Radical urethrectomy with bladder preservation and continent catheterizable stoma (Yang-Monti technique)

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ABSTRACT

Malignant melanoma originating in the urethra is considered extremely rare and has a very poor prognosis. Consequently, therapeutic reviews are retrospective describing assorted treatments. We report how to perform a radical urethrectomy with bladder preservation and a continent catheterizable stoma (Yang-Monti technique). Radical urethrectomy with bladder preservation and a continent catheterizable stoma may be appropriate in selected patients with tumors that do not invade the bladder neck.

Key words: Female genital, primary urethral melanoma, radical urethrectomy, Yang-Monti technique

INTRODUCTION

Malignant melanoma of the female urethra is rare. Consequently, therapeutics reviews are retrospective describing assorted treatments. The prognosis is poor with less than 10 reported cases of survival greater than 5 years. In the published cases, treatment differs in the radicality of the surgery, from excision with or without the addition of radiation therapy to extensive surgery including cystourethrectomy, vaginectomy, vulvectomy, and lymph node dissections. In this article we explain how to perform a radical urethrectomy with bladder preservation and a catheterizable stoma (Yang-Monti Technique).

CASE REPORT

A 65-year-old female patient was referred to our hospital with a 6-month history of difficulty in voiding associated with a small mass at the urethral meatus. She denied any history of dysuria, hematuria or urethral discharge. At physical examination, she was a healthy elderly woman with a small tan-colored polypoid growth protruding from the external urethral meatus (mimicking a urethral caruncle). There was no evidence of inguinal lymphadenopathy. Abdominal and pelvic ultrasound showed no significant abnormality. Urethral carunclectomy was performed and the patient made an uneventful postoperative recovery. Histological examination of the specimen showed malignant melanoma of the urethra with positive surgical margins. Immunohistochemical staining was positive for HMB-45, S-100 and vimentine. The tumor cells were negative for cytokeratin. Urethrocystoscopy revealed the presence of residual tumor in the last 0.5 cm of the urethra with no abnormal findings in either the proximal urethra or the urinary bladder. Abdominal computed tomography of the chest and abdomen as well as bone scan demonstrated no abnormal lesion in the upper urinary tract, inguinal and pelvic lymph nodes and no evidence of metastases. Malignant melanomas may be difficult to distinguish from urethral polyp, caruncle, mucosal prolapse, chancre or more common malignant urethral tumors (all about if they are hypomelanotic to amelanotic).

Based on these findings, we performed a radical urethrectomy [Figure 1] with bladder preservation and a continent catheterizable stoma (Yang-Monti technique) to achieve 2.5 cm margins. The patient was placed in the low lithotomy position with the legs extended to allow simultaneous perineal and abdominal access. A weighted vaginal speculum was placed and no. 0 silk sutures were used to tack the labia laterally in order to improve exposure of the...
vaginal introitus. A Scott ring retractor was placed in order to provide further vaginal exposure. A Foley catheter was inserted to facilitate palpation of the urethra, bladder neck and the tumor during dissection. Dissection was initiated through a transvaginal approach to excise the urethra and distal bladder neck along with a margin of anterior vaginal wall. This was done by extending the circumscibing perimeatal incision along the anterior vaginal wall. The urethra was dissected bluntly from the surrounding tissue and the pubourethral ligaments that connect the distal urethra to the interior surface of the symphysis were divided with electrocautery. The dissection was extended under the pubic arch to the base of the bladder. Frozen sections of proximal urethra were obtained to confirm negative surgical margins.

The vagina was closed primarily with 2-0 polyglactin. The continent cutaneous diversion was then performed with the patient in supine position. A midline transperitoneal lower abdominal incision was used. We fashioned a continent cutaneous stoma using the Yang-Monti technique (transversely retubularized ileum): an ileal segment [Figure 2] 2-2.5 cm long was excised and opened longitudinally about 1 cm. from mesentery, resulting in a pedicled rectangle (2 x 6 to 7 cm.). Retubularization [Figure 3] in transverse direction using interrupted sutures (5/0 polydioxanone) results in a small caliber tube that is divided by mesentery into a short branch (stoma formation, anastomosis with umbilical tunnel) and long branch (for submucosal embedding).

Yang-Monti technique has been credited with a novel modification of the tapered intestinal segment that can be reimplanted according to the Mitrofanoff principle. Postoperatively, the cutaneous conduit remained catheterized with a 12 Fr for 1 month (there were no complications). Thereafter, the patient was instructed to perform self-intermittent catheterization and remained continent.

Final pathology revealed a small focus (3 mm) of a residual malignant melanoma with negative surgical margins. One year after the surgery, the patient is in complete clinical remission with no evidence of disease.

**DISCUSSION**

Cancers involving the female urethra and other urinary organs (excluding the bladder, kidney and renal pelvis) are rare with an estimated 780 cases (0.1% of total new cancers) in women in 2008. Primary urethral cancers were noted in a total of 540 women with an age-adjusted incidence incidence of 1.5 per million in an analysis of the SEER database from 1973 to 2002.

Squamous cell carcinoma is the most common histological type of urethral’s neoplasms, accounting for 50-70% of all cases. Transitional cell carcinoma and adenocarcinomas are the next most common cell types (10-25% each). Other rarer cell types include lymphoma, neuroendocrine carcinoma, sarcomas, paragangliomas, melanoma and metastasis. Only 23
(1.4%) patients had urethral melanomas. Within urethral diverticula, an increased incidence of adenocarcinomas seems to exist, substantiating the theory that urethral diverticula in women may arise from a glandular origin.

Although the urethra is the most frequent site of primary melanoma of the urinary tract, primary urethral melanoma is rare. It accounts for 0.2% of all melanoma and about 4% of urethral cancer; more than 160 cases have been reported in the English literature in men and women since the initial description dating more than one century ago, with the largest single institutional review being a clinical pathological analysis of 15 cases, nine in women and six in men. Age at diagnosis ranges from 32 years to 80 years (mean 63). Urethral melanoma occurs more frequently in women than in men, and is most common in women of white ethnic origin. The distal urethra is more affected (80%) than the proximal urethra. In a series of 11 women [Table 1] with urethral melanoma, overall 3-year survival was 27%. Microscopic examination of urethral melanoma is even more varied, with patterns including diffuse, nested or fascicular growths of pleomorphic cells. Furthermore, amelanotic melanoma represents 1.8-8.1% of all melanomas, resulting in greater diagnostic difficulty.

As a result; immunohistochemical markers S-100 protein and HMB-45 antigen have become an important component of the histological examination. The S-100 protein is sensitive for melanocyte differentiation but lacks specificity. In contrast, HMB-45 immunoreactivity is specific to melanoma; however, it lacks sensitivity, and 10% to 40% of S-100-positive tumors are found to be HMB-45 negative. A particular challenge is present in patients with S-100-positive, HMB-45-negative tumors. As a result, investigation continues for improved detection through such markers as tyrosinase, melan-A and KBA62. Nonetheless, S-100 and HMB-45 markers are extremely useful in the diagnosis of poorly or undifferentiated tumors and when the tumor presentation is atypical.

Of the nine women with urethral melanoma reported by Oliva and colleagues, two had local excision; two had urethrectomy; one had urethrectomy, vulvectomy, groin lymphadenectomy and chemotherapy; one had urethrectomy and vaginectomy; two had cystourethrectomy, vulvectomy and vaginectomy; and one had radiotherapy alone. Of nine women with urethral melanoma, six died 13–49 months after diagnosis, one died intraoperatively, and two were alive and well at 11 months and 23 months, respectively, after diagnosis.

DiMarco and coworkers identified 11 female patients with urethral melanoma and found that eight of the 11 had vaginal invasion at their clinical surgery. There were seven recurrences, six of which were within 1 year of surgery and five of seven patients who had partial urethrectomy developed recurrent disease in the remaining urethra. The high rate of urethral recurrences suggested that inadequate surgical margins were obtained with only a partial urethrectomy. The researchers therefore advocated total urethrectomy for early-stage urethral melanoma. Optimum surgery for women with urethral melanoma remains undefined, due to low occurrence of malignant melanoma in the urethra. No therapy has been established in the urological field. However, for early-stage disease complete urethrectomy seems more beneficial than does partial urethrectomy in reducing the risk of local failure.

In this type of surgery it is difficult to obtain negative margins and leave adequate urethral length to maintain continence, which has encouraged some authors to advocate anterior exenteration as primary therapy. In our opinion, this approach is appropriate for tumors invading the bladder neck but may not necessary if the tumor involves only the...
distal urethra. Moreover, we did not perform inguinal lymphadenectomy, because the value of inguinal lymph dissection for staging and therapy for urethral melanoma has been questionable.

Although the survival benefit of groin (with or without pelvic) lymphadenectomy is uncertain, it might have a role in staging and planning of adjuvant therapy such as radiation and immunotherapy. The role of chemotherapy in patients with urethral melanoma has not been established. Occasional long-term survivors have been noted after radiation therapy to the tumor bed or to the regional lymphatics in high-risk patients. Biotherapy for urethral melanoma has been anecdotal.

CONCLUSIONS

We recommend that radical urethrectomy with bladder preservation and a continent catheterizable stoma may be appropriate in selected patients with tumors that do not invade the bladder neck, with an attempt to obtain 2.5-cm surgical margins. Optimum surgery for women with urethral melanoma remains undefined, due to low occurrence of malignant melanoma in the urethra. No therapy has been established in the urological field.

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