Circumcaval Ureter With A Synchronous Ipsilateral Transitional Cell Carcinoma of the Upper Urinary Tract and the Urinary Bladder

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Uretère rétrocave avec double localisation synchrome d'un carcinome à cellules transitionnelles de la vessie et de l'urtére.

LA TUNISIE MEDICALE - 2010 ; Vol 88 (n°05) : 357 - 359

RÉSUMÉ

But : Nous rapportons un cas d'une double localisation concomitante d'un carcinome à cellules transitionnelles développé au niveau d'un uretère retro-cave associé à un carcinome urothélial vésical.

Observation : Le diagnostic a été suspecté sur les données de l'urographie intra-veineuse et l'examen tomodensitométrique qui ont montré un aspect typique de 'J' inversé de l'uretère avec une dilation en amont et d'une image d'amputation de la vessie.

Le patient a eu une cysto-prostatectomie avec une nephro-ureterctomie.

Aucune récidive n'a été constatée après un recul de 12 mois.

Mots-clés

Uretère rétrocave, carcinome, chirurgie

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SUMMARY

Aim : We report a case of concomitant ureteral transitional cell carcinoma (TCC) developed in a circumcaval ureter associated to an invasive bladder cancer.

Case : Diagnosis was made by intravenous urography (IVU) and contrast-enhanced computed tomography (CT) scanner which showed a typical 'J' shaped deformity in the dilated proximal ureteric segment with moderate right hydronephrosis and pelvic filling defect associated to bladder filling defect due to a bladder tumor. The patient underwent a radical cystoprostatectomy and nephroureterectomy; no recurrence was detected after a 12 months period of follow-up.

Key-words

Circumcaval Ureter, Carcinoma, surgery

الحالب خلف الجوف مع توضّع مزدوج متزامن لسرطانة ذات خلايا انتقالية في المثانة وفي الحالب

الباحثون : ساطع السلامي - سامي بن رحومة - منية تنقور - صابر الرباعي - كريم الشريف - نظام الدين كشير - ياسين نويرة - نوفل بن رايس - علي الحرشاني.

تستعرض دراستنا حالة توضّع مزدوج متزامن لسرطانة ذات خلايا انتقالية متكونة علي مستوى حالب خلف الجوف و متزامنة مع سرطانة في الإحليل و لمثانة .آعتمد التشخيص على المفراس و التصوير و خضع المريض إلى عمليّة آستئصال للمثانة و الموثة و إلي الكلية و الحالب .لم نسّجل أيّ تنكّس ..12 شهرا بعد العملية

حالب خلف الجوف ـ سرطانة ـ جراحة

Retrocaval ureter is a rare vascular congenital anomaly which leads to external ureteral compression by the inferior vena cava (IVC)1. Only sporadic cases were reported in the literature.
Ureteral transitional cell carcinoma (TCC) developed in a retrocaval ureter is an exceptional condition and to our knowledge only 2 such cases have been reported 2,3. The combination of TCC in a retrocaval ureter in association to bladder TCC has never been reported. We report, herein, a case of such combination which, to the best of our knowledge, is the first case reported in urological literature.

CASE REPORT

A 56-year-old man, heavy smoker, presented with a history of intermittent right frank pain and gross hematuria of one year duration.
Clinical examination was unremarkable except moderate right flank tenderness. The intravenous urogram (IVU) showed a normal left kidney while the right renoureteral unit showed a typical 'J' shaped deformity in the dilated proximal ureteric segment with moderate hydronephrosis and a filling defect in the renal pelvis (Figure 1). Abdominal computed tomography (CT) scan with contrast showed a right hydronephrosis due to an intra-pelvic tumor (Figures 2-a&b).

Figure 1 : IVU: 'J' shaped deformity in the dilated proximal right ureter with hydronephrosis (arrow) and pelvic filling defect (*)

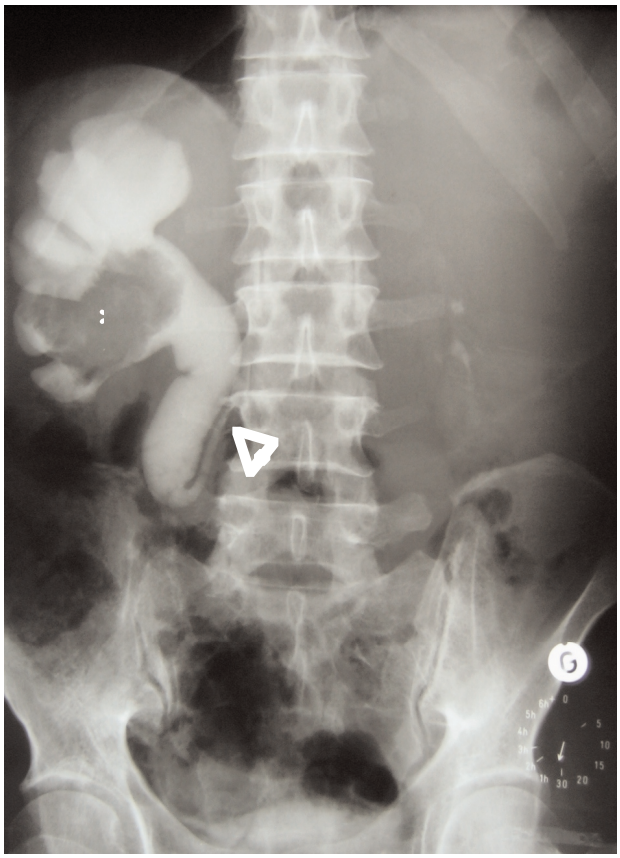
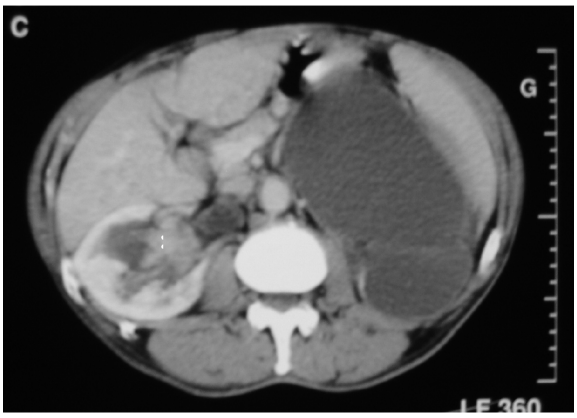
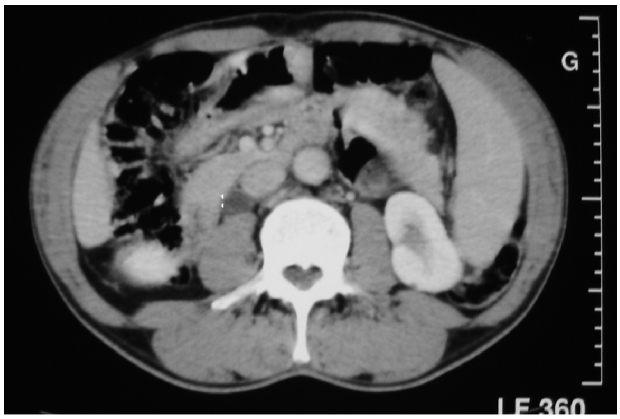


Figure 2 : Contrast-enhanced CT scan of the abdomen.
a) Axial image at the level of the right renal pelvis shows enhancing soft tissue mass occupying the dilated renal pelvis (*).
b) Axial image caudal to (a) demonstrates dilated proximal part of retrocavacal ureter (*).





Urine culture was negative. There was no evidence of bone metastasis in the radioisotope bone scan. Cystoscopy found a huge bladder tumor and biopsy revealed a grade III TCC infiltrating the superficial muscles. Surgical treatment consisted on a 'en bloc' right nephroureterectomy combined with a radical cystoprostatectomy with ileal conduit urinary diversion. Upon opening the specimen the tumor was developed either within the renal pelvis and proximal ureter, above the site where the ureter passed behind the vena cava. Definitive pathologic examinations classified the tumors as a pT4a N0 M0 and the ureteral tumor as pT2 N0 M0. With a follow-up of 12 months, radiological investigations didn't reveal any recurrence.

DISCUSSION

A retrocaval ureter is a rare congenital anomaly usually causing upper urinary tract stasis and a ''J'' or ''fish hook'' deformity of the ureter, in which the ureter passes behind the inferior vena cava (IVC). Although this anomaly is commonly known to

urologists as a circumcaval or retrocaval ureter, it is not the result of an abnormality in ureteral development, but rather an anomaly in the development of the IVC 4. The abnormal position of the ureter results from the anomalous development of the infrarenal IVC from the right posterior cardinal vein that is embryologically more laterally placed instead of from the supracardinal vein which is embryologically more medial 4. This anomaly entraps a segment of the proximal ureter as it wraps around the IVC, and often results in obstruction and hydronephrosis 5. The main causes of hydronephrosis are compression of the ureter by the psoas muscle, the pinal column, and the IVC 6; may be due also to lumen stenosis, torsion, and adhesion of the retrocaval segment 7.

Since its first description by Hochstetter in 1893 6,8, approximately 200 cases of retrocaval ureter have been reported in the reported literature 9. The incidence of this congenital anomaly ranges from 1 in 1000 live births 10 to about 1 in 1500 in autopsy studies 11. The ratio is 3 to 4:1 male to female in cadavers 12 and circumcaval ureter occurs 2.8 times more in males than in females clinically as well 6, 13.

Although the lesion is congenital, symptoms usually present at the third or the fourth decade of life 6,13. In our case, the patient was 56 years old. In most patients who become symptomatic, symptoms are due to ureteral obstruction and consequent hydronephrosis. Patients usually present with right flank pain or discomfort. This pain can be intermittent, dull, and aching 6. Rarely, a lumbar mass could be observed due to a hydronephrotic affected kidney 2. The other clinical features included recurrent urinary tract infections, microscopic or gross haematuria as in our reported case 7.

Imagings are usually sufficient to make diagnosis of retrocaval ureter. Ultrasonography is a non-invasive method to demonstrate the anatomy of the retrocaval ureter and to do follow-up of the patients for hydronephrosis, parenchymal atrophy, and nephrolithiasis 7. IVU and retrograde urography were commonly used to diagnose a circumcaval ureter. The renal pelvis and upper ureter are typically elongated and dilated in a ''J'' or fish-hook shape before they pass behind the IVC. Typically, IVU can fail to visualize the portion of ureter that extends behind the IVC; a retrograde ureteropyelogram, however, may help demonstrate the typical fish-hook curve of the upper ureter towards the midline, with the retrocaval segment at the level of the 3rd or 4th lumbar vertebra 6,13 . A spiral CT scan has been recently considered the tool of choice for the diagnosis of IVC abnormalities and circumcaval ureter 14-16 as it is non-invasive and accurately determines the anatomic relationship of the IVC and ureter 17.

Ureteral TCC developing in a retrocaval ureter is an exceptional condition. To our knowledge, only two such cases were reported in the urological literature 2,3. Some investigators have suggested that urinary stasis, often associated with this type of anomaly, may be tumorigenic 9. Urinary stasis increases the contact time of urinary metabolites with the urothelium of the dilated collecting system. Smoking, as reported in the present case, is an important predisposing factor. Synchronous TCC of the bladder and ureter was found in 2.3% of patients with bladder TCC, 39% of those with ureteral TCC, and 24% of those with renal TCC 18 as in our case. To our knowledge, no cases of synchronous upper urinary tract TCC in a circumcaval ureter and bladder tumor were reported in the literature. Our case seems to be the first report of this association.

As recommended for treatment of such cases, our patient underwent a radical cystoprostatectomy and radical nephroureterectomy. No recurrence has been observed after one year follow-up period.

CONCLUSION

An accurate preoperative diagnosis of a retrocaval ureter can be achieved by imagings. As in normal upper urinary tract, association of bladder and upper urinary tract TCC may coexist.

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