Original Research Article

Evaluation of the Clinicopathological Pattern and Treatment outcome of Vulval Carcinoma at a Tertiary Hospital in Port Harcourt, Nigeria.

Abstract

Background : Vulva cancer is a rare but important genital tract malignancy. **Aims :** This pioneer study was to determine the prevalence, sociodemographics, clinical présentation, histological pattern and treatment outcome at the University of Port Harcourt Teaching Hospital (UPTH). **Materials and methods:** This was a retrospective study of all cases of vulva cancer at the UPTH over a 10-year period from January 1st 2010 to December 31st 2019. Case notes of patients who had vulva cancer were retrospectively reviewed and relevant information were retrieved. Data was analyzed using SPSS version 25 software package. **Results:** Vulval carcinoma represented 3.6% of all gynaecological cancers. The mean age of patients was 49.7 years, with 41.7% of patients between the ages of 50 to < 65 years. Multiple sexual partners, (50%) and immunosuppression (37.5%) were the most identified risk factors. Vulval pain, vaginal discharge and pruritus vulvae were the major presenting complaints. All the patients had either stage 111 or IV disease at présentation. Excision biopsy (82.6%), partial vulvectomy (60.7%) and total vulvectomy (21.7%) were the commonly performed surgical procedures. Only 17.4% of cases had lymph node dissections. Surgery was mostly complicated by wound infection (47.6%) and wound breakdown (28.6%). Many (91.7%) of the patients were lost to follow up. **Conclusion:** Vulva cancer is a rare form of gynaecological cancer. Most patients with vulva cancer presented mainly with pain and pruritus vulvae. Late présentation and advanced-stage disease were the norm in this study. Most patients had inadequate evaluation and surgery due to lack of appropraite skill. There is an urgent need to train specialist in the art of radical vulvectomy.

Keywords : Vulva cancer, clinical presentation, surgical outcome, Port Harcourt.

Introduction

Vulva cancer is any malignancy involving the external genitalia of the female genital tract. Vulval carcinoma is not a common gynaecological cancer.[1] Cancer of the vulva is a relatively rare neoplasm accounting for approximately 4% of all gynaecological malignancies and less than 1% of all cancers in women. Vulva cancer is the 4th most common gynaecological cancer after cervical (62.5%), ovarian (17.0%) and endometrial cancers (6.8%).[2-6] Vulva cancer are commonly diagnosed in the 6th and 7th decades (51-60 years, 36%; 61-70yrs, 28.6%) of life.[7,8,9] Predominantly, it is a disease of elderly women, with the median age being 67 years, although it is now becoming common in younger age group.[10] Incidence of vulva cancer is significantly high in multiparous (90%) and postmenopausal (65%) women.[11]

The increased age is itself a high risk factor. Other risk factors for vulval carcinoma include smoking, young age at first sexual intercourse, history of multiple sexual partners, history of sexually transmitted infection (STI), presence of genital warts, immunosuppression and history of chronic vulval inflammation or Lichen sclerosis.[11] Human Papilloma virus (HPV) infection, immunosuppression and advanced age are the strongest risk factors identified for vulva neoplasms.[12,13]

The most prominent presenting symptom of vulva cancer is localized pruritus. Other common symptoms are a vulval mass with endophytic or exophytic lesions ranging from a small vulval swelling to massive vulval masses with ulcerations, bleeding, local pain, surface drainage from the tumour, vaginal discharge or urinary tract symptoms.[9,14,15]

Squamous cell carcinoma is the most common histological variant of vulva cancer. Other less common histological subtypes are malignant melanoma, Bartholin's gland carcinoma, basal cell carcinoma, soft tissue sarcoma and adenocarcinomas.[6,16] Strong association between HPV

infection and the later development of vulval carcinoma has been identified.[6,12] Labia majora was the predominant site of disease in 80%. Labia minor in 14.3% and clitoris in 5.7%.[10]

The staging of vulva cancer is popularly done according to the International Federation of Gynaecology and Obstetrics (FIGO).[17] According to the disease's stage, 5-year survival rates range from 86% for early-stage disease (FIGO stage 1) to 19% for metastatic disease (FIGO stage IVB); and the life-time risk of developing vulva carcinoma is 0.3%.[18]

Lymphatics is the primary route of spread while haematogenous spread is associated with sarcomas and large tumours. The strict adherence to FIGO staging is important for disease prognostication and treatment outcome therefore, every vulval lesion must be staged and biopsied for histological diagnosis.[6,19] Lymph node positivity is an independent bad prognostic factor.[17,20]

The treatment depends on the disease histology, stage and patient's performance status, and treatment is either surgical, chemotherapy, radiotherapy, palliative or multimodality care.[21] Surgery which is the mainstay of treatment for vulva cancer has evolved from extensive radical vulvectomy with dire post-operative complications to less radical surgery with better quality of life outcome. Lymphadenectomy is an integral part of radical vulvectomy as lymph node posivity is a strong prognostic factor. All centrally placed vulva tumours require bilateral lymph node dissection using seperate incisions on the groin. Commonly, employed surgical modalities include hemivulvectomy, excision biopsy and wide local excision especially in early stage disease. Unfortunately, majority of patients in developing countries for several reasons (social stigma, low socioeconomic status, illiteracy) present in advanced stage disease thus requiring a multidisplinary, multimodality treatment approach however with poor outcome.[22-25]

Objectives Aims

1. To determine the prevalence and sociodemographic pattern of vulva cancer at the University of Port Harcourt Teaching Hospital (UPTH).
2. To evaluate the risk factors, clinical profiles, treatment modalities, treatment outcome and histological pattern of the disease.

Materials and methods

Case notes of patients who had vulva cancer at the University of Port Harcourt Teaching Hospital, over a 10-year period between 1st January 2010 and 31st December 2019 were retrospectively reviewed. Gynaecological admission register and histopathology Department records were reviewed and the case notes of patients with vulva cancer were obtained from the Medical records Department and studied. Permission were obtained from the Heads of department of medical records, gynaecology and histopathological departments for the use of hospital records.

During the 10-year period, there were 729 gynaecological cancers and 26 were vulva cancers. Two case notes were not retrieved, hence 24 (92.4%) cases were analyzed. Information obtained were age, menopausal status, parity, clinical features, stage of the disease, treatment modality and outcome, treatment complications and histological types. Data was analyzed using SPSS version 25 software package. Results were presented in percentages and simple frequency tables.

Results

During the 10-year period under review, there were 729 gynaecological cancers admitted and vulval carcinoma accounted for 26 cases giving a prevalence of vulval carcinoma

of 3.6% of gynaecological cancers. However, only twenty-four (92.3%) case file were available for analysis.

The socio-demographic characteristics of patients are as shown in table 1. Majority of these patients were 50 years and above and those between the ages of 50 to < 65 years had the highest incidence of vulva cancer (41.7%). Most of the patients were multiparous 14 (58.3%). Nineteen (79.2%%) patients had either primary or no formal level of education. Most of the patients with vulva cancer were postmenopausal 17 (70.8%).

The most common risk factors for vulva cancer, as shown in table 2 were multiple sexual partners 12 (50%), immunosuppressive states 9 (37.5%) and history of sexually transmitted infections (STI) 6 (25%).

Pain and vaginal discharge were the most common presenting symptoms in majority of the patients 18 (75%) respectively. Other clinical presentations were pruritus 17 (70.8%), ulceration/bleeding 14 (58.3%) and vulva mass 8(33.3%).

Table 3 shows the various histological types of vulva cancer. Majority of the patients had Squamous cell carcinoma 19 (79.2%); others were basal cell carcinoma 3 (20.8%) and Adenocarcinoma 1 (4.2%).

All the patients had vulva cancer involving the labia majora 24 (100%), as well as occurring in the labia minora 14 (58.3%) while the clitoris was involved in five (20.8%) cases as shown in table 4. Twenty-three (95.8%) patients presented in advanced stages of the disease (stages 3 and 4).

In table 5, majority of the patients with vulva cancer had excision biopsy 19 (82.6%), following which they had partial vulvectomy 14 (60.7%) or total vulvectomy 5 (21.7%) while 4 (17.4%) had wide local excision only. Only 4 (17.4%) patients had lymph node dissections

during surgery. One patient declined surgery. Critical evaluation of the surgical notes revealed that most (82.6%) of the patients who would have benefitted from lymph node sampling or therapeutic lymphadenectomy did not have such intervention probably due to lack of skill and attention was not paid to tumour free margins during surgery.

Post-operative wound infection observed in 10 (47.6%) patients was the most common post-operative complication followed by wound breakdown noted in 6 (25.0%) patients. Most of the patients were lost to follow-up. Only 2 (8.3%) patients had some form of follow-up care between 6 months to 1 year post operatively while the rest were lost to follow up post operatively.

Discussion

Vulva cancers are relatively rare in our environments and even worldwide.[2,7,13,18] Independent reports from Nigeria, United Kingdom and the United States have shown vulva cancers to be rare.[2,26] It is reported that 60% of vulva cancers occur in developing countries.[2] The increase in the incidence and prevalence of this cancer in our environment is basically because of human papilloma virus (HPV) which is implicated as a risk factor for vulva cancers, especially the squamous cell carcinoma.[17] The increase is also likely due to increased health awareness among the population with the increasing use of hospital services. In Nigeria, cancer of the cervix had previously been shown to be the most common gynecological cancer, with cancers of the vagina and the vulva being the least common.[2] In Zaria, a prevalence of vulva cancer of 2.6% was reported, Lagos 3-5%, Ibadan 1.3% and in the USA 0.6%.[6,12,13,20] Vulva cancer contributes to about 3.6% of all gynaecological malignances in this study done in UPTH. The world wide range of vulva cancer was 0.3/100,000 women in Asia to 1.6/100,000 in North America and Europe.[6]

The average age at presentation in this study was 49.7yrs, a similar finding was reported from Ibadan and Lagos.[6,20] The age at presentation in this study, is relatively early compared to what is obtainable in developed countries and previous study, where majority presented in the fifth and sixth decades of life.[4,6,20,26,27]

Studies have revealed that 40% of vulva cancers in the USA are HPV related.[6,20] Most likely the percentages will be higher in developing countries because of the prevalence and incidence of HIV, which are relatively higher as we have seen in cases of carcinoma of the cervix.[21,28] The advent of human immunodeficiency virus (HIV)/acquired immune deficiency syndrome (AIDS), which compromises the immunity of these patients, makes the HPV more virulent.[17] The risk factors predominant in our setting: immunosuppression, multiple sexual partners, history of sexually transmitted diseases, genital wart and early age at first intercourse. Other risk factors for carcinoma of the vulva include chronic inflammatory diseases of the vulva such as granulomatous infection of the vulva, prior cervical or vaginal cancers from the concept of field carcinogenesis.

Squamous cell carcinoma (SCC), was the most common histological type seen in this study, accounting for 79.2%. Squamous cell carcinoma has previously been reported to be the most common malignant tumour of the vulva by a previous study from Nigeria,[4] and several reports from the developed nations,[13,26] sometimes constituting as high as 95% of the cases. In this study, ~~3~~ three cases of basal cell carcinoma were seen, constituting 16.6% of vulva cancer while adenocarcinoma was reported in 4.2% of cases.

All the cases of vulva cancer in this study were diagnosed at advanced stages of the disease (3 and 4). The population of patients who presentated at stage 3 was 61.4%. None was diagnosed at an early stage. This is worrisome considering the fact that the external genitalia is

easily accessible but most patients will not seek help until the condition becomes unbearable. Another reason for such late présentation may be because some patients may find it embarassing discussing or exposing their genitalia to be examined due to cultural beliefs and barriers. Similar finding was reported by Babarinsa[4] et al, who noted that most patients with vulva cancer tend to present late and to default during treatment, thus more likely to be diagnosed with late-stage disease. Pruritus vulvae which featured prominently in this study as a very common complaint in patients with vulva cancer should not be treated with levity especially in post menopausal women as it could be a pointer to premalignant or invasive carcinoma of the vulva. General practitioners should have a high index of suspicion when elderly women present with pruritus vulvae and refer such cases to the specialist rather than wasting precious time treating such patients for recurrent candidiasis.

With early detection, vulva cancer is highly curable. When lymph nodes are not involved, the five year survival rate is slightly higher than 90%.[25] Unfortunately, most patients in this study who would have benefitted from lymph node sampling or therapeutic lymphadenectomy did not have such intervention done probably due to lack of skill condering the fact all the patients presentated with late- stage disease. Surgical resection margins were not considered at the time of surgery and these patients stand a high risk of having recurrent disease. Another worrisome finding is that most of these patients who had inadequate surgical intervention should have benefitted from additional treatment with chemoradiation. This may have been hindered by the fact that radiation therapy are not readily available with pausity of knowledge regarding it's role in the management of advanced vulva malignancy. Even in developed nations, the management of vulva carcinoma is hampered by the fact that diagnosis is delayed in most cases and by the choice of the proper surgical procedures.[19] The greatest

difficulty in surgical management is with primary wound closure and healing, and wound breakdown and sepsis occur commonly.[16,19] Similar complications were seen in this study. There were no recorded deaths within the study period, but this is difficult to conclude since majority of the patients were lost to follow-up.

Most patients defaulted follow-up as seen in this study, only ~~2~~ two of the patients complied with follow-up but for a brief period. Poor follow-up due to several factors like long travelling distance, low socioeconomic status and elderly age is not unusual.[22,25]

Conclusion

Vulval cancinoma is still a rare genital tract malignancy but the mean age at présentation seems to be reducing. Late présentation and late stage disease are commonly associated with vulva malignancy. Most patients had inadequate surgical evaluation and treatment. Elderly patients with pruritus vulvae should be properly evaluated for the likelihood of invasive cancer. There is need to train specialist in the management of vulva cancers.

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Table 1. Socio-demographic pattern of patients with invasive vulvar cancer

Variables	Frequency (n =24)	Percentage (%)
Age (years)		
30 - <50	6	25.0
50 - <65	10	41.7
≥ 65	8	33.3
Mean ±S.D	49 ± 0.776	
Parity		
Nullipara	2	8.3
Multiparity	14	58.3
Grand Multiparity	8	33.3
Level Education		
No Formal Education	12	50.0
Primary	7	29.2
Secondary	4	16.7
Tertiary	1	4.2
Occupation		

Trader	10	41.7
Farmer	6	25.0
Housewife	5	20.8
Business	2	8.3
Civil Servant	1	4.2
Menopausal Status		
Premenopausal	5	20.8
Peri-menopausal	2	8.3
Post-menopausal	17	70.8

Table 2. Risk Factors and Clinical features of Patients

Variables	Number(s)	Percentage (%)
RISK FACTORS		
MSP	12	50.0
Immunosuppression (HIV)	9	37.5
History of STIs	6	25.0
Genital Wart	3	12.5
Early coitarche	1	4.2
Smoking	1	4.2
CLINICAL FEATURES		
Pain	18	75.0
Vaginal Discharge	18	75.0
Pruritus	17	70.8

Ulceration/bleeding	14	58.3
Vulvar Mass	8	33.3

MSP --- multiple sexual partners ; STIs --- sexually transmitted infections

Table 3. Histological Types of Vulva Cancer

Variables	Frequency (n =24)	Percentage (%)
Squamous Cell Carcinoma	19	79.2
Basal Cell Carcinoma	3	16.6
Adenocarcinoma	1	4.2

Table 4. Site and stage a presentation

Variables	Number	Percentage (%)
SITE		
Labia majora	24	100.0
Labia minora	14	58.3
Clitoris	5	20.8
Other sites	5	20.8
STAGE AT PRESENTATION		
I	0	0.0
II	0	0.0

111	18	78.3
IV	5	21.7

~~TABLE 5:~~ **Table 5**. Type of Surgery, Surgery Complication and Follow-up

Variables	Number	Percentage (%)
TYPE OF SURGERY		
Excision biopsy	19	82.6
Partial vulvectomy	14	60.7
Total vulvectomy	5	21.7
Wide local excision	4	17.4
Lymph node dissection	4	17.4
SURGERY COMPLICATIONS		
Wound Infection	10	47.6
Wound Breakdown	6	28.6
None	5	23.8
FOLLOW-UP		
Yes	2	8.3