

Melanoma of the female urethra

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ABSTRACT

Melanoma is a malignant tumor that can affect any area of the anatomical economy. Its appearance in the female urethra is extremely rare, with approximately 121 cases in indexed literature since 1966. The subject to be described is an 86-year-old woman who seeks assessment for intermittent macroscopic hematuria with blood clots of 3 months progression. On physical examination, there are no suspicious lesions detected on the surface of the skin. On external genital examination, it is observed a friable lesion at the level of the urethral meatus, with heterogeneous digitations, dark brown to black, and irregular polycyclic borders. No inguinal adenomegalies were palpated. Cystourethroscopy and biopsy of the lesion confirm the diagnosis. Melanoma of the female urethra is an extremely infrequent pathology. Due to lack of published case reports and the absence of prospective randomized trials on treatment outcomes, treatment must be directed using the same anatomical and surgical criteria for female urethral tumors, adding also the concepts of treatment of mucosal melanoma, even though its prognosis is different from the before mentioned.

Key words: Female urethra cancer, melanoma, urethral neoplasm

INTRODUCTION

Melanoma is a malignant tumor that can affect any area of the anatomical economy. Its appearance in the female urethra is extremely rare, with approximately 121 cases in indexed literature since 1966; it was described for the first time in 1896 by Reed,^[1] most of them being published as case reports. The most extensive case series is by DiMarco from the Mayo Clinic, describing 11 cases between 1950 and 1999.^[2]

From a histological point of view, tumors affecting the female urethra are: Squamous cell carcinoma (60% of cases), transitional cell carcinoma (20%), adenocarcinoma (10%), undifferentiated tumors and sarcomas (8%) and in the last place melanoma (2%).^[2] It is more frequent in women in a 3:2 ratio.^[3] This location represents 0.2% of all melanomas.^[4] The

average age of presentation is 68 years,^[2] ranging from 32 to 80 years of age. There are only six cases published that describe disease survival over 5 years time.^[5]

MATERIALS AND METHODS

A PubMed search was conducted for manuscripts published regardless of the publication date, which contained the terms “melanoma”, “urethra”, “melanoma urethra”, and “female urethral neoplasm”, giving preference to most recent publications. Articles identified were screened for their relevance to the field of melanoma and likely interest to both urologists and oncologists. This mini review focuses on the presentation, evaluation, treatment, and prognosis of melanoma occurring in the urethra, describing our own case.

CASE PRESENTATION

The subject, an 86-year-old woman seeks medical consultation at our institution for intermittent macroscopic hematuria with blood clots of 3 weeks progression. No significant medical history is observed. Past surgical history is positive for cystocele repair 23 years ago by a combined approach, without any complications or pertinent information.

On physical examination no lesions suspicious of melanoma are observed on skin. An external genital exam showed a friable lesion at the borders of the urethral meatus, with heterogeneously colored digitations ranging from

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dark-brown to black, with irregular polycyclic borders. No inguinal adenomegalies are palpated. Biopsy of the lesion shows results consistent with infiltrating nodular melanoma, with evidence of ulceration and hemorrhage. On rigid cystourethroscopy, it is evident that the lesion partially occludes the distal third of the urethra, and extends throughout the before-mentioned sparing the neck of the bladder. A plain chest X-ray shows no evidence of metastatic lesions, and contrast abdomino-pelvic CT scan does not reveal the presence of adenomegalies, with perivesical and periurethral tissues being lesion free. The patient refuses any invasive surgical approach and has not shown clinical progression of disease to date with irregular follow-up.

Review

Clinical review

Clinical presentation is very florid, and may range from dysuria to hemorrhage. It may co-present with vulvar melanoma^[6] or vulvar, vaginal, or vesical melanosis.^[7] Lesions can vary from millimetric up to 6 cm.^[3] It has been described that up to 20% can be amelanotic, generating differential diagnosis such as transitional cell carcinoma or sarcoma,^[3,8] pagetoid infiltrations of urothelial carcinomas^[6] or even benign lesions.

Evaluation

Clinical staging includes biopsy of the lesion with or without urine cytology.^[9] The localization is almost always in the distal third of the urethra.^[2] Very rarely it has been described in the proximal or middle thirds.^[3] Metastasis from a primary cutaneous melanoma must always be excluded.

Surgical staging consists of complete resection of the lesion and lymphadenectomy of the first lymph groups associated, preferably marked by lymph node scintigraphy with technetium 99 (Tc99).^[2,6]

Anatomo-pathologic staging includes extension, depth, necrosis and lymph node, and vascular invasion.^[2] It may present with a horizontal and/or vertical pattern of growth, diffuse, or mixed.^[3]

It is convenient to use the TNM classification by the American Joint Committee on Cancer (AJCC),^[10] although for staging from a depth point, Chung's Index^[11] if more useful, which is applied in mucosal melanoma (Level 1: Limited to the epithelium, level 2: Less than 1 mm, level 3: Between 1 and 2 mm, and level 4: Over 2 mm of depth).

Treatment

The principles of melanoma treatment are wide local excision^[12] with sentinel lymph node dissection.^[13] This is based on the concept that the sentinel lymph node is the first station of metastasis, and it was described for penile cancer.^[14] A very important aspect is patient selection. In conventional melanoma, the criteria of selection are <10% of lymph node

metastasis and <1 mm of depth by Chung's Index.^[11] A possible treatment option could be partial urethrectomy, radical urethrectomy with a continent urinary diversion^[5,6] or even anterior pelvic exenteration with or without vulvectomy,^[2,5] as long as there is no clinical evidence of metastasis.^[5,15] Regional lymphadenectomy is done only in the case of positive metastasis in the sentinel lymph node, decreasing the morbidity of the procedure.^[13] Lymphadenectomy is also used for staging.^[5] Those patients with depth up to 3 mm can benefit from anterior pelvic exenteration.^[16] Radical surgery is contraindicated in cases of inguinal metastasis and in large-sized tumors.^[15] Different regimens of adjuvant immunotherapy have been described with interferon alpha^[5] or interferon beta,^[17] with or without dacarbazine, vincristine and cyclophosphamide chemotherapy.^[5,16]

Prognosis

The most significant prognostic factors for local control and overall survival is anatomic localization and tumor extension. Generally speaking, the prognosis in this case is poor due to the tendency to locally invade the vagina and the vulva.^[15] The recurrence rate is about 69%,^[2] usually in the first postoperative year, locally if partial urethrectomy was performed, or pelvic for exenteration. Also, distant recurrence may occur after primary resection at the level of lymph nodes or in the lung^[2,9] and less frequently in the liver.^[7] The survival rate at 3 years is about 38%.^[2] This is usually due to inadequate resection.^[2] It seems however, unlike cutaneous melanoma, prognostic factors such as depth of tumor invasion or tumor stage do not appear to have that much of an impact at predicting survival in mucosal localized melanoma, mostly because of its growth is usually nodular.^[3]

CONCLUSION

Melanoma of the female urethra is an extremely uncommon pathology leading to paucity of literature and any definite recommendations regarding management. Radical surgery with adjuvant chemotherapy appears to provide some response in an otherwise very poor prognostic scenario.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Abbreviations

Tc99, Technetium 99; AJCC, American Joint Committee on Cancer; CT scan, Computed axial tomography scan

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