Case Report

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Primary Malignant Melanoma of Female Urethra: A Case Report

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ABSTRACT

Primary malignant melanoma of the urethrais avery rare tumor constituting 0.2% of allmelanomas. Around 120 cases are published inliteraturetilldate. Wereportacaseofmalignantmelanomaoriginatingin theurethrainapostmenopausalwoman whichwas diagnosedonbiopsy and subsequentradicalsurgeryperformed.

Keywords: *Malignant Melanoma, Primary, Urethra*

Introduction

Malignant melanoma is a rare neoplasm accounting for 1.2% of all malignancies. Though it may affect any part of human body, genitourinary tract involvement is observed in less than 1% of cases while primary malignant melanoma of the urethra in females represents only 0.2% of all malignant melanomas. [1] Since the clinical presentation of urethral melanoma is similar to that of urothelial carcinoma and a few other benign conditions, there is frequently a delay in diagnosis. Common clinical signs include bleeding, discharge, voiding dysfunction and the presence of tumorous masses. Clinical misdiagnosis and delayed diagnosis of urethral melanomas result in a poor prognosis. [2] In this report, we present a case of malignant melanoma originating from the urethra and involving the neck of the bladder in a postmenopausal female.

Case Report

A 55 year old female came with complaints of mass at urethra since six months which was gradually increasing in size associated with bleeding, itching and dysuria. Local examination revealed a 2*2 cm sized black, indurated, non-tender mass present around urethra. Scopy showed evidence of a brownish black mass around urethra with blood clot covering it (Fig 1). An endophytic mass was seen between 11 to 3 o'clock positions compressing urethral lumen. Also there was evidence of a papillary mass at 3 o'clock position at bladder neck. The mucosa throughout the urethra was black coloured. USG showed a heterogeneous lesion in urethral region measuring 3.9*2.5*2.9cm with another lobulated hypoechoic area within measuring 2.2*1.4cm in size and shows mild vascularity within. MRI scan of pelvis showed a well defined heterogeneous soft tissue lesion measuring approximately 2.2*2.2*3.5cm in anterolateral urethra protruding into the bladder. A biopsy was performed which showed features of malignant melanoma. Thorough external examination of the patient was carried out and no cutaneous lesions were identenfied. Metastatic evaluation was negative. Abdominal CTscan showed no evidence of lymphadenopathy or metastatic foci.

The surgeons performed a radical cystourethrectomy along with total hysterectomy and appendicectomy. Bilateral external iliac lymph nodes were also removed. Grossly there was a black coloured papillary mass measuring 7*7*1.5 cm entirely occupying the urethra and extending into the neck of the bladder (Fig 2). Microscopy showed tumor cells arranged in nested pattern and scattered showing enlarged nucleus with prominent nucleoli and cytoplasm showing coarse brown pigment (Fig 3). There was brisk mitotic activity and areas of necrosis were seen. The tumor cells were also identified in the urothelial lining of the bladder neck. After bleaching the pigment disappeared confirming it to be the melanin pigment of the melanocytes. Immunohistochemistry was done for the further confirmation of melanoma.

S-100, HMB-45 and Melan A were strongly positive in the tumour cells (Fig 4). Thus, the diagnosis of primary urethral malignant melanoma was confirmed. The margins of the surgical spcecimen were clear. However, the external iliac lymph nodes dissected and vaginal flap showed evidence of metastasis.

Discussion

The first case of urethral melanoma was reported by Tyrell and Reed more than 100 years ago. It is a rare malignancy and accounts for 0.2% of all malignant melanomas and 4% of all urethral cancers. [3] Till date around 121 cases of female urethral malignant melanoma have been reported. [4]

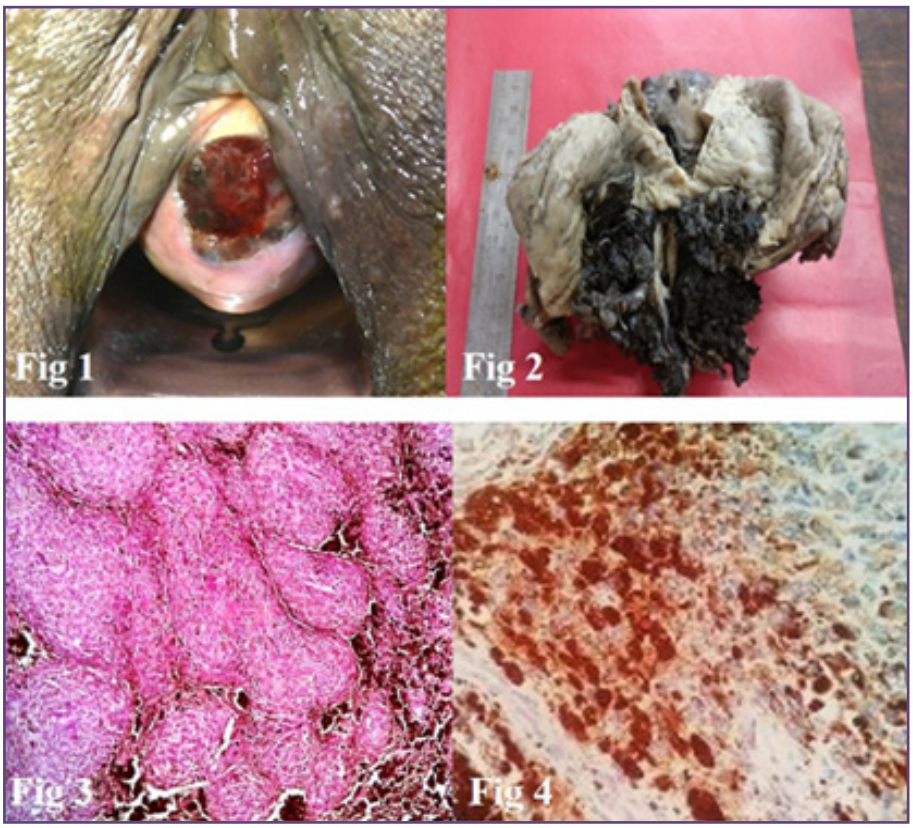

Fig. 1: Pigmented mass protruding from the urethra. Fig. 2 Black coloured urethral mass extending into bladder neck. Fig. 3 (H & E, 400 X) Tumour cells arranged in nodules with abundant brown black pigment. Fig 4. (400 X) Melan A positive tumour cells.

The histogenesis of melanoma arising in mucous membrane is still unclear and various theories have been suggested. One theory suggests that melanoblasts originating from neural crests may migrate with mesodermal cells to sites where they usually do not exist. Another mechanism put forth is that of metaplasia of squamous and glandular epithelium into pigment producing cells. [5]

Primary urethral melanoma is three times more common in females than in males. In females it commonly affects the distal urethral meatus. In males the most common site of occurrence is fossa navicularis and this predominates over pendulous, bulbous and prostatic urethra. [6]

Symptoms of urethral melanomas are nonspecific and develop rapidly. They include urethral mass, nonspecific perineal pain, dysuria, frequency incontinence and haematuria or local bleeding. [5] Extensive, thorough examination of other sites is essential to rule out a primary source of melanoma elsewhere.

The urethral malignant melanoma has a worse prognosis than a cutaneous malignant melanoma , partly because of the frequent finding of a vertical growth phase and lymph node metastasis at the time of initial diagnosis and partly because the location often results significant delays in making the diagnosis. Clinically, urethral malignant melanomas are frequently polypoid and are usually mistaken for other malignant diseases or even benign lesions, including urethral polyps, caruncle, mucosal prolapse, or chancre. A few cases of urethral malignant melanoma that were initially suspected to be urethral caruncle have been reported. [3] The tumour is usually pigmented, varying in colour from black to blue or light brownish lesions, which are firm, nodular and often ulcerated. [5] The amelanotic variant may mimic urothelial carcinoma histologically. S100, HMB-45 and Melan A are markers frequently used in the clinical practice for diagnosis of melanoma. S100 is more sensitive, HMB-45 is more specific and Melan A has both good sensitivity and specificity. [7]

Primary malignant melanoma of the female urethra tends to metastasize at an early stage to adjacent areas, the regional lymph nodes, and occasionally distant sites by the hematogenous route. The TNM classification by AJCC on cancer is useful for clinical staging although for staging

from a depth point, Chung's Index is 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). [8]

The treatment recommendations primarily depend on the tumor location and the clinical stage. The treatment varies from local excision with or without the addition of radiation therapy to extensive surgery, including cystourethrectomy, vaginectomy, vulvectomy, and lymph node dissection. The survival rate at 3 years is about 38%. This is usually due to inadequate resection. 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 its growth is usually nodular. [5, 8]

Conclusion

Urethral melanomas although rare must be considered in the differential diagnosis of pigmented lesions of urethra. Extensive workup to rule out a primary melanoma has to be done. Histological and IHC findings are helpful in the early and accurate diagnosis in these cases.

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