

Malignant melanoma of the vagina

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ABSTRACT

Carcinoma of the vagina is uncommon and only constitutes two percent of malignant neoplasms of the female genital tract. Malignant melanoma of the vagina is rarer and accounts for only 3.2% of all vaginal cancers. It presumably arises from melanocytes found in the vagina of three percent of normal women. It is a very aggressive tumour and the overall prognosis is poor despite the treatment. We report the case of an 81-year-old woman who presented with per vagina bleeding secondary to a malignant melanoma of the vagina. She was treated with radiotherapy as her disease was locally extensive and she was not a candidate for pelvis exenteration. This is the first recorded case of malignant melanoma to be diagnosed in Brunei Darussalam.

Keywords: Melanoma, vaginal neoplasm, surgery,

INTRODUCTION

Carcinoma of the vagina is uncommon and only constitutes two percent of malignant neoplasms of the female genital tract. Primary malignant melanoma of the vagina is rare with less than 250 reported cases worldwide. It accounts for less than one percent of all melanomas in women and 3.2% of all vaginal cancers. In the female genital tract it is more common on the vulva.¹ The average age of diagnosis is 58 years but can range from the third to ninth decade of life.² Almost all cases of primary malignant melanoma of the vagina have been reported in Caucasian. Its clinical behaviour is notoriously more

aggressive than that of cutaneous and vulvar melanomas with a five year survival rate ranging from 5 to 25%. The tumour size is the strongest predictor of survival unlike cutaneous melanoma where tumour thickness predicts survival.³

We report the case of an 81-year-old woman with malignant melanoma presenting with bleeding. To our knowledge this is the first recorded case of vaginal malignant melanoma to be diagnosed in Brunei Darussalam.

CASE REPORT

An 81-year-old Chinese woman presented to the Department of Obstetrics and Gynaecology clinic with an initial episode of post menopausal bleeding of 17 days duration. The bleeding was considerable and was soaking

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two sanitary pads per day. There were no associated symptoms such as anorexia and weight loss. She had had three children, all normal vaginal deliveries and her last childbirth was 46 years earlier. She had been menopausal for more than 30 years and she had never had any Pap smear previously. She had hypertension (on perindopril 4 mg daily and Nifedipine LA 30 mg daily) and hyperlipidaemia (Atorvastatin 10 mg daily). She had no other relevant surgical and family history.

Systemic examination was unremarkable. She had no lymphadenopathy, ascites, hepatosplenomegaly or abdominal mass. Per vaginal examination revealed a bluish black growth at the lower posterior vaginal wall. She underwent an examination under general anaesthesia (EUA) and this revealed a blackish vaginal epithelium at the vestibule with a normal urethral orifice. There were two black coloured deposits measuring 0.5 x 0.5 cm and 0.2 x 0.5cm on the posterior lip of cervix with a circular dark haemorrhagic growth measuring 3 x 3 cm in diameter (Figure 1) arising from the lower half of the posterior vaginal wall with minimal surrounding induration.

There were a few satellite lesions on

both lateral vaginal walls. The right inner one third of the parametrium was involved but the left parametrium was not affected. The pre-rectal space and the rectal mucosa were not affected. The uterus was atrophic, retroverted and no adnexal mass was palpable. Punch biopsies were taken from the two cervical lesions and three biopsies were taken from the junction of vaginal growth and normal epithelium. Although endometrial curettage was performed no sample was obtained.

The histopathology confirmed that the tumour was a malignant melanoma of the vagina and cervix. Few melanoma cells were seen with high nuclear-cytoplasmic ratio, nuclear hyperchromasia and dark brown melanin pigment in some (Figure 2a). The cells were positive for S-100 (Figure 2b) and HMB-45 (Figure 2c). Urine cytology was negative for malignant cells. Pap smear showed an atrophic pattern with few melanoma cells.

A staging computed tomography (CT) scan of pelvis, abdomen and thorax showed a 6.2 x 5.6 cm mass in the lower part of the vagina without any evidence of metastases. Based on the findings, a diagnosis of malignant melanoma of the vagina, Clark's level

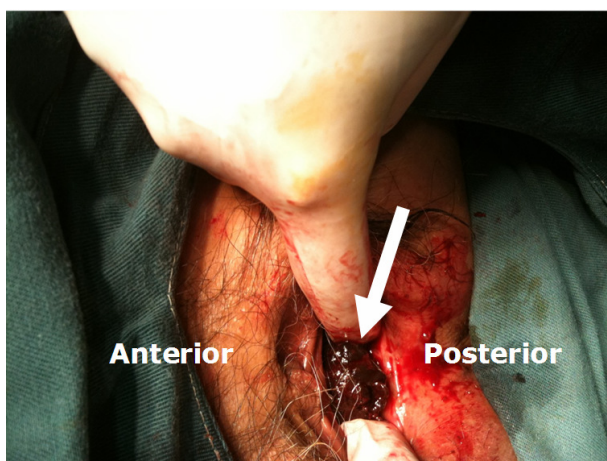


Fig. 1: A black pigmented mass (Malignant melanoma) (white arrow) arising from the posterior wall of the vagina as seen during examination under anaesthesia (EUA).

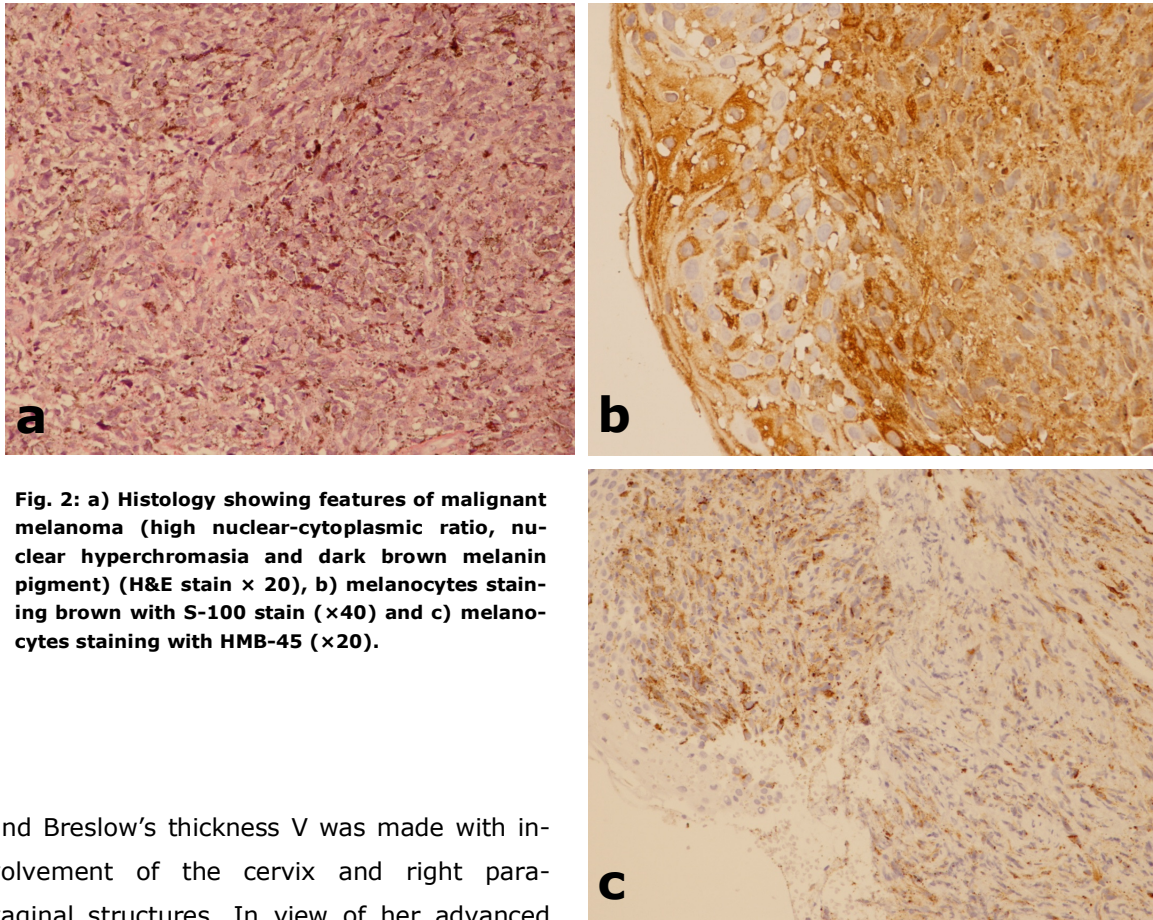


Fig. 2: a) Histology showing features of malignant melanoma (high nuclear-cytoplasmic ratio, nuclear hyperchromasia and dark brown melanin pigment) (H&E stain $\times 20$), b) melanocytes staining brown with S-100 stain ($\times 40$) and c) melanocytes staining with HMB-45 ($\times 20$).

and Breslow's thickness V was made with involvement of the cervix and right paravaginal structures. In view of her advanced age and extensive local spread, neither a pelvic exenteration nor a wide local excision were recommended. Therefore she was offered radiotherapy for local control of the disease for which she was sent abroad for the treatment. The patient received a course of radiotherapy in a referral centre in a neighbouring country. The patient passed away five months later.

DISCUSSION

Primary malignant melanoma of the vagina is a rare entity and was first reported by Poronus in 1887.⁴ It is mainly a disease of post menopausal women.⁵ Clinically most patients present with vaginal bleeding, mass or discharge.⁶ The lesions arise most commonly in the distal part of the vagina particularly on the anterior wall.⁴ Grossly the tumour

appears as a black or greyish nodular or polypoid or fungating mass. In the majority of cases the tumours are polypoid to nodular. Most are deeply invasive. In 60% of cases, there is spread of melanocytic cells into the adjacent epithelium and in approximately 30% of cases the lateral spread is extensive.⁴ They may be non-pigmented and are frequently ulcerated making them easily confused with squamous carcinomas.⁶

The International Federation of Gynecology and Obstetrics (FIGO) system used for classification of squamous lesions of the vagina is not applicable for melanomas. Instead the Breslow and Clark systems (Table 1) are used as part of staging.⁶

Table 1: Histological staging of malignant melanoma, both level of invasion (Clark's level) and tumour thickness (Breslow's thickness) should be measured. ¹¹

	Clark's levels	Chung et al	Breslow's thickness
I	intra-epithelial	intra-epithelial	<0.76 mm
II	into papillary dermis	≤ 1 mm from granular lesion	0.76-1.50 mm
III	filling dermal papillae	1.1-2 mm from granular lesion	1.51-2.25 mm
IV	into reticular dermis	>2 mm from granular lesion	2.26 -3.0 mm
V	into subcutaneous fat	into subcutaneous fat	>3 mm

The differential diagnosis of malignant melanoma of the vagina includes metastasis from malignant melanoma from other sites, poorly differentiated squamous cell carcinoma, sarcoma, lymphoma and blue naevus. ⁹ Immunohistochemical studies are of value in differentiating the differential diagnosis. ⁵

Microscopically the lesion shows proliferation of atypical melanocytes along the dermo-epidermal junction with extension into the squamous epithelium. The atypical melanocytes may be epithelioid, spindled or mixed and occur singly or in clusters. Malignant melanoma is positive for S-100 antigen and the melanoma associated marker HMB-45.

Radical surgery has traditionally been the mainstay of treatment and this has often involved anterior, posterior or total pelvic exenteration. ⁶ Small upper vaginal lesions have been treated with radical hysterectomy, sub-total vaginectomy and pelvic lymphadenectomy whereas small distal vaginal lesions have been treated by partial vaginectomy, total or partial vulvectomy and bilateral inguinal lymphadenectomy. ^{7, 8} More recently conservative options like wide local excision have

been used followed frequently by pelvic radiotherapy. However, there appears to be no significant benefit in the survival or disease-free interval for radical versus conservative surgery. ^{1, 3, 7, 8} Chemotherapy using semustine or dacarbazine has also been disappointing. ⁶

The overall prognosis is poor as most patients have locally advanced disease at the time of diagnosis. ⁶ Reviews by Reid *et al.* and Buchane *et al.* noted that the size of the lesion was the best prognostic factor with lesions more than 3cm having a worse prognosis. ^{3, 4} Once recurrence is noted, prognosis is extremely poor with a mean survival time of 8.5 months. ⁶

Adjuvant therapies with interferon alpha-2 have been shown to improve the relapse free period and overall survival in patients with high risk cutaneous melanoma but there are as yet no data on these for vaginal melanoma. ¹⁰

In conclusion, although rare, melanoma forms one of the differential diagnosis of primary vaginal malignancy and carries a poor prognosis.

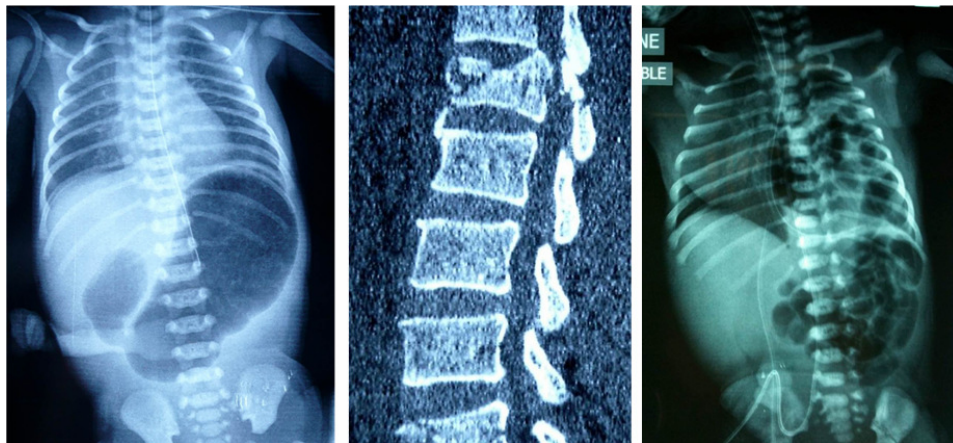
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