Unusual Cause of Hematuria

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Abstract

Primary urethral mucosal melanoma is a rare but aggressive melanoma that affects patients in their sixth and seventh decade of life. The tumor appears as a black nodule or polyp but can also be amelanotic. One of the prognostic factors is the size of the tumor. Surgery seems to be the only effective treatment. Treatments add to surgery, more particularly chemotherapy, seem to improve the prognosis and prevent recurrences, this in the absence of guidelines and clear consensus in the treatment of mucosal melanomas. The survival rate largely depends on the lymph node and visceral metastases of the disease.

Keywords

Melanoma; Vaginal; Urethral; Mucosa; Hematuria

Introduction

Genital mucosal melanomas are extremely rare, about 1% of all melanoma localizations, most frequently localized on the vulva [1]. Exceptional on the vagina or on the urogenital tract [2], generally these melanomas are diagnosed late due to their localization. Thus, they are associated with a poor prognosis. This article intends to discuss a case of primitive ureter melanoma.

Observations

A female patient 41 years-old, single; complaining of hematuria, presenting a polypoid and pigmented tumor on the urethral orifice. Clinical examination of the teguments didn’t show any suspected skin lesion or lymph nodes. The examination under anesthesia showed a pigmented urofication on the peri-ureteral area, extending to the urethral orifice, the ureter and to the vagina, extends over 4 cm intra vaginally. A biopsy confirmed the diagnosis of melanoma. The cancer is inoperable; the radiological extension assessment revealed showed the presence of enlarged lymph nodes and metastatic localizations. The patient was put on chemotherapy in the absence of surgical indication. The evolution was marked by the patient’s deaths after 6 months of chemotherapy (Figure 1).

Discussion

Urogenital tract melanomas are very rare and extremely aggressive. Vagina melanomas represent about 0.3% of all malignant melanomas end less than 3% of vagina cancers [3,4]. Urethral localizations are even rarer, there are only 121 cases described in the literature since 1966 [5,6], generally originate in the distal urethra. They represent only 0.1% of all malignant melanomas and about 2% of malignant tumors affecting female ureters [7-10]. The physiopathology is not yet quite understood, but many theories and risk factors are proposed to explain this ailment, among these, the CKIT gene mutation is highly incriminated [11]. Another theory suggests that the causal factor could be micro-environmental factors and not associated with ultraviolet radiation vue [12]. The disease affects people between 38 and 90 years old, often in women in their sixth to seventh decade [13], generally discovered as a polypoid pigmented and hemorrhagic mass, sometimes ulcerated. Amelanotic tumors have also been reported in patients, which often leads to misdiagnosis and delayed treatment [3,14]. This often leads to delayed diagnosis, treatment, and affect badly the prognosis. The confirmation of the diagnosis is based on immunehistochemistry, which is S-100 +, mélanA +, HMB-45 + and vimentin + [5,15]. The investigation is the same as skin melanoma. Nodal metastasis and distant metastasis are frequent, 50% and 20% respectively [16]. There are no well-established guidelines

Figure1: Clinical image showing a blackish tumor in the urethral meat and vaginal

Concerning the treatment. When the cancer is resectable, surgery is recommended [17], with surgical safety margins varying between 1 to 2 cm [3]. Radical surgery associated with lymph nodes dissection, chemotherapy or radiation therapy exclusive or additional to other treatments, are all options for advanced-stage malignant primitive vagina melanomas [4,18,19]. The urogenital localization is associated with very poor prognosis even if it’s a locally evolutive lesion [14]. The current treatment strategy includes surgery, chemotherapy, immunotherapy, and palliative care [5]. The available data suggest a survival rate of a relative 5 years survival rate between 0 to 25%, in case of primitive vagina melanoma regardless of the treatment strategy [16].

Conclusion
The urogenital tract melanoma is a very aggressive and rare disease, an early diagnosis is essential to an adequate treatment strategy, since the scarcity of data and guidelines regarding treatment measures, is making the management of this disease very difficult and challenging.

References