Epidemiology, aetiology, classification and diagnosis

EPIDEMIOLOGY, AETIOLOGY AND PATHOLOGY:

Primary urethral carcinoma (PUC) is considered a rare cancer, accounting for < 1% of all genitourinary malignancies. Male/female prevalence of 2.9:1.

For male primary urethral carcinoma, various predisposing factors have been reported, including urethral strictures, chronic irritation after intermittent catheterisation/urethroplasty, external beam irradiation therapy (EBRT), radioactive seed implantation, chronic urethral inflammation/urethritis following sexually transmitted diseases and lichen sclerosus.

In female urethral carcinoma, urethral diverticula and recurrent urinary tract infections have been associated with primary urethral carcinoma.

Urothelial carcinoma (UC) is the predominant histological type (54-65%), followed by squamous cell carcinoma (SCC) (16-22%) and adenocarcinoma (AC) (10-16%). In women, AC is the more frequent histology (38-46.7%) followed by SCC (25.4-28%), UC (24.9-28%) and other histological entities (6%).

STAGING AND CLASSIFICATION SYSTEMS:

In men and women, urethral carcinoma is classified according to the 8th edition of the TNM classification. There is a separate TNM staging system for prostatic UC. For cancers occurring in the urethral diverticulum, stage T2 is not applicable.

TUMOUR GRADE:

World Health Organization (WHO) grading system of 2004

<table>
<thead>
<tr>
<th>Grade</th>
<th>Non-muscle invasive papillary carcinoma (NMIBC)</th>
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<tbody>
<tr>
<td>Low grade</td>
<td>Well differentiated (G1)</td>
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<tr>
<td>High grade</td>
<td>Poorly differentiated (G3)</td>
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PUNLMP (papillary urothelial neoplasm of low malignant potential) can rarely be seen in PUC.

DIAGNOSTIC EVALUATION:

HISTORY:

When becoming clinically apparent, most patients (45-57%) present with symptoms associated with locally advanced disease (T3/T4): extravesical urethral mass, bladder outlet obstruction, pelvic pain, urethrocutaneous fistula, abscess formation or dyspareunia.

At initial presentation visible haematuria or bloody urethral discharge is reported in up to 62% of the cases.

CLINICAL EXAMINATION:

Physical examination should comprise palpation of the external genitalia for suspicious indurations or masses and digital rectal examination. In women, further pelvic examination with careful inspection and palpation of the urethra should be performed. Bilateral inguinal palpation should be conducted to assess the presence of enlarged lymph nodes (LNs), describing location, size and mobility.

URINARY CYTOLOGY:

Cytological assessment in suspect cases of primary urethral carcinoma should be conducted according to the Paris system although its role is limited since its sensitivity ranges between 55% and 59%. In male patients, the sensitivity for UC and SCC was reported to be 80% and 50%, respectively, whereas in female patients, sensitivity was found to be 50% for UC and 77% for SCC.

BIOPSY:

Careful cystoscopic examination is necessary to exclude the presence of concomitant bladder tumours. In patients with larger lesions, transurethral resection (optionally in men under penile blood arrest using a tourniquet) can be performed for histological diagnosis.

RADIOLOGICAL IMAGING AND REGIONAL LYMPH NODES ASSESS:

Enlarged LNs in urethral carcinoma often represent metastatic disease (84%).