HPV related malignancy : Vulvar carcinoma at a young age

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A 28-year-old Fijian female presented with a two-month history of a vulvar growth.
Office biopsy showed severe dysplasia but clinically the lesion was invasive, consistent with a stage II vulvar carcinoma. She underwent radical wide excision with bilateral inguinofemoral lymphadenectomy and final pathology confirmed invasive disease.

Introduction :

Vulvar cancer is relatively uncommon, accounting for 3 to 5 percent of female genital-tract malignancies. Over the past decade, an increase in vulvar intraepithelial neoplasia (VIN) and VIN-related invasive vulvar cancer has been noted in women younger than 50 years.[1]

Case report :

A nullipara presented with a two month history of a posterior vulvar growth .The growth was increasing in size and associated with pain and discharge which was blood stained at times.

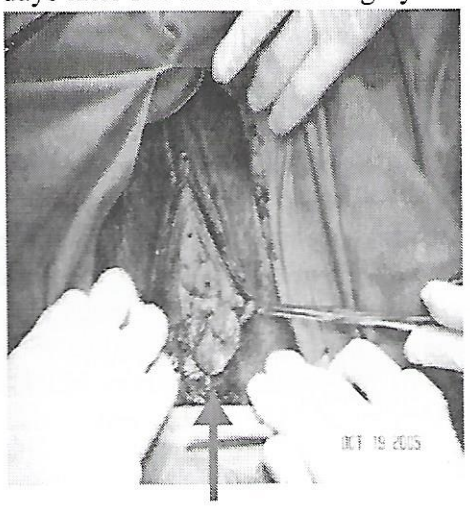
She was sexually active and reported one current partner. She did have multiple partners in past and had been sexually active since the age of 15 years. She was also a chronic smoker since the age of 15 years.

Her VDRL titre was positive in 1:64 dilutions and was treated with Benzathine Penicillin

2.4 MU weekly for 3 weeks. HIV and HBsAg tests were non-reactive.

On examination she was found to have a ulcerative growth, 2x2cm in size at the posterior fourchette, starting from midline extending to the left side. The remainder of the physical examination was normal and the inguinal lymph nodes were not enlarged. A punch biopsy was taken and the histology showed severe dysplasia with squamous cell carcinoma in-situ.

On return visit three weeks later she reported that the size was increasing. The lesion was noted to be 3x3cm in size and involving deeper tissues of the perineal body. The clinical diagnosis was invasive cancer. She was counseled about the recommendation for surgical treatment. 8 days later she underwent surgery.



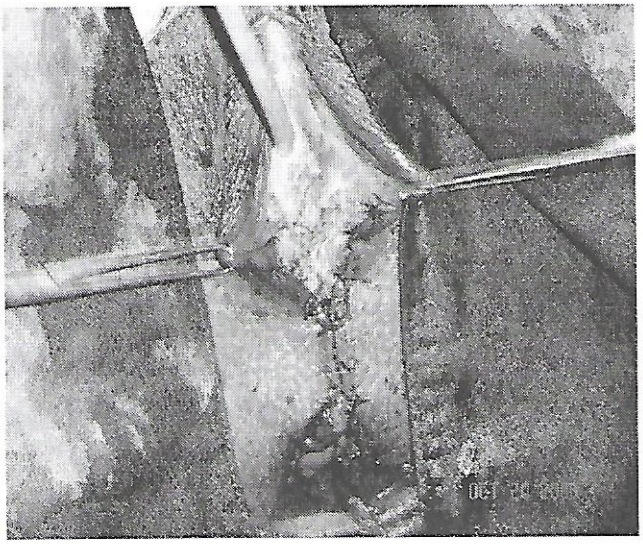
Growth at Vulva

Wide radical excision with bilateral groin lymph node dissection was performed. The clinical margin of resection was 2 cm from the lesion except at the anus where only 1 cm was obtained in order to preserve the external anal sphincter. All perineal body tissues anterior to the external anal sphincter and the rectal wall were excised, via

development of the rectovaginal space, dissecting anterior to the sphincter muscle but deep to the muscular capsule. Primary closure of the surgical defect was done with minimal tension after plication of the levator ani muscles in the midline and mobilization of the vagina.

Postoperatively the inguinal drains were kept for 10 days and the wounds at both groins were clean and healing at the time of discharge after 10 days. Histology confirmed the diagnosis of an invasive, well differentiated squamous cell carcinoma with margins free of tumour and 17 lymph nodes without metastasis.

She was followed in the outpatient clinic and at the followup visit one month later the wounds had completely healed.



Vulva after reconstruction

Discussion :

Vulvar cancer most frequently occurs in women 65 to 75 years of age. [2] It can develop in younger patients, and a recent review [3] noted that approximately 15 percent of all vulvar cancers occur in women younger than 40 years of age. One study [4] has noted the increasing incidence of VIN in young women during the past 2 decades and is being reflected now in VIN-associated squamous cell carcinoma of the vulva in younger women.

It has been suggested that vulvar cancer exists as two separate diseases. The first type involves human papillomavirus (HPV) infection, which leads to VIN and predisposes the patient to vulvar cancer occurs at younger age and has strong association with history of other sexually transmitted infections and cigarette smoking whereas other type is unrelated to HPV infections occurs in elderly women.[5] A retrospective review [6] of women with vulvar cancer has found a statistically significant correlation between patients younger than 45 years and HPV , cigarette smoking , having more than two sexual partners , sexual initiation before age 19 years and low socioeconomic status. Most of these features were present in our case, suggesting HPV causality, but unfortunately we could not perform HPV testing in our case due to financial constraints.

One case report describes a patient with SLE who developed invasive squamous cell carcinoma of the vulva at age 25 years and it was speculated that the SLE itself and/or the treatment with immunosuppressive drugs permitted malignant transformation and the development of invasive disease in such a young patient.[7]
A recent review [8] has described three cases of human immunodeficiency virus-infected women who were diagnosed with vulvar cancer before age 40 years .

The unusual feature of our case is the diagnosis of invasive vulvar cancer at 28 years of age without identifiable predisposing immunosuppressive disease or therapy. She tested negative for HIV infection and had no other significant medical history other than syphilis and smoking.

She has undergone the appropriate surgical treatment for patients with invasive vulvar cancer. We expect a good prognosis for her as groin lymph nodes did not show metastasis. We are concerned that the age at presentation of vulvar cancer may be decreasing and encourage clinicians to consider the diagnosis and perform biopsy as needed, even in younger women if the clinical findings are suspicious.

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