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Letter to the Editor

Presentation of mucinous adenocarcinoma of renal pelvis masquerading as gross hydronephrosis: A histopathological surprise

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Dear Editor,

The majority of malignant renal pelvic tumors are epithelial in origin, of these approximately 90% are transitional cell carcinomas (TCCs). Adenocarcinomas are rare and account for <1% of renal pelvic neoplasms. A mucinous adenocarcinoma is a subset of adenocarcinoma in which cancer cells produce abundant extracellular mucin.[1] Squamous and glandular neoplasm may arise within the pelvic calyceal system through the metaplastic transformation of epithelium due to long-standing obstruction, infection, and chronic irritation.[2] Ackerman reported the first case in 1946.[3]

Reports of renal pelvic mucinous adenocarcinomas in the literature are limited to small series and isolated case reports. Because the tumor is uncommon, a preoperative diagnosis is rarely made.[4] The rarity of this tumor justifies reporting the case.

A 45-year-old female patient presented with a history of dull aching pain on the right side of abdomen since 2 months. She had no history of hematuria. Laboratory investigations showed serum creatinine 0.8 mg%, urine albumin: Trace with 120–140 pus cells. Routine hematological investigations were unremarkable. In transabdominal sonography, gross hydronephrosis with marked thinning of renal parenchyma,

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internal echoes, and debris within it was observed that mimicked long-standing hydronephrosis accompanied pyonephrosis.

A double phase contrast enhanced multidetector computed tomography (CT) of the abdomen was performed on 64 slice Somatom sensation machine from Siemens Company with an oral and intravenous injection of 100 ml nonionic contrast medium. The CT examination includes plain, arterial, venous, and delayed phase. The study reveals multicystic mass in the right lumbar region involving entire kidney. The lesion appears hypodense with multiple septations. Multiple foci of calcification are seen in septa and in wall of the lower aspect of cystic mass [Figure 1a]. No significant nodular or septal enhancement seen on postcontrast study [Figure 1b]. Lesion measures about 17.4 cm × 14.0 cm × 7.6 cm in size. Renal vessels appear unremarkable. No evidence of excretion of contrast is seen in the right kidney. No hydroureter was seen on ultrasonography or CT scan. The rest of abdominal organs appeared normal. Normally functioning left kidney was observed.

Radical nephrectomy was performed.

Grossly right kidney was completely replaced by lobulated multicystic mass measuring 17 cm × 13 cm × 7 cm, filled with pus and a large amount of mucin.

Histopathology revealed that tumor show a malignant tumor comprising of cells arranged in papillary fronds. Individual cells are large, columnar having fine granular basophilic/clear cytoplasm with well-depicted borders and round vesicular nuclei with mild anisonucleosis. Some cells show prominent

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nucleoli [Figure 1c]. Foci of necrosis and hemorrhage are seen. The tumor shows extensive mucin production. The adjacent parenchyma shows changes of advanced hydronephrosis. Ureter shows the absence of tumor infiltration. Perinephric fat, renal vein and adrenal gland are free of tumor infiltration. Tumor multi-centricity and invasion in Gerota's fascia is absent. All surgical margins are free from tumor.

Final pathologic diagnosis was mucinous adenocarcinoma of the renal pelvis, stage PT2NxMx.

The patient was absolutely normal up to 6 months of follow-up in terms of the radiological investigation.

Tumors of renal pelvis are uncommon neoplasm (2.6–10.7% of all renal tumors). TCC accounts for the great majority of such tumors (approximately 90%). Ten percentages are squamous cell carcinoma and 1% adenocarcinoma.[2]

The first major review of this condition was presented by Aufderheide and Streitz in 1974, with 28 cases summarized. They found that the majority of the cases occurred in patients older than middle age and were usually associated with a long history of infection, stones, and hydronephrosis.[5] These authors emphasized the criteria for malignancy so that mucinous metaplasia is not confused with an adenocarcinoma. These criteria are (1) histological evidence of architectural or cellular atypism, (2) microscopic evidence of invasion of renal pelvis wall, renal parenchyma or nodal or distant metastases, and (3) evidence of overt invasion, recurrence or metastasis. These tumors occur as a result of metaplasia of transitional epithelium of the calyces and pelvis into the glandular epithelium, which then undergoes malignant transformation.[6]

Most reported cases were identified in patients older than 60 years. There is no evidence of any sex difference in prevalence.[7] Our patient was 45-year-old female with a tumor in right kidney. However, the relationship between this ratio and different races is difficult to accurately interpret given the small number of cases. In spite of causative mechanism of carcinoma not having been proven, it is proposed that chronic infection or stone irritation is associated with the development of mucinous adenocarcinoma.[5,6] No evidence of stone or sign of chronic infection was noted in our patient.

In contrast to cases reported a long time ago, which were diagnosed by intravenous urography (identification of renal masses, calyceal/pyelic distortion or amputation, either associated with stones or not), diagnosis is currently based on ultrasonography and CT findings. Both procedures usually show the presence of complex cystic masses requiring adequate management.[8] Contrast enhanced CT scan shows multicystic mass lesion with enhancing internal septation and calcification. Commonly, entire kidney is involved by mass and does not show any contrast excretion. Contrast enhanced CT scans also demonstrates extension of mass within Gerota's fascia, perinephric region and into the ureter, nodal involvement, distal metastasis, and renal vessels involvement.

As regards treatment, the literature unanimously agrees in that surgery is the treatment of choice. Because of few reports available and the resultant lack of minimally representative series, no guidance may be provided with regard to the use of neoconjuvant or adjacent chemotherapy or radiotherapy.[8]

From the published data, the prognosis appears to be poor, with about half of the patients dying within 2 years of surgery.[9] As reported in the literature, the diagnosis was ultimately achieved through the pathological study.

Mucinous adenocarcinoma refers to an extremely rare neoplasm. Its exact etiopathogenesis has not been elucidated. Despite the fact that the condition causes long-term symptoms in most cases due to the coexistence of lithiasis/hydronephrosis, diagnosis is essentially histological and is rarely suspected before surgery.

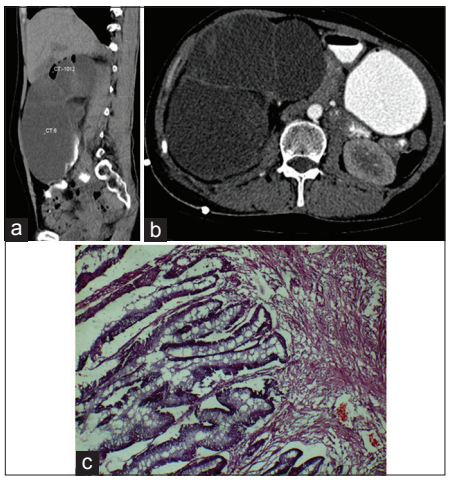

Figure 1: Computed tomography scan and histopatholgy findings in a 45-year-old female. (a) Sagittal reformatted plain computed tomography scan abdomen with oral contrast shows a multicystic mass with calcification in the lower part of the mass. (b) Axial computed tomography after contrast in arterial phase shows multicystic mass lesion involving right kidney with nonenhancing septa within it. (c) H and E stain (×20) shows mucinous glands lined by columnar epithelium with goblet cell

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Conflicts of interest

There are no conflicts of interest.

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