

MALIGNANT MELANOMAS OF THE VULVA: TWO CASE REPORTS

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INTRODUCTION

Malignant melanomas of the vulva are rare tumours which constitute only 5–10% of all vulvar cancers but are the second commonest vulvar malignancy after squamous carcinomas. Almost all vulvar naevi are of the junctional type from which melanomas are believed to arise and this may explain why in women, 3–5% of melanomas occur in the vulva though it occupies only 1–2% of the body surface area. Vulvar melanomas have a poor prognosis with an overall 5-year survival rate of only 34% in a recent review.¹

Vulvar melanomas are much more common in fair skinned Caucasian women but very rare in our local women who have a much darker skin. We therefore felt it important to report and record our experience with vulvar melanomas occurring in both types of individuals to emphasize that melanomas should be considered as a diagnosis in malignant lesions of the vulva, and the poor prognosis and unusual spread pattern of this neoplasm compared to squamous cancers. Microstaging of vulvar melanomas as a valuable prognostic method to enable therapeutic indi-

dualization and less extensive surgical treatment of early lesions is discussed.

CASE REPORTS

Case 1

M.K., a 38-year old Caucasian female, presented in June 1977 with a 9-month history of vulval soreness and a vulvar ulcer. Examination revealed a 1 cm diameter ulcer on the right labium majus and a smaller nodule on the ipsilateral labium minus without inguinal lymphadenopathy. The rest of the examination was entirely negative and an excision biopsy of both lesions reported an invasive squamous cell carcinoma. By criteria of the International Federation of Gynaecology and Obstetrics (FIGO), she was in Stage I of the disease, and a radical vulvectomy and bilateral inguinal lymphadenectomy were performed; a histopathologic examination confirmed an invasive squamous carcinoma without lymph node involvement. Eighteen months later an enlarged liver with a hard nodular edge was detected clinically and a liver biopsy confirmed a metastatic poorly differentiated carcinoma. Due to unusual behaviour of an early squamous carcinoma with negative lymph nodes the histopathologies of all specimens were reviewed and the diagnosis changed to that of malignant melanoma. The patient died 10 months after the liver biopsy.

Case 2

M.B.M., a 68-year old Malay lady, presented in July 1985 because of a 2-month history of a lump in the vulva. Examination revealed a darkly pigmented nodular area at the distal 2 cm of the urethra involving the meatus and upper half of the right labium minus, and 1 cm laterally and three darkly pigmented satellite nodules mea-

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suring 0.5 cm in diameter. A separate mass affected the lower portion of the right labium minus with spread to the right lateral and posterior vaginal walls. The right inguinal nodes were firm, enlarged and matted together, forming a 4 x 4 cm mass fixed to the underlying fascia. Apart from obesity, the rest of the examination was negative. Biopsy of the upper right labium minus mass showed a nodular melanoma (Fig. 1) with surface ulceration and invasion of the subcutaneous fat. The thickness of the tumour could not be determined as the deep margin was not clear but it was more than 1.5 mm. Biopsy of the satellite nodules showed junctional activity in adjacent skin (Fig. 2). There was a moderate lymphoid response and vascular invasion and neurotropism could be seen. A diagnosis of FIGO Stage III disease was made; a chest radiograph was normal and a computerized tomographic (CT) scan of the abdomen and pelvis revealed no enlarged pelvic or paraaortic nodes or liver metastases. The patient refused any treatment and died 8 months later.

DISCUSSION

Malignant melanomas of the vulva are extremely rare in local women and only 4 cases have been recorded in the Singapore Cancer Registry since 1967. Also, 80% of vulvar melanomas occur in the labia minora and clitoris and are elevated, darkly pigmented, nodular, and frequently ulcerated lesions. Diagnosis may be difficult when pigmentation is lacking or slight due to pseudo-epitheliomatous hyperplasia and keratinization of the overlying epidermis¹ as in the first case. But an inflammatory reaction around the tumour or the presence of satellite nodules as in our second case, may suggest melanoma. Small suspect lesions are best evaluated by excision biopsy, but in large lesions a wedge biopsy is sufficient for diagnosis.

The extent of clinical disease at presentation is a major determinant of prognosis in vulvar melanomas, and is poor when regional nodes are clinically involved with only a 15% 5-year survival rate,¹ a point illustrated by the second patient who died 8 months following diagnosis. However, even when regional nodes are not clinically involved

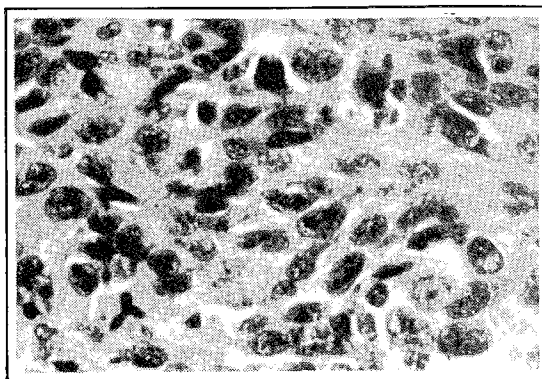


Fig. 1 Malignant melanoma. Pigmented tumour cells are irregular with several mitotic figures. A tri-polar mitosis is seen in the left lower quadrant of the illustration. (Haematoxylin and eosin x 650)

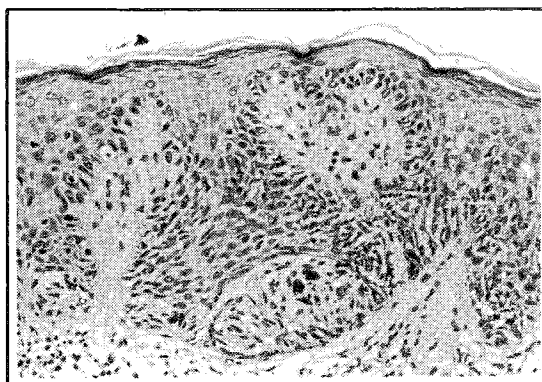


Fig. 2 Adjacent skin showing junctional activity (Haematoxylin and eosin x 240)

the 5-year survival rate is only 45% for vulvar melanomas compared to 90% in squamous carcinomas since spread often occurs to the liver as illustrated by the first case or commonly to the lungs or brain. These features clearly indicate a need to identify more accurate and significant prognostic criteria in early vulvar melanomas than clinical staging.

In cutaneous melanomas five anatomic levels of invasion (Clark's levels) relate to prognosis and survival. In level 1, the melanoma is intraepithelial; in level II, the melanoma invades the papillary dermis but not the reticular dermis; in level III, the tumour cells abut on but do not invade the reticular dermis whereas in level IV, there is invasion of the reticular dermis and in level V of the subcutaneous fat. The alternative

method of Breslow² which measures depth of invasion and vertical tumour thickness with an ocular micrometer has shown that melanomas of less than 0.76 mm thickness have the best prognosis, but is infavourable in those exceeding 1.5 mm thickness. While direct measurement is more objective than determination of histological levels, errors may occur due to mal-orientation or miscalculation. Thus a combination of methods is usually used. An extensive study³ which evaluated both Clark's levels and Breslow's tumour thickness in vulvar melanomas confirmed their validity as good prognostic indices with a fall in 10-year survival from 100% in level II to 23% in level V lesions. Similarly, the study confirmed the excellent prognosis (100% survival) of lesions less than 0.76 mm in thickness emphasizing the value of microstaging. Microstaging identifies lesions which by the above criteria have an excellent prognosis and allows omission of groin lymphadenectomy as part of the radical surgical procedure in certain FIGO Stage I vulvar melanomas with consequently less morbidity.

With advanced disease, large nodular lesions and clinically positive nodes appear as in the

second case. The prognosis is so poor that the value of radical or ultra-radical surgery is uncertain. However, there may be place for simple surgery for a local "toilet" effect to palliate local symptoms and facilitate nursing care until, a reasonable prospect of cure is likely in such case, which will require some form of effective systemic adjuvant therapy in addition to surgery.

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