EPIDEMIOLOGY:

Upper urinary tract urothelial carcinomas (UTUCs) are uncommon and account for only 5–10% of UCs. Peak incidence in individuals aged 70–90 years. Pyelocalyceal tumours are twice as common as ureteral tumours. Multifocal tumours are found in 10–20%. Concomitant CIS: 11–36%. Concurrent bladder cancer: 17%.

2/3 have invasive disease at diagnosis compared to 15–25% with muscle-invasive bladder tumours. 9% of patients present with metastasis. Genomic characterisation of UTUC provides information regarding the risk of bladder recurrence and can identify tumours associated with Lynch syndrome. Patients identified at high risk for Lynch syndrome should undergo DNA sequencing for patient and family counselling.

HISTOLOGY AND CLASSIFICATION:

Almost always urothelial carcinomas and pure non-urothelial histology is rare. Pure squamous cell carcinoma is often assumed to be associated with chronic inflammatory diseases and infections arising from urolithiasis. Urothelial carcinoma with divergent squamous differentiation is present in approximately 15% of cases. UTUCs with variant histology are high-grade and have a worse prognosis compared with pure UC.

STAGING: TNM, 2017:

DIAGNOSIS:

- Symptoms:
  Most common symptom: visible or nonvisible haematuria (70–80%) Flank pain (clot or tumour obstruction or less often due to local growth).

- Imaging, cystoscopy, cytology and diagnostic ureteroscopy:

  Summary of evidence

  The diagnosis and staging of UTUC is best done with computed tomography urography and URS.
  Selective urinary cytology has high sensitivity in high-grade tumours, including carcinoma in situ.
  Ureteroscopy can detect concomitant bladder cancer.

  Recommendations

  strong

  1. Perform a ureteroscopy to rule out bladder tumour.
  2. Perform a computed tomography (CT) urogram for diagnosis and staging.
  3. Use diagnostic ureteroscopy and biopsy if imaging and cytology are not sufficient for the diagnosis and/or risk stratification of the tumour.
  4. Magnetic resonance urography or 18F-Fluorodeoxyglucose positron emission tomography/computed tomography may be used when CT is contra-indicated.

PROGNOSIS:

Risk stratification of non-metastatic UTUC

Low-risk UTUC*
- Unifocal disease
- Tumour size < 2 cm
- Negative for high-grade cytology
- Low-grade URS biopsy
- No invasive aspecto on CT

High-risk UTUC**
- Multifocal disease
- Tumour size ≥ 2 cm
- High-grade cytology
- High-grade URS biopsy
- Local invasion on CT
- Hydronephrosis
- Previous radical cystectomy for high-grade bladder cancer
- Variant histology

UTUC - prognostic factors included in prognostic models

Pre-operative

High-risk UTUC
- Advanced age
- Poor social status
- Chronic kidney disease
- Ureteral tumour location
- Sessile tumour architecture
- Advanced clinical stage
- High grade (biopsy, cytology)
- Blood based biomarkers

CT = computed tomography; URS = ureteroscopy; UTUC = upper urinary tract urothelial carcinoma.
* All these factors need to be present.
** Any of these factors need to be present.

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