorted kidney and pelvis. The kidney was thick walled and showed areas of hemorrhage and necrosis, and a smooth-walled unilocular cyst was seen in the pelvis (Figure 1). Copious amount of thick viscid mucinous fluid extruded out on cutting open the kidney. The cyst wall and the renal pelvis were in apposition. The calyxes were dilated. No calculi could be found either in the kidney or in the pelvis.

On histology, the cyst wall showed a single-layered tall columnar mucinous epithelium that resembled endocervical cells. There was focal pseudostratification (Figure 2). However, there were

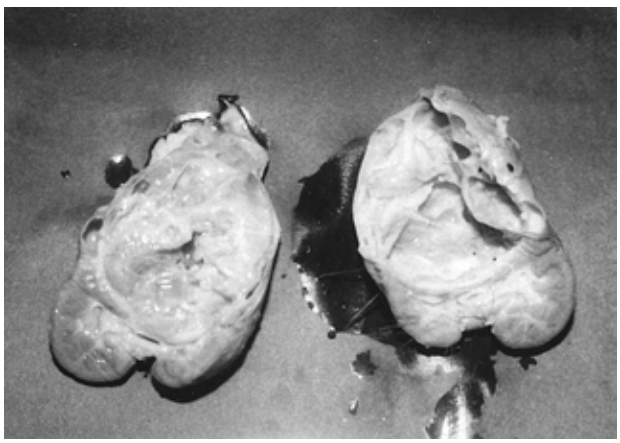

Figure 1. Unilocular cyst in the pelvic region with glistening mucoid cyst wall.

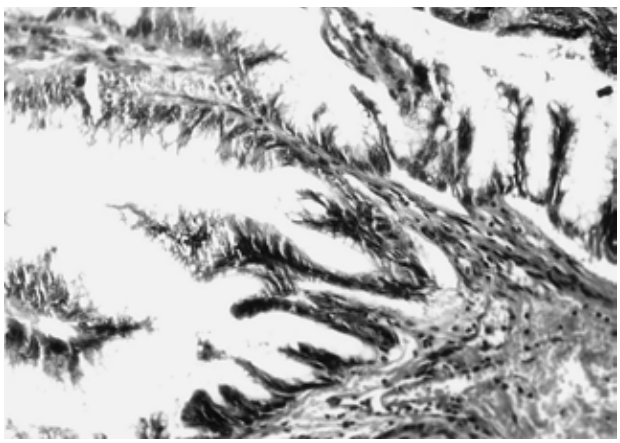

Figure 2. Papillary projections and marked stratification of the epithelium lining the cyst wall (hematoxylin-eosin, × 100).

no areas of nuclear atypia, multilayering, or stromal invasion. Sections from the adjacent areas of transitional epithelium failed to reveal any inflammatory or metaplastic changes. A diagnosis of mucinous cystadenoma arising from the renal pelvis was made. During the 2-year follow-up, the patient was well and no recurrence of the neoplasm occurred.

Case 2

A 65-year-old man presented with pain in the abdomen, weakness and loss of appetite of 8 days duration. On physical examination, a tender lump was palpable in his left lumbar region, extending up to the left iliac fossa. The lump was firm and ballottable. He was found to have prostatomegaly on rectal examination. The primary clinical diagnosis was left renal mass with prostatic hyperplasia. Ultrasonographic examination of the abdomen revealed an enlarged left kidney, which was labeled as pyonephrosis. A calculus was present in his left lower calyx, and the proximal left ureter was dilated. Peripheral smear examination revealed anemia (hemoglobin, 5.4 g/dL) and neutrophil leukocytosis with presence of toxic granules. Urine microscopy showed pyuria. Urine culture showed growth of *Escherichia coli* and *Klebsiella pneumoniae*.

The patient was operated on and a grossly dilated left kidney measuring 13 × 9 × 4 cm^3 was extracted. The renal capsule could not be stripped off. On the cut section, the pyelocaliceal system was extensively dilated and distorted. The cortex was thinned out to a fibrous rim. There were multiple smooth-walled dilated cystic areas, some of which were filled with grey opaque gelatinous clots. No distinct mass could be outlined.

On pathology examination, only occasional glomeruli were seen. The pelvic lining epithelium showed dense fibrocollagenous tissue and infiltration by lymphocytes and plasma cells. There were some foci where the lining was made of transitional epithelium, but largely, the cystically dilated areas were lined by single layers of mucin secreting cells with basally located nuclei. There was minimal architectural complexity. At places, these cells showed pseudostratification and were thrown into delicate papillary folds. The epithelium was reminiscent of the lining

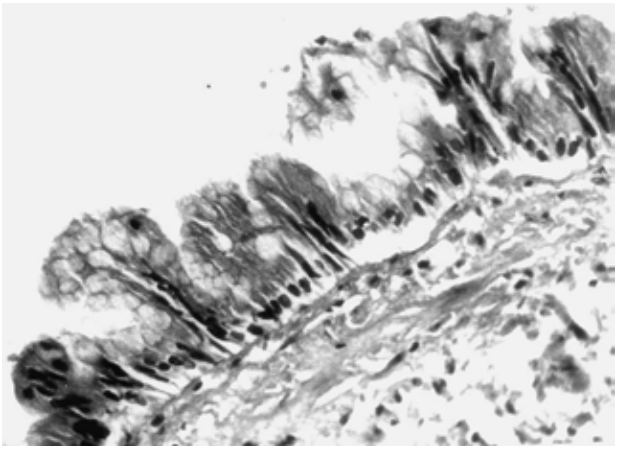

Figure 3. Pseudostratified epithelium with mucinous metaplasia (hematoxylin-eosin, × 100).

epithelium of mucinous cystadenoma of the ovary. The nuclei were bland looking, and foci of stromal invasion were absent. A transition from non-neoplastic urothelium to mucinous epithelium was demonstrable (Figure 3). A diagnosis of mucinous cystadenoma arising from the renal pelvis with changes of pyonephrosis was made. The patient did well postoperatively and was symptom free during the 8-month follow-up period.

DISCUSSION

Adenocarcinomas arising from the renal pelvic epithelium are rare, accounting for less than 1% of all epithelial malignancies found in the renal pelvis.[1] The existence of a benign counterpart of mucinous adenocarcinoma is an unsettled issue.[3] Auferderheide and Streitz[2] reviewed 32 cases of glandular neoplasm occurring in the renal pelvis. Of the 31 cases with adequate information, only 3 showed no malignant changes. Thus, the majority of the cases were interpreted as malignant.

Ross and D'Amato[3] reported a case of papillary mucinous cystadenoma of probable renal pelvic origin in a horseshoe kidney. They suggested that the same criteria used for evaluating mucinous ovarian tumors might also be appropriate for mucinous tumors of the kidney. Similarly, the striking resemblance of this lesion to mucinous cystadenoma of the ovary suggests the possibility of a similar histogenesis, ie, either mesodermal coelomic epithelium or teratomatous in derivation.

The normal transitional epithelium is capable of metaplasia into squamous, columnar, or cuboidal form.[4] Transitional or squamous carcinomas are

more frequently seen, as squamous metaplasia is relatively more common. Protoplastic conversion of transitional epithelium to colonic type of epithelium is exceedingly rare, and by that logic, mucin producing adenocarcinoma should be exceedingly uncommon. We believe that mucinous cystadenoma, like its malignant counterpart, arises in foci of intestinal metaplasia, as we were able to demonstrate histological transition from transitional epithelium to metaplastic mucinous areas in the same lesion. Toyoda and colleagues[5] and Mardi and colleagues[6] reported a case of mucinous cystadenoma arising from the renal pelvis which showed malignant transformation. They suggested the possibility that adenoma-carcinoma sequence might exist among the glandular neoplasms arising from the renal pelvis. However, we are documenting cases of mucinous metaplasia progressing to adenoma which may progress to malignancy.

Urothelial glandular metaplasia often develops nonspecifically in response to injury.[4] Glandular metaplasia and chronic inflammation with or without obstruction frequently coexist with carcinoma, leading to speculation by many authors that intestinal metaplasia is the precursor lesion for adenocarcinoma.[4] This is supported by compelling reports of a high incidence of associated adenocarcinoma (30%) in cases of significant grossly identifiable intestinal metaplasia followed up for more than 2 years.[2]

Long-standing chronic infection and kidney calculi have been invoked as possible etiologic factors in mucinous adenocarcinoma of the pelvis.[7] It is not clear if the same hypothesis holds true for adenomas as well. While one of our patients had a long-standing infection and calculus, the other one had no such preceding lesion. However, Liwnicz and associates[8] postulated that formation of the calculi might be initiated by the abundant glycoproteins secreted by the tumor. These glycoproteins can bind to cations such as sodium, calcium, and magnesium, forming larger calculi. Thus, calculi may be the result, and not the cause of the neoplasm. In most of the previously reported cases of adenocarcinoma, however, the kidneys were chronically infected.[2,7]

It is noteworthy that the literature from India[9-11] indicates a greater incidence of adenocarcinoma of the renal pelvis than data published from other geographical areas. The predisposing factors leading to this apparent increased incidence needs to be evaluated. There have been series of adenocarcinomas that show lower frequencies of calculi.[12] Epidemiological studies in specific geographic locations like India are essential if the etiology and progression of this rare neoplasm is to be unraveled.

CONFLICT OF INTEREST

None declared.

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Case Report

High-Grade Vesicoureteral Reflux in Pfeiffer Syndrome

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Keywords: Pfeiffer syndrome, cleft lip, cleft palate, craniosynostoses, vesicoureteral reflux

Urol J. 2008;5:200-2.
www.uj.unrc.ir

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Received November 2007
Accepted March 2008

INTRODUCTION

In 1964, Pfeiffer described an acrocephalosyndactyly syndrome consisting of bicoronal craniosynostosis, midface hypoplasia, broad thumbs, broad big toes, and partial and variable soft-tissue syndactyly of the hands and feet.[1] Autosomal dominant inheritance with complete penetrance is the main characteristic despite variable expressivity related to the presence or absence of syndactyly and its degree of severity. Based on the severity of the phenotype, Cohen proposed a classification of Pfeiffer syndrome into 3 clinical subtypes.[2] We report, a case of Pfeiffer syndrome type 2 with high-grade bilateral vesicoureteral reflux (VUR), and discuss the importance of surveillance for urogenital problems in patients with this syndrome.

CASE REPORT

A 4-month-old male infant was admitted to our hospital because of fever since 3 days earlier and generalized tonic-clonic seizure. He was a product of term normal vaginal delivery. He had 3 normal siblings. There was a history of abortion at the third month of gestation in the first maternal pregnancy. The parents had a normal phenotype and were not consanguineous. The mother was 34 years and the father was 36 years old.

The child had failure to thrive with a birth weight of 3.8 kg. His present weight was 4.5 kg. Developmental delay was also noticed. On physical examination, the patient had a cloverleaf skull, cleft palate, cleft lip, flat nasal bridge, broad toes, and low-set ears. Proptosis and some degree of strabismus were also noticed (Figure 1). On cardiac examination, a grade 2/6 systolic murmur was auscultated at the pulmonary area. Mild valvular pulmonary stenosis was documented by echocardiography.

Skull radiography showed acrocephaly and the prominence of temporal bones (cloverleaf skull). Computed tomography of the skull and brain showed bicoronal craniosynostosis and enlargement of lateral ventricles suggestive of moderate hydrocephalus. Electroencephalography showed paroxysmal discharge. According to the abovementioned findings, diagnosis of Pfeiffer syndrome was made clinically.

Ultrasonography of the urogenital