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Review

Primary malignant melanoma of the vagina: a case report and review of the current treatment options

Hüsnü Gökaslan, Alper Şişmanoğlu *, Tanju Pekin, Handan Kaya, Nejat Ceyhan

Marmara University Hospital, Marmara School of Medicine, Department of Obstetrics and Gynecology, Altunizade, Istanbul, Turkey

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Abstract

A case with primary malignant melanoma located in the posterior lower third of the vagina was encountered and treated by surgery, postoperative immunotherapy and chemotherapy. The tumor was close to the anal sphincter and posterior exenteration was done to achieve tumor-free surgical margins. The need for such radical treatment prompted us to review the literature and discuss our case with the rarest localization of the tumor in the vagina. We focused on the treatment options and the possible complication that may arise during the treatment of the primary malignant melanoma of the vagina. The need for radical surgery was discussed and the current treatment options were reviewed.

Keywords: Vagina; Melanoma; Posterior exenteration

Contents

1. Background

Malignant melanoma which has a highly malignant potential is a tumor of the melanocytes of the skin and mucosal membranes. Melanocytes are embryologically derived from neural crest cells and can be found in the basal portion of the vaginal epidermis in 3% of healthy women. Some of these melanocytes *are located aberrantly* in the vaginal mucosa and these are the sites that vaginal melanoma is thought to arise [1]. Vaginal melanoma is *rare* and there are less than 250 cases in the literature [2]. It accounts for 2.6–2.8% of all primary malignant tumors of the vagina and 0.4–0.8% of all malignant melanomas in women [1]. The average age at presentation is 57, and the most common presenting symptoms are vaginal bleeding, vaginal discharge or a palpable mass [3]. Due to the ability of the tumor to spread hematogenously, early metastases are common and regardless of primary therapy chosen, the prognosis is poor with reported 8.4% 5-year survival rate [1]. The tumor is primarily found in the distal one-third (58%) of the vagina and mostly on the anterior wall (45%). A low occurrence of this type of tumor makes the assessment of various treatment options difficult. As treatment options, there are some standard modalities used individually or in combination, such as conservative wide local excision, radical surgical extirpation, irradiation and chemotherapy [1]. In the present report, due to the tumor's localization in

* Corresponding author. Tel.: +90 5325159713; fax: +90 2163276809.
E-mail address: docalper@hotmail.com (A. Şişmanoğlu).

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the lowest posterior part of the vagina near the rectum, we preferred and performed posterior exenteration as a radical surgical extirpation. It is necessary to discuss the difficulty in making correct decision for therapy with the available current literature. As we have noticed, the review of references quoted in the previous publications and the computer search by using the key words ''melanoma'' and ''vagina'' in the Pubmed may not help adequately solve the problem. Therefore, our experience in this case related with the clinical presentation and evaluation, preferred method of treatment, complication and outcome is a particular source of information.

2. Case

Ten years postmenopausal, 56-year-old patient was complaining of vaginal discharge and vaginal bleeding in spotting type, first appeared 3 months ago. With these complaints, the patient was referred and evaluated in Gynecologic Oncology Section of Marmara University. The medical history was unremarkable except a tonsillectomy at 10 years old. The patient was cigarette smoker for 20 years. There was no malignancy in the family history. During the examination, a nodular lesion was felt in the posterior wall near the introitus. The location of the tumor was in the lowest part of distal vagina. The greatest diameter of the lesion was 1.5–2 cm and it *bleeds* easily in contact. The general physical examination was negative. Pap smear showed no atypical cells.

An excisional biopsy of the lesion taken under local anesthesia was reported as malignant melanoma and the whole biopsy material contained these malignant cells. Since the tumor was nodular in type and the base was fixed to the anterior rectal wall and anal sphincter, it was dissected from the rectum under the guidance of a finger inserted into the anal canal. Around the lesion, 1 or 2 cm in width of vaginal wall was also removed. Microscopically, the squamous epithelium was ulcerated and infiltrated by the neoplastic cells. Nodular tumor growth pattern was both pseudoglandular and solid in nature. The tumor cells were with wide eosinophilic or clear cytoplasm containing large and prominent nucleus with nucleolus. Focal necrotic areas, high percentage of typical and atypical mitotic figures (10–15/10 hpf) were seen. There were no perineural, lymphatic and/or intravascular invasion of the tumor (Fig. 1).

Immunocytochemistry was done using cytokeratin, chromogranin, HMB-45 and S-100 protein with avidin–biotin complex method. Tumoral cells were positive for the HMB-45 and S-100 proteins showing either intranuclear or intracytoplasmic immunoreactivity (Fig. 2). On the contrary, cytokeratin and chromogranin were both negative.

It was reported as nodular type malignant melanoma and the depth of invasion was more than 2 mm. According to Clark Classification [3] or Chung Classification [3] which is more widely used in vaginal melanomas, the stage was Level IV.

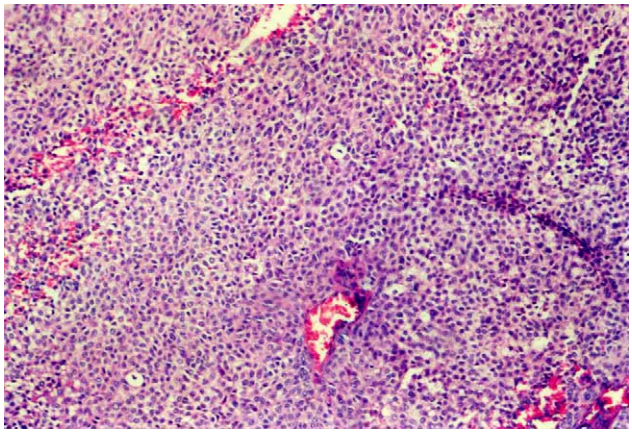

Fig. 1. Malignant melanoma in vagina (H&E ×20).

Preoperative evaluation of the patient showed no clinical evidence of metastasis. Routine laboratory tests were normal. Chest X-ray revealed no sign of metastatic lesion or pleural effusion. Computerized tomography (CT) of the pelvis clearly showed no evidence of local spread and lymphadenomegaly. Transabdominal sonogram proved negative for liver metastasis and abdominal mass or fluid.

The surgical procedure was posterior exenteration including total abdominal hysterectomy with bilateral salpingo-oophorectomy and the removal of the whole vagina with rectum. Bilateral inguinofemoral lymphadenectomy was also added. The patient was operated in the dorsal lithotomy position and the surgery was performed by abdomino-perineal approach. Infralevator posterior exenteration was preferred due to the location of the lesion and the surgery was completed with sigmoid end colostomy. In the exploration of the abdomen, the abdominal organs were found normal and there was no grossly palpable lymph node in the pelvic and paraaortic area. Since the diagnosis was confirmed preoperatively and the most extensive surgery was contemplated for the patient, there was no need to do frozen section during the surgery (Fig. 3).

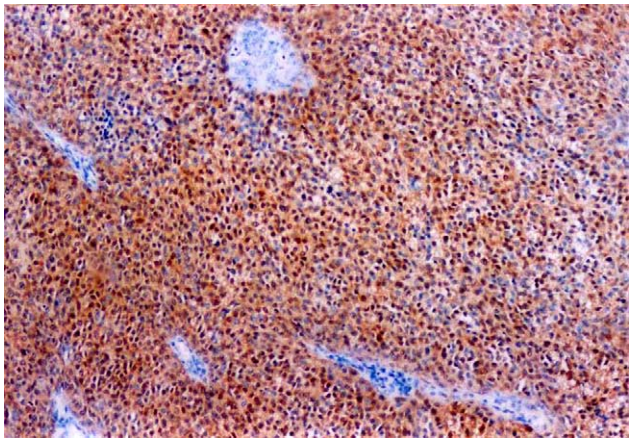

Fig. 2. Intracytoplasmic and intranuclear S-100 immunoexpression of the malignant melanoma cells.