Case Report

Squamous Cell Carcinoma Vulva in a Young Woman

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ABSTRACT

Carcinoma of vulva is an uncommon malignancy accounting for only 2% of all female genital malignancies.[1] It is usually seen in postmenopausal women in age group ≥65 yrs. Increased life expectancy has given place to carcinoma vulva among gynaecological malignancies. However in recent years there is an increased incidence (almost doubled) of vulvar cancer in younger women.[2] We report a case of invasive squamous cell carcinoma of vulva stage IIIb in a 36 year old woman, indeed first only in thirty years of gynaecological practical experience. The relative rarity of vulvar cancer in young age and the general lack of awareness of typical signs and symptoms even by medical professionals frequently lead to a delay in diagnosis.

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Introduction

Carcinoma of vulva is an uncommon malignancy accounting for only 2% of female genital tract malignancies.[1] It is a disease of postmenopausal women with a median age at diagnosis of about 65 years. In the past three decades the incidence has doubled in younger women in U.K.[2] Squamous cell carcinoma accounts for more than 90% of vulvar cancers, while 10% include melanomas, sarcomas, basal cell carcinomas and adenocarcinoms.[3] Toki and collaborators[4] divided vulvar squamous cell carcinoma into two distinct groups. The first group comprises the vast majority of vulvar malignancies that occur mostly in older women. These neoplasias are keratinizing squamous cell carcinomas (KSCs), and human papillomavirus (HPV) is usually not found in them. The second group comprises a minority of vulvar malignancies occurring in younger women.This second group of tumors usually show a basaloid or warty histology and frequently are positive for HPV DNA.[4] We report a case of keratinizing squamous cell carcinoma vulva in a 36-year-old woman; which is otherwise seen in the postmenopausal age group.

Case Report(S)

A 36 year old obese woman Para2 presented to Out Patient Department of Gynaecology, Government Medical College & Rajindra Hospital Patiala with chief complaint of intense vulval itching & burning sensation with dysuria and burning micturition for last three months. On local examination 5x4 cm angry red, ulcerated, oozing lesion was seen on upper part of right labia majora and minora (fig 1).

The growth was involving part of clitoris and right half of urethral meatus. Thick white discharge was oozing out of the lesion. Non tender matted lymph nodes about 3x3cm were palpable in right inguinal region. Vagina, cervix and uterus appeared normal on per speculum and per vaginum examination. There was no history of smoking, diabetes mellitus or hypertension. There was no history of genital malignancy in the family. Human Papilloma Virus Real time PCR was found negative. PAP smear was negative for intraepithelial lesions of malignancy. Patient was negative for VDRL and HIV infection. CT scan abdomen and pelvis depicted enlarged nodes measuring 3.4x3.2 cm in right inguinal region. Subcentimetric sized nodes are seen in left inguinal region. Biopsy from growth confirmed diagnosis of moderately differentiated keratinising squamous cell carcinoma vulva, usually not seen in young women. FNAC from right inguinal nodes show dysplastic cells with metastatic squamous cell carcinoma. A diagnosis of keratinizing squamous cell carcinoma vulva stage IIIb was made as per revised FIGO staging for vulvar cancer 2009.[5] Patient was treated with chemoradiotherapy

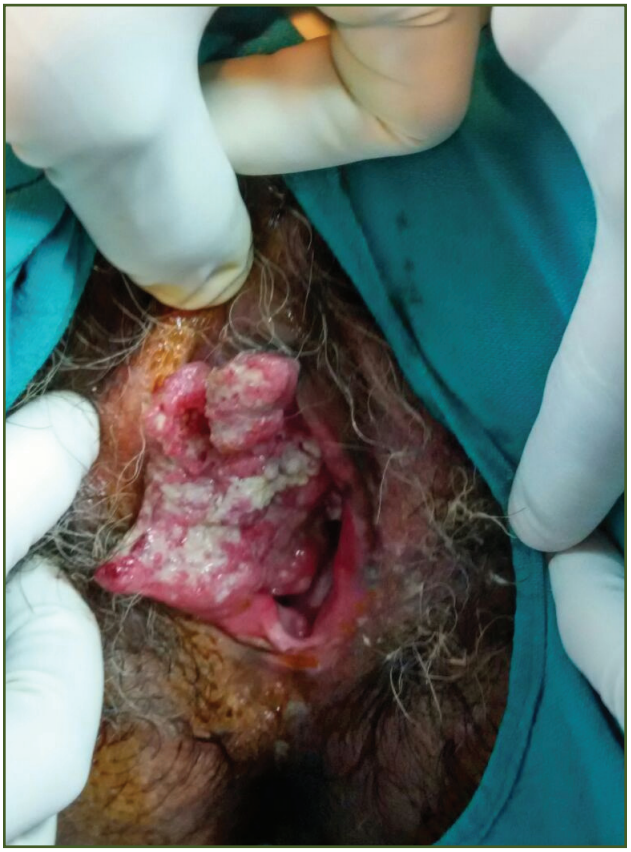

Fig. 1: Angry red, ulcerated, oozing lesion on upper part of right labia majora and minora

without surgical resection. 57.6 Gy intensity modulated radiotherapy (IMRT) with chemosensitising dose of cisplatin. Patient will be followed every three months for one year, six monthly for next five years and then annually. However patient advised to report earlier in event of any ongoing problem or recurrence of growth.

Discussion

Vulvar cancer is primarily a disease of postmenopausal women yet upto 15 % of vulvar cancers are diagnosed in women less than 40 years .[6] Risk factors for vulvar cancer include cigarette smoking, vulvar dystrophy (eg lichen sclerosis), vulvar or cervical intraepithelial neoplasia, human papillomavirus infection, immunodeficiency syndromes, a prior history of cervical cancer etc.[7] Our patient was 36 year old with a diagnosis of moderately differentiated squamous cell carcinoma vulva however none of these risk factors were present in our patient. Vulvar carcinoma in situ (VIN) tends to be multifocal with a lower risk of invasive cancer in younger women but higher risk in older women. Thus VIN 3 should be treated with mandatory long term follow up. Two independent pathways of vulvar carcinogenesis are felt to currently exist, the first related

to mucosal HPV infection and second related to chronic inflammatory (vulvar dystrophy) or autoimmune processes.[8] Women with vulvar carcinoma may present with a vulvar lesion or vulvar pruritis with burning sensation and pain. Cervical cytology and/or cervical examination are recommended, as women with carcinoma of the vulva are at an increased risk of developing other anogenital cancers, particularly cervical cancer.[9,10] In early vulvar tumours the risk of lymph node metastases is reported as low. Clinical assessment of the lymph nodes alone is not recommended.[11] Due to the significant morbidity associated with groin lymphadenectomy, high-resolution imaging may be used preoperatively to stage disease and potentially detect those women who may not require lymphadenectomy. After FNAC when metastasis in inguinal nodes is confirmed disease is staged as stage IIIb (FIGO 2009). Our patient was also stage IIIb vulvar squamous cell carcinoma. Thirty per cent of patients are reported as having lymph node metastasis at presentation.[12]

Modified radical vulvectomy with unilateral/bilateral inguinal/femoral lymphadenectomy depending on stage of disease remains standard therapy applied to most patients with vulvar carcinoma. Certain tumour characterstics may preclude an otherwise medically fit patient for undergoing primary surgery. Proximity to important functional structures such as urethra, clitoris and anal sphincter must be considered. If adequate surgical margins can not be obtained without sacrificing such a structure neoadjuvant treatment with radiation and/or chemotherapy should be considered.[1] Our patient was also treated with IMRT with chemosensitising dose of cisplatin. Advanced-stage disease (FIGO III–IV) may be difficult to manage, especially in cases where primary disease involves the anus, rectum, urethra, bladder or bulky groin nodes. Preoperative radiotherapy may allow for shrinkage of primary tumour (eg when trying to achieve sphincter-preserving surgery). Radical radiotherapy is used in patients for whom surgery is not an option and is usually combined with chemotherapy.[13] Primary tumour factors that appear to have prognostic importance include tumour diameter, depth of invasion or tumour thickness, tumour differentiation, lymphovascular space involvement and surgical margin status. Tumour involvement of distal urethra, vagina or perineum is also an adverse prognostic factor.[1] In our patient growth was involving right half of urethra and clitoris along with involvement of right inguinal lymph nodes. The five-year survival rate for women with positive nodal disease (<50 per cent), however, is much lower than for those with node-negative disease (>80 per cent).[6] Groningen international study on sentinel nodes in vulvar cancer (GROINSS-VII)[14], is looking at whether it is safe to have radiotherapy instead of surgery when vulvar cancer has spread to the sentinel nodes.

Invasive squamous cell carcinoma in our patient (young woman) is not completely clear as none of the associated risk factors were present. HPV infection is a known cause of vulvar carcinoma in young patients but our patient was negative for HPV/HIV infection with normal pap smear. she might have an unknown immune deficiency that allowed abnormal cells to proliferate and progress to cancer. Long term follow up after treatment of vulvar cancer is recommended. Every three months for one year, every six months for five years and annually thereafter. Annual cervical or vaginal cuff cytology is warranted as 15-40% patients may have local recurrence.[1]

Conclusion

Vulvar self examination for all women on monthly basis should be promoted for early detection of vulvar cancer as recommended by vulval health awareness campaign (VHAC). Biopsy of any questionable vulvar lesion prior to empirical treatment is warranted. A careful pelvic examination and pap smear screening must be done in patients with vulvar carcinoma as these women have increased incidence of cervical and vaginal neoplasia.

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Competing Interests

None Declared

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