Oncology

Periurethral adenocarcinoma of mesonephric origin: A case report and review of the literature

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ABSTRACT

Periurethral mesonephric adenocarcinoma is a rare tumor. To the best of our knowledge, only 13 cases have been reported in the literature to date. We report the case of a 36-year-old lady who presented with periurethral mesonephric adenocarcinoma, treated by surgery followed by adjuvant chemotherapy and pelvic radiotherapy. We demonstrate the unusual histology of mesonephric adenocarcinoma and the necessity to consider this tumor in the differential diagnosis of all unusual genito-urologic tumours. In the present literature, combination of surgery followed by chemotherapy and radiotherapy is the most suitable treatment for locally advanced periurethral mesonephric adenocarcinoma.

Introduction

Mesonephric adenocarcinoma (MA) is a rare neoplasm arises from mesonephric (Wolffian) duct remnants along the female genital tract. Most of the documented cases are of cervical origin.

Urethral, periurethral and vaginal mesonephric adenocarcinomas are even rarer. To the best of our knowledge, 77 cases have been described in literature, of which only 13 are of periurethral origin. In addition, minimal information about their behavior and optimal management is known.

Case presentation

A 36-year-old female patient presented with urinary frequency and urgency of two years duration. This was complicated by abnormal vaginal bleeding and right thigh pain just two months prior to presentation.

Patient sought medical consultation in a private clinic where physical examination and ultrasound revealed two vaginal masses that were surgically excised.

Histopathology of the masses showed a poorly differentiated adenocarcinoma with focal signet ring cell-like component. The tumor cells were positive for CK7, PAX8 and CD10 immunohistochemical stains. Besides, patchy nuclear positivity for calretinin was also evident.

On CT scan (Fig. 1) and MRI of the pelvis, a large (6.3 cm) circumferential soft tissue mass was identified in the urethra, just under the urinary bladder neck. The mass showed extension into the anterior vaginal wall and urinary bladder neck. Minimal numbers of enlarged pelvic and inguinal lymph nodes were also identified. No ascites was noted.

A single hypermetabolic left inguinal lymph node was picked up on PET scan. Whole body bone scan showed no evidence of bone metastases. Colonoscopy, endoscopy and laboratory studies were unremarkable.

The patient underwent radical cystectomy, urethrectomy, and anterior vaginectomy along with bilateral inguinal lymph nodes dissection. Ilial conduit for urine diversion was also performed.

Histologic examination revealed a high grade tumor with mixture of tubular, papillary and retiform growth patterns. Multiple foci of dense eosinophilic secretions within the tubules are also present (Fig. 2). In addition, a mesonephric remnant-like cystic area was seen in close proximity to the tumor. Tumor cells were positive for CD10, CK7, EMA, vimentin, calretinin and PAX8. They were negative for ER, CEA, P16, P63, CK20, GATA3, WT1, P53 and NapsinA immunohistochemical stains.

The final diagnosis was mesonephric adenocarcinoma, stage pT4N2.

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Vaginal wall, bladder neck, anterior and lateral soft tissue margins were microscopically invaded by the tumor. However, no urethral mucosal involvement was noted.

Given the locally advanced stage of the tumor and the positive margins; surgical treatment was followed by adjuvant chemotherapy and pelvic radiotherapy in the form of sandwich technique. In which 50.4 Gray of pelvic radiotherapy were sandwiched in between six cycles of carbotaxol.

Follow up CT scan of the patient after nine months of treatment did not show any evidence of local recurrence or distant metastasis.

Review of the literature

Methods

We conducted a review of the English-written medical literature using PubMed looking for these terms: “periurethral mesonephric adenocarcinoma”, “urethral diverticular mesonephric adenocarcinoma” and “vaginal mesonephric adenocarcinoma”. Besides, we reviewed the references of the available articles. Papers that did not have obvious histopathologic typing of the carcinoma as “mesonephric” and those that did not mention the frequency of mesonephric adenocarcinoma were excluded. Twelve articles fulfilled our criteria.

Results

About 77 cases of periurethral, urethral diverticular and vaginal mesonephric adenocarcinomas are reported in literature (Table 1). Of which, only 13 cases are of periurethral origin, which accounts for 17% of the total number of female patients with lower urogenital mesonephric adenocarcinomas.

Discussion

Mesonephric adenocarcinoma is a rare tumor. Cervical, uterine, vesical, urethral and periurethral cases have been reported.

To the best of our knowledge, this case is the fourteenth case of mesonephric adenocarcinoma of periurethral origin.

Usually the diagnosis of mesonephric adenocarcinoma is made with caution given the lack of distinctive morphologic or immunohistochemical features specific for the mesonephric duct. Therefore, adenocarcinoma of other pelvic organs should be excluded. The presence of tumor in sites of normally developing mesonephric duct along with the presence of mesonephric vestiges may be of help.

In our case, the location of the tumor, the presence of mesonephric-like cystic remnant and the absence of typical in situ or papillary urethelial component were all in keeping with mesonephric adenocarcinoma. This was also supported by the immunoprofile of tumor cells.

The cervix and other genital structures were unremarkable upon physical examination and imaging, excluding them as possible sites of origin. Furthermore, the lack of clearing or hobnailing of tumor cells along with the negativity for NapsinA, ruled out a tumor of mullarian origin.

Regarding clinical presentation, mesonephric adenocarcinoma is characterized by a wide range of ages at presentation. One vaginal mesonephric adenocarcinoma presented in a 7 month old infant.

Table 1

| Study Author Year | Total numbers of cases reviewed | Periurethral MA | Urethral Diverticular MA | Vaginal MA | Notes |
|-------------------|---------------------------------|-----------------|--------------------------|------------|-------|
| Harold G. et al., 1967 | 3 | NA | NA | 2 | Ultrastructural study of the W.Leistenschen et al, 1981 case |
| I.D. Truskett 1968 | 15 | NA | NA | 2 | |
| Hano A. Siegel 1970 | 18 | NA | NA | 19 | |
| Philip et al., 1977 | 31 | NA | 2 | NA | |
| N.Schnoy et al., 1982 | 42 | 7 | 3 | NA | |
| William W. Hinchey et al., 1983 | 3 | NA | NA | 1 | First case without DES exposure |
| E. Geisler et al., 1998 | NA | NA | 12 | NA | The rest are benign |
| YoshiyShimao et. Al 2000 | 8 | 4 | NA | 1 | |
| D.Doddamani et. Al 2002 | 109 | 2 | 9 | NA | |
| CagatayErsahin et. Al 2005 | 3 | NA | NA | 2 | |
| G.Bifulco et. Al 2007 | 3 | NA | NA | 4 | |
| Iris Mueller et al., 2016 | 6 | NA | NA | 7 | |

Fig. 1. Pelvic axial CT scan with contrast showing a large mass extending to the urinary blabber.

Fig. 2. Tumor with back-to-back round tubules and dense eosinophilic intraluminal secretions (H&E original magnification, × 400).
female. Among previous cases of urethral and periurethral mesonephric adenocarcinoma patients’ ages at presentation ranged from 35 to 72 years.

Histological features of our case resemble previous reported cases of urethral and vaginal mesonephric adenocarcinoma. However, the locally advanced circumferential urethral tumor with extension into urinary bladder and anterior vagina was unusual.

Given the rarity of mesonephric adenocarcinoma, little is known about its prognosis and the best modality of treatment. However, some studies stated a 5-year survival rate of 42% and 33% in primary urethral and vaginal carcinomas, treated with radical surgery or with adjuvant radiotherapy, respectively.

For vaginal mesonephric adenocarcinoma, adjuvant chemotherapy is recommended especially in cases with microscopic tumor remnants, as these tumours are hypothesized to be sensitive for chemotherapy.

Conclusion

We demonstrate the unusual histology of mesonephric adenocarcinoma and the necessity to consider this tumor in the differential diagnosis of all unusual genito-urologic tumours. In the present literature, combination of surgery followed by chemotherapy and radiotherapy is the most suitable treatment for locally advanced periurethral mesonephric adenocarcinoma.

Conflict of interest

All authors declare that they have no conflict of interest.

References

1. Siegel HA, Sagerman R, Berdon WE, Wigger HJ. Mesonephric adenocarcinoma of the vagina in a 7-month-old infant simulating sarcoma botryoides successful control with supervoltage radiotherapy. J Pediatr Surg. 1970;5(4):468–470.
2. Pointon RCS, Poole-Wilson DS. Primary carcinoma of the urethra. Br J Urol. 1968;40(6):682–693.
3. Herbst AL, Green TH, Ulfelder H. Primary carcinoma of the vagina: an analysis of 68 cases. Am J Obstet Gynecol. 1970;106(2):210–218.
4. Mueller I, Komettrier G, Jacobs VR, et al. Mesonephric adenocarcinoma of the vagina. Strahlenther Onkol. 2016;192(9):668–671.