# Rare renal tumors – Bellini duct carcinoma

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**KEY WORDS**

renal cancer ◗ Bellini duct carcinoma

**ABSTRACT**

Bellini duct carcinoma is a rare type of renal tumor characterized by high malignancy. Race or gender-related differences in its incidence have not been demonstrated. There are, however, reports of a more frequent incidence in dialyzed patients as well as in those with renal failure and nephrolithiasis. The disease is usually diagnosed at a late stage. Radical nephrectomy is the fundamental method of management. Adjuvant therapy is usually not appropriate for Bellini duct carcinoma. Most patients fail to survive 2 years after the diagnosis has been established. We report two cases of Bellini duct carcinoma treated in our department.

## INTRODUCTION

Renal carcinoma is an epithelial malignant tumor, which accounts for 2-3% of all human cancers. The incidence increases with age, reaching a peak in the sixth decade of life. Men are affected more often than women (1.5:1).

In 2007, deaths due to renal carcinoma accounted for 2.9% of the total mortality associated with malignant tumors in Poland, whereas the incidence of new renal carcinoma cases reached 3.6% [1].

The term proposed by Murphy in 1994 – renal cell carcinoma (RCC) – has been commonly used to date. The classification of renal carcinomas based on different histological presentations has been controversial for a long time. The classification, presented in 1986 by Thoenes, distinguished the following types: clear cell carcinoma, chromaffin type carcinoma (basophilic, acidophilic, and neutrophilic), as well as chromophobic (typical, acidophilic) and Bellini duct carcinoma. In 1998, the WHO supplemented this entry with granular cell and spindle cell types as well as tumors accompanying cysts, or originating from them, were added to the classification.

Clear cell carcinoma accounts for 65-75 % of all renal carcinoma cases; chromaffin type - for 10-15%; chromophobic – for 5%; whereas the prevalence of other types reaches approximately 1%.

## CASES REPORTS

### Case 1

The patient, S.G., a female, aged 76, was referred to the urology department due to symptoms resembling right-sided renal colic along with the suspicion of a calculus within the pelvic ureteral segment. The painful symptoms presented in the right lumbar region, persisted for a few weeks without accompanying hematuria or fever, and were treated pharmacologically without any improvement. The anamnesis revealed arterial hypertension, conservative treatment of nephrolithiasis, and ischemic heart disease.

Laboratory investigations included urinalysis, which revealed 6-8 erythrocytes per visual field, and blood cell count, which indicated signs of anemia.

Ultrasound scan of the urinary tract revealed the pyelocaliceal systems (PCS) of both kidneys without any signs of calculi or urine retention. The PCS of the right kidney was markedly visible, without signs of calculi. There was, however, an irregular outline of the right kidney, the corticomedullary borderline was obscured.

Chest radiography did not reveal any pathological changes.

Urography: abdominal radiography showed that the kidneys and the iliopsoas muscles were normal. A shadow that may have corresponded to a calculus approximately 6 mm in diameter was present in the right ureter in the region of its vesical ostium. Five minutes after contrast administration the outlines of both kidneys were normal. Cortical secretion of the right kidney (nephrogram) was present without contrast enhancement of the right kidney's PCS. The pyelocaliceal systems were of normal shape bilaterally. Fifteen minutes after contrast administration, secretion of the left kidney was normal. Nephrogram of the right side revealed that the left ureter was visible along its full course and a smooth outline of the vesical walls was seen.

An attempted right-sided ureterorenoscopy failed due to kinking of the right ureter at the level of approximately 10 cm away from its vesical ostium, which could not be penetrated.

After insertion of a ureteral catheter into the right ureter, ascending pyelography was performed (Fig. 1a, b).

Ascending pyelography: abdominal radiogram showing the shadow of the ureteral catheter in the area of the right ureter. Following contrast administration the renal pelvis was partly contrast-enhanced with an irregular outline.

The patient was qualified for surgical treatment because of a suspected tumor of the right renal pelvis. Intraoperatively, a tumor-like lesion was found in the central part of the right kidney. Right-sided nephrectomy was performed. The perioperative course was non-complicated. In the postoperative period, the patient received 2 units of erythrocyte concentrate. He was discharged home in good general condition.

Histopathological investigation results obtained after the surgery:

- *carcinoma ex ductibus Bellini*
(carcinoma of the collecting ducts of Bellini) (Fig. 2a)
- *partim in statu necrosis renis dextri*
(part of the right kidney is necrotic).
*pT3aNxMx*
(malignancy grading according to Fuhrman = 3).
- *emboliae carcinomatosae vasorum*
(arterial embolization of neoplasm).
- *nephritis chronica interstitialis*
(chronic interstitial nephritis).
- *signae morphologicae haematuriae*
(hematuria).

The tumor extends beyond the fibrous renal capsule and infiltrates the perirenal adipose tissue (Fig. 2b). The lateral surgical dissection line (on the side of perirenal adipose tissue) runs across the tumor surface (Fig. 2c). The ureteral stump free of tumor infiltration.

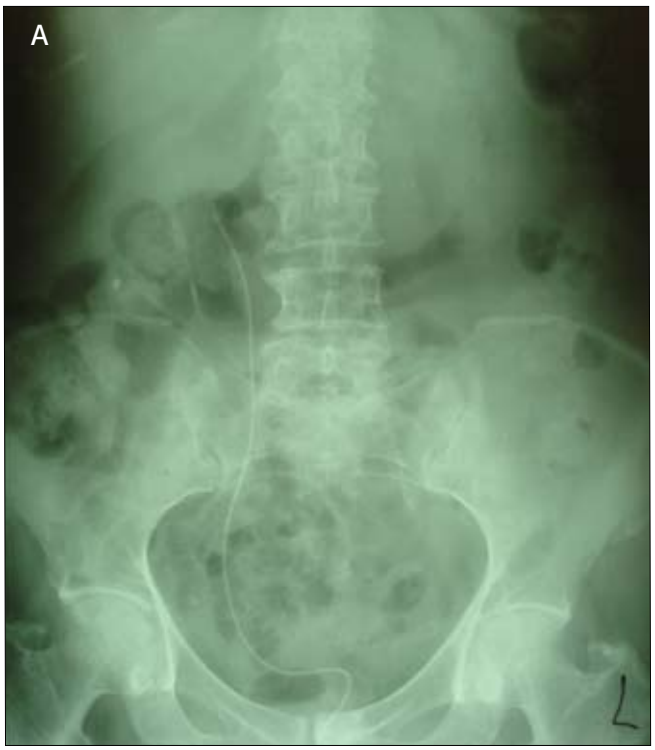


**Fig. 1a.** Plain abdominal X-ray.

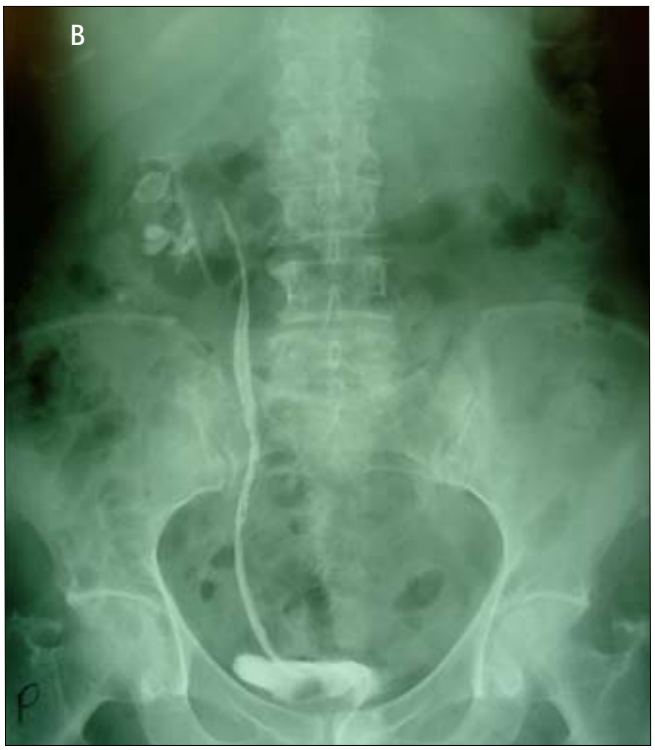


**Fig. 1b.** Ascending pygelography.

In the specimen collected from the vascular stumps, tumor growth infiltrating the walls of large blood vessels, was detected (Fig.2 d). An immunohistochemical profile confirmed the diagnosis.

Cytokeratin cocktail – positive (Fig. 2e), high-molecule cytokeratins (HMW-34be12) – positive (Fig. 2f), and vimentin – positive (Fig. 2g).

### Case 2

The patient, L.S., a male aged 48 years, was admitted to the Urology Department because a tumor was accidentally discovered in the right kidney on an ultrasound scan of the abdominal cavity. On admission, the patient reported no symptoms associated with the urinary tract. Laboratory investigations: urinalysis – no abnormalities; blood cell count - indicated minor signs of anemia.

Chest radiography: no pathologic changes.

Computed tomography of the abdominal cavity: in the central and interior part of the right kidney, a solid tumor containing cyst-like formations sized 66 x 49 mm and was connected in its lower portion with a trick-walled cyst composed of multiple chambers approximately 44 mm in diameter. Signs of infiltration to the surrounding tissues was evident. The liver was normal in size, but contained metastatic type lesions in segment 8, sized 36 x 21 mm, as well as segments 2 and 4, sized 36 and 23 mm in diameter respectively. A bone window revealed no evident metastatic lesions.

The patient was qualified for surgical treatment. Right-sided nephrectomy was performed. The peri- and postoperative course was uncomplicated. The patient was discharged home in good general condition and referred for further care to the Regional Oncology Center.

Postoperative histopathology results revealed *carcinoma solidum partim adenogenes G3* (solid carcinoma partial adenoma). The microscopic image prompted distinction between the Bellini duct carcinoma and immature urothelial carcinoma with glandular differentiation. No atypical lesions were located at the vascular or ureteral stumps. Extensive tumor infiltration of the renal parenchyma, renal pelvis, and perirenal adipose tissue was marked. Tumor cell emboli were visible in thin-walled blood vessels. A supplementary finding from numerous specimens collected from the renal pelvis region did not show atypical features of the urothelium. Immunohistochemical investigations were also performed: CK 7 (-) and Vimentin (+). The histological presentation, together with the immunophenotype, point towards a Bellini duct carcinoma rather than a urothelial carcinoma with glandular differentiation.

## DISCUSSION

Bellini duct carcinoma is a rare type of renal tumor (accounting for less than 1% of all cases). However, it is characterized by high malignancy. The First Department of Urology of the Military Medical Academy Memorial University Teaching Hospital – Central Veterans Hospital surgically treated 231 renal carcinoma patients from 2004 to 2009. Among these cases were 2 patients (0.8%) with Bellini duct carcinoma. No race or gender related differences in its incidence have been demonstrated. A more frequent occurrence of Bellini duct carcinoma has been described in dialyzed patients as well as in those with renal failure and nephrolithiasis [2, 3]. It often develops in younger age groups when compared to other malignant renal tumors and also occurs in children [4].

The tumor originates from the renal collective ducts (Bellini ducts) in the renal medulla. Microscopically, it consists of papules covered with a single layer of cube-like cells with characteristic knob-like widened portions („cobblestones") together with connective tissue elements and blood vessels forming the stroma. The cytoplasm of the epithelial cells is acidophilic and hyperchromatic nuclei are rarely present. Within the tubular structures, infiltrating the renal parenchyma, a pleomorphism of the cell nuclei is observed much more frequently, including a large number of atypical mitoses and anaplastic giant cells [5]. Macroscopically, the tumor is white to gray in color and is often accompanied with polycystic formations resulting from distension of tubule-like tumor struc-

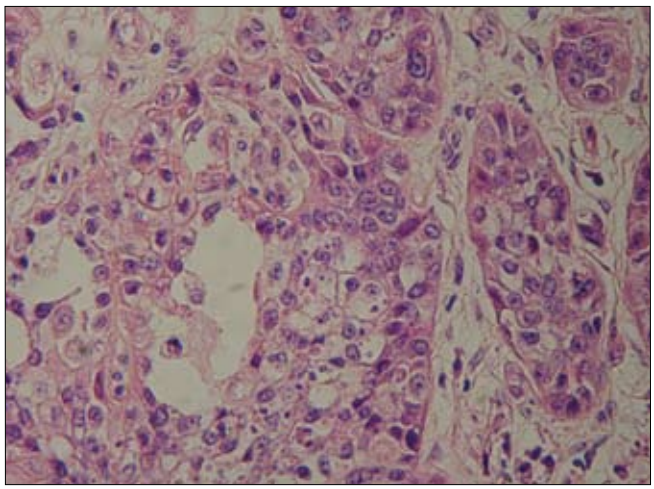

**Fig. 2a.** Carcinoma of the collecting ducts of Bellini.

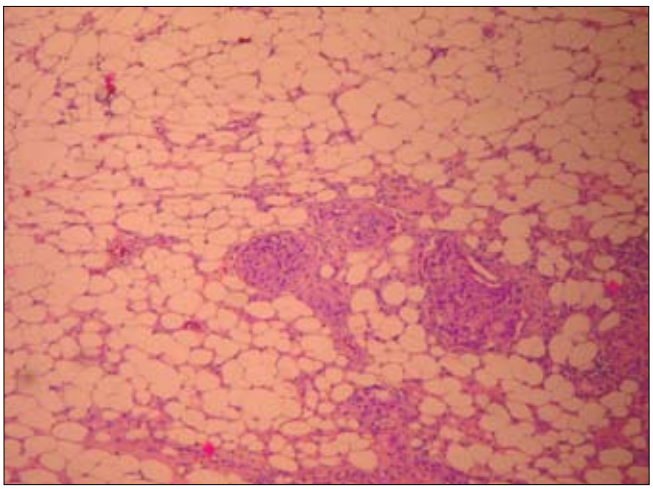

**Fig. 2b.** Infiltration the perirenal adipose tissue.

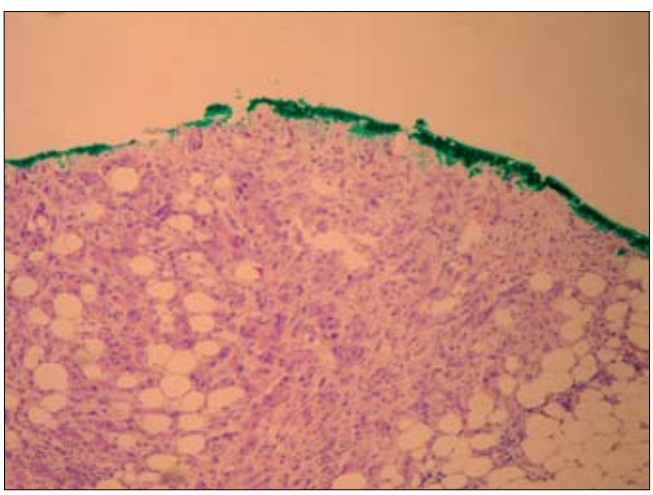

**Fig. 2c.** Lateral surgical dissection line across the tumor surface.

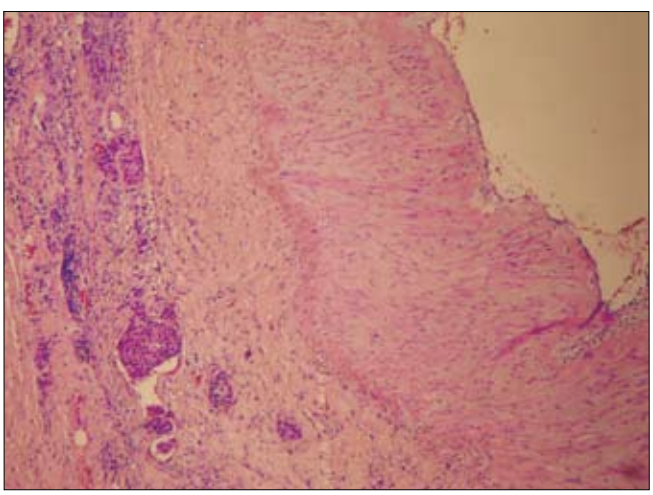

**Fig. 2d.** Inflitration the walls of large blood vessels.

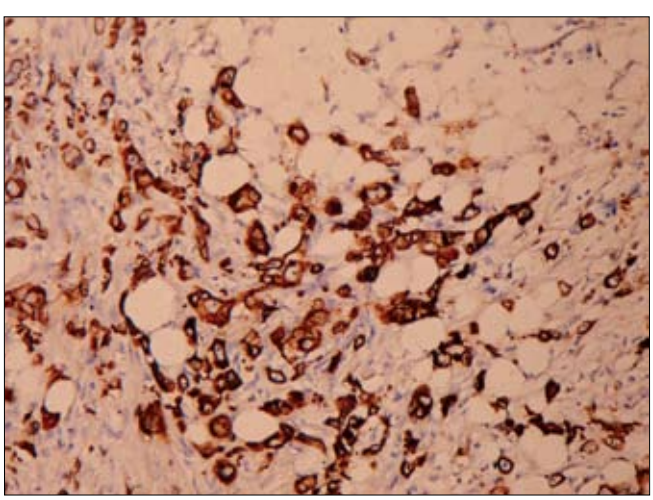

**Fig. 2e.** Cytokeratin cocktail – positive.

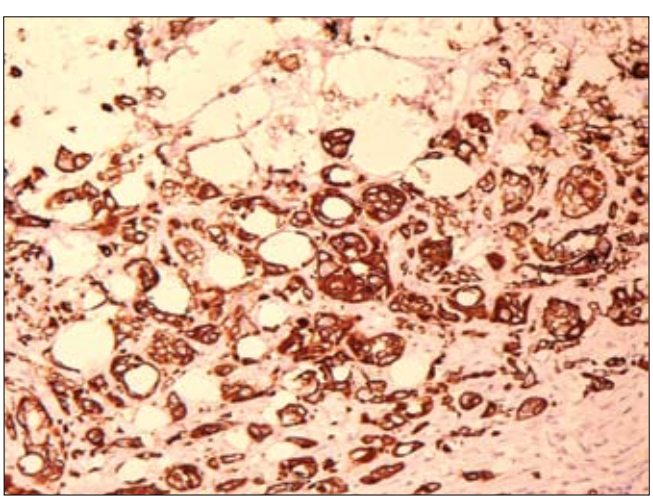

**Fig. 2f.** High-molecule cytokeratins (HMW-34be12) – positive.

tures. The tumor can infiltrate the renal cortex to obscure the corticomedullary borderline or can involve the renal hilus or vessels to cause deformation of the pyelocaliceal system. A typical feature of these tumors is the coincidence of dysplastic lesions or *carcinoma in situ* within the adjacent collective ducts.

The histopathological presentation may resemble that of a pancreatic or ovarian carcinoma metastasis. It should also be differentiated from squamous cell carcinoma of the renal pelvis, urothelial transitional cell carcinoma, and non-Hodgkin lymphoma. Immunohistochemical investigations produce positive results of staining for cytokeratin presence – cytokeratin cocktail, high-molecule cytokeratins, and vimentin [6]. Bellini duct carcinoma may be accompanied with elevation in alpha-fetoprotein (AFP) and cancer embryonal antigen (CEA) and may reveal a positive urine cytology

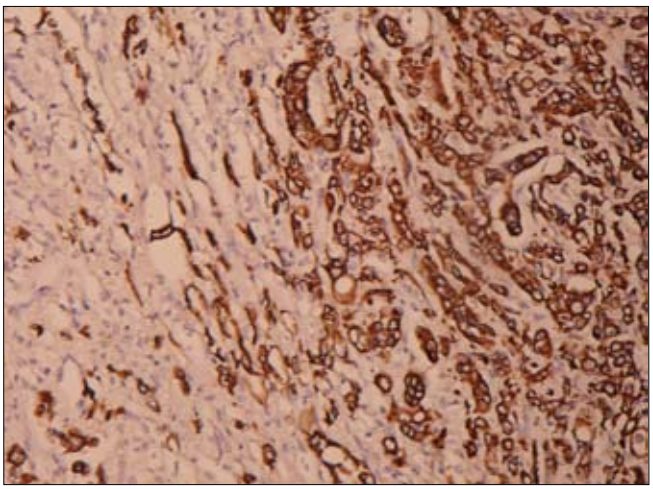

**Fig. 2g.** Vimentin – positive.

[7, 8]. Genetic abnormalities associated with this tumor are insufficiently known at the molecular level due to its rare occurrence. Bellini duct carcinoma has been associated with deletion of chromosome 1q and loss of chromosomes 1, 6, 8, 11, 18, and 21 [9].

It is usually diagnosed at a late stage with metastases present both in the lymph nodes and in the lungs, liver, bones, and adrenals (35-40% of patients) [10]. Bone metastases are usually osteoblastic in character, a quality which differentiates this tumor from e.g. chromophobic carcinoma (osteoclastic metastases). In computed tomography and angiography, Bellini duct carcinoma demonstrates signs of poor vascularization. In ultrasound scans, these tumors are hyperechogenic. In T2-weighted magnetic resonance images they are visualized as hyperintensive foci. Most patients fail to survive 2 years after the diagnosis is established. Radical nephrectomy is the fundamental method of management. Adjuvant therapies, such as radiotherapy, are not applicable for Bellini duct carcinoma. In addition to surgical treatment, chemotherapy can be instituted according to the MVAC schedule (methotrexate, vinblastin, adriblastin, and cisplatin) or gemcitabin because immunohistochemical investigations and molecular analyses suggest a similarity between transitional cell carcinoma and Bellini duct carcinoma.

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