Original Article

Carcinoma of Vulva in Nepalese Women Managed at B. P. Koirala Memorial Cancer Hospital

Jitendra Pariyar[1], Binuma Shrestha[2], Bijaya Chandra Acharya[2], Suresh Shrestha[3], Jaya Shrestha[4], Sadina Shrestha[5]

[1]*Gynecologic Oncology Unit, Civil Service Hospital, Kathmandu, Nepal*

[2]*Gyneoncology Unit, Department of Surgical Oncology, B.P. Koirala Memorial Cancer Hospital, Bharatpur, Chitwan, Nepal*

[3] *Department of Radiation Oncology, B.P. Koirala Memorial Cancer Hospital, Bharatpur, Chitwan, Nepal*

[4]*Department of Medical Oncology, B.P. Koirala Memorial Cancer Hospital, Bharatpur, Chitwan, Nepal*

[5]*Department of Pathology, B.P. Koirala Memorial Cancer Hospital, Bharatpur, Chitwan, Nepal*

ABSTRACT

Introduction: This study aims to analyze the clinical presentation and management outcomes carcinoma of vulva managed B. P. Koirala Memorial Cancer Hospital.

Methods: A descriptive study was conducted of all carcinoma of vulva cases managed at B. P. Koirala Memorial Cancer Hospital from 1999 to 2009. The case record of all women diagnosed to have carcinoma of vulva were retrieved and socio-demographic characteristics, clinical presentations, histological type, treatment modalities and outcome were obtained and analyzed.

Results: There were 5152 gynecological malignancies and vulvar cancer accounted for 87, giving a prevalence of 1.7%. The ages ranged from 17 to 86 years (mean of 48.6 years). Parity was 0-10. Vulva wound and pruritus were the most frequent clinical features with presentations in stage I -8%, stage II- 28%, stage III – 52 % and stage IV -12%. Squamous cell carcinoma (93%) predominated and 62% were grade I. Among the 87 cases, 32% were treated primarily with surgery, 34% primarily with concurrent chemo-radiation and 28% with combined modality. Clinical follow-up of one to five years showed that 26 (30%) cases had local recurrence and 22 (25%) died of disease.

Conclusion: Carcinoma of the vulva is a rare gynecological malignancy in Nepal. Surgery and radiotherapy remain to be the mainstay of treatment. Delayed presentation still results in greater morbidity and mortality rates.

Keywords: carcinoma of vulva, chemotherapy, groin node dissection, radiation therapy, radical vulvectomy

Introduction

Female genital tract cancers are very common among Nepalese women. Hospital based data compiled by B. P. Koirala Memorial Cancer Hospital, the national cancer center of Nepal reveals that cervical cancer is the commonest followed by ovarian and endometrial cancers. Cancer of vulva and vagina are less common.[1] Globally also carcinoma of vulva is noted as a rare tumor accounting for only 0.3% of all cancers in women and 3–5% of all female genital tract malignancies, with an estimated incidence of 1–2/100,000 women.[2-4]

Carcinoma of vulva occurs predominantly in elderly women although recently, an increased incidence has been observed among women under 50 years.[5] In older women, the vulvar cancer is unrelated to human

Correspondence
Dr. Jitendra Pariyar, *Gynecologic Oncology Unit, Civil Service Hospital, Kathmandu, Nepal, email: jipariyar@yahoo.com, phone: 9855060371.*

papillomavirus infection but in younger patients the disease is associated with HPV.[6,7] Carcinoma of the vulva may arise from the skin, subcutaneous tissue and glandular elements of the vulva, thus, early diagnosis can be made only by physical examination and tissue biopsy. However, late presentation is not uncommom where the 5-year survival rates for stage III and IV diseases according to the International Federation of Gynecology and Obstetrics (FIGO) system is 43.2% and 13% respectively.[8]

Surgery, historically, radical vulvectomy with bilateral dissection of the inguinal groin nodes and more recently, radical local excision, with inguinal lymph node dissection has been the mainstay of treatment of vulvar cancer. Over the past three decades surgeons are opting for conservative surgery with combination of radiation and chemotherapy either neoadjuvant or adjuvant treatment to reduce morbidity while improving survival.[9]

In Nepal, very few studies have looked at the management outcomes for vulva cancer. This study tired to analyze the clinical presentation and management outcomes carcinoma of vulva managed B. P. Koirala Memorial Cancer Hospital.

Methods

This review was conducted at B.P. Koirala Memorial Cancer Hospital, Chitwan, Nepal from January 1999 to December 2009. The case records of all carcinoma of vulva cases during the study period were analyzed regarding their identification, illness history, clinical examination, investigations, treatment and follow-up. The total number of gynecological malignancies over the same period was also obtained. Patients without histology proof of vulvar carcinoma were excluded from the study.

Surgical treatment consisted of either radical vulvectomy and bilateral groin lymphadenectomy with use of the three-separate incision technique, or radical vulvectomy and unilateral groin lymphadenectomy, or radical vulvectomy without groin lymphadenectomy, or simple vulvectomy or wide local excision. The groin lymph node dissection consisted of removal of all superficial and deep lymphatic fatty tissue around the femoral artery, femoral vein, and great saphenous vein in the femoral triangle. For patients who received radiotherapy to the whole pelvis including vulva, it consisted of external beam radiation therapy employing 6 MV energy using linear accelerator or irradiation by Cobalt 60 machine delivering 4,600–5,000 cGy in daily fractions of 180 – 200 cGy with concurrent chemotherapy cisplatin 40mg/m^2/wkly. Adjuvant chemotherapy consisted of cisplatin 70 mg/m^2 (day 1) and 5-fluorouracil (5-FU) 500 mg/m^2 (days 1–5) or paclitaxol 175mg/m^2 and carboplatin AUC 6-7.

The main outcomes were measured in terms of age, duration, parity, histopathology, treatment, follow-up and mortality associated with this disease. Statistical analysis was done using descriptive statistics: mean, range and standard deviation.

Results

The hospital record of B. P. Koirala Memorial cancer hospital from January 1999 to December 2009 showed that there were 5152 gynecological malignancies among which vulvar cancer accounted for 87, giving a prevalence of 1.7%. The mean age of the cases at the time of diagnosis was 48.6 years; the youngest patient was 17 years and the oldest 86 years (Table 1).

All the cases were married. Parity was recorded in all cases: two (2.3%) were nulliparous and 11 (12.7%) had at least one child at the time of diagnosis. The mean parity of ranged from 0 to 10. Two young patients conceived with vulvar cancer, had vaginal delivery and were diagnosed and treated during their postnatal period.

Table 1: Demographic characteristics of participants (N=87)

Characteristics	Frequency (n)	Percentage (%)
Age		
≤ 40 years	26	30
40 - 50 years	30	34.5
> 50 years	31	35.5
Parity		
0	2	2.3
1	11	12.7
2	21	24.0
3	25	28.8
≥ 4	28	32.2

All the cases with vulvar carcinoma were symptomatic at the time of diagnosis. The usual presenting symptoms were vulvar lump or wound; itching and discomfort in vulva and discharge and abnormal bleeding per vaginum. Most of the cases had more than one symptom. The time interval from the beginning of symptoms until seeking medical attention recorded ranged from few weeks to several months.

The commonest histological type of vulvar carcinoma noted among the 87 cases that presented with vulvar carcinoma was squamous cell carcinoma, seen in 81

(93%) of the patients. There were three (3.5%) cases of Bartholin's gland adenocarcinoma and two (2.3%) cases of vulvar sarcoma of the vulva and one case (1.2%) of malignant melanoma of vulva (Table 2).

Table 2: Histopathological patterns of carcinoma of vulva of the participants (N=87)

Histopathology	Frequency (n)	Percentage (%)
Squamous cell carcinoma (n=81)	81	93.0
Grade I	54	62.0
Grade II	23	26.5
Grade III	10	11.5
Adenocarcinoma of Bartholin's gland	3	3.5
Sarcoma	2	2.3
Malignant melanoma	1	1.2

Seven cases (8%) had Stage I, 24 (28%) had Stage II, 45 (52%) had Stage III and 11 (12%) had Stage IV disease at the time of presentation (Table 3).

Table 3: Distribution of participants by Stage of carcinoma of vulva (N=87)

Stage	Frequency (n)	Percentage (%)
Stage I	7	8
Stage II	24	28
Stage III	45	52
Stage IV	11	12

Among the 87 cases, 32% were treated primarily with surgery. Regarding surgical procedures, radical vulvectomy and bilateral groin lymphadenectomy with use of the three-separate incision technique in 27 cases, radical local excision and groin lymphadenectomy in 18 cases and wide local excision in 12 cases were carried out. Radiation therapy with or without concurrent chemotherapy was delivered to 30 (34%) cases who presented with advanced staged disease or who were unfit, or unwilling undergo any surgical procedure. Thirty (34%) of the cases were treated with combined modalities, radiation with or without chemotherapy as an adjuvant treatment to surgery (Table 4). Clinical follow-up of one to five years showed that 30% (26) cases had local recurrence and 25% (22) died of disease.

Table 4: Treatment done of carcinoma of vulva of the participants (N=87)

Treatment Modality	Frequency (n)	Percentage (%)
Surgery Radical vulvectomy and bilateral groin lymphadenectomy -27 Radical local excision and unilateral groin lymphadenectomy- 18 Wide local excision - 12	57	65.5
Radiation Therapy with or without chemotherapy	30	34
Combined modality (Surgery + Chemo-radiation)	30	34

Discussion

In women, gynecological malignancies, cancer of cervix causes a significant amount of morbidity and mortality. Cancer of vulva though rare as noted in the present study, also contributes to a significant morbidity. Due to its anatomical location near the urinary and genital tract, the woman's quality of life may be affected.

In the present study, a significant number of cases (30%) were young aged ≤ 40 years and another 34.5% of the cases were between 40 to 50 years of age and mean age 48.6 years was similar to the earlier study from Jamaica.[10] Another study also observes an increased incidence vulvar cancer among women under 50 years and better survival outcome.[5] However, this finding does not match with the previous studies that demonstrated that the most common age group of patients with vulvar carcinoma is 60–69.9 years.[11,12]

The most common presenting symptoms of vulvar carcinoma are vulvar mass, ulcer, and itching, which were noted in the present study as well another study from Israel.[13] Bleeding was reported as rare symptom as in Way's series.[11]

Vulva being an external genital organ, pathology like cancer could be suspected by the presence of abnormal growth and early diagnosis could be made only by physical examination and tissue biopsy. Despite this fact most of the cases delay in seeking medical help and are diagnosed in advanced stage. In the present study majority of the cases 45 (52%) and 11 (12%) had stage III and stage IV disease respectively which is contrast to the findings of early stage disease reported in other studies like that done by Piura B et al.[13] Vulvar cancer tends to have very slow progression which could be the reason behind to seeking medical assistance late. Sometimes, delay may be caused by the patient's health care provider

who fails to diagnose the disease because of failure to examine the patient as reported in a study done by Gyenwali D *et al* where 70% cases of cervical cancer patients did not have examination by speculum.[14]

Vulvar cancer in most cases is local or loco-regional at the time of diagnosis. Definitive therapy is associated with high cure rates. Radical vulvectomy and bilateral groin lymphadenectomy has been the established mainstay of treatment in invasive vulvar carcinoma and radiotherapy may be used preoperatively to reduce the size of the tumor, or it may be used as primary treatment, or it may be used as postoperative adjuvant therapy.[15] In the present study also majority of the cases underwent radical surgery in the form of radical vulvectomy and bilateral groin lymphadenectomy , radical local excision and unilateral groin lymphadenectomy or simply wide local excision. Radiation therapy with concurrent chemotherapy was used more often to reduce surgical morbidity and physical debility. Neoadjuvant chemothrapy is being explored in place of mutilating surgery and application of high dose radiation therapy to achieve better treatment results with less side-effect. However, due to late presentation and bulky disease even in early stage, higher recurrence and mortality were observed in the present study with surgery, radiation therapy and chemotherapy.

Conclusion

Carcinoma of the vulva is a rare gynecological malignancy in Nepal. Though known to be a disease of elderly women, an increased incidence was observed among younger women. Surgery and radiotherapy remain to be the mainstay of treatment. In Nepal's context, delayed presentation result into greater morbidity and mortality rates. Thus, focus should be made in early diagnosis by creating awareness among women to seek early medical care and among health care providers to perform clinical examination.

References

1. Dhakal HP, Pradhan M. Histological pattern of gynecological cancer. J Nepal Med Assoc.2009; 48(4):176.
2. Monaghan JM. The management of carcinoma of the vulva. In Sheperd JH, Monaghan JM (eds): Clinical Gynecologic Oncology. 2nd ed. London: Blackwell, 1990;140–167.
3. Hoffman MS, Cavanagh D. Malignancies of the vulva. In Rock JA, Thompson JD (eds): Te Linde's Operative Gynecology.8th ed. Philadelphia: Lippincott-Raven, 1997;1331–1383.
4. Mack TM, Cozen W, Quinn MA. Epidemiology of cancer of the endometrium, ovary, vulva and vagina. In Coppleson M, Monaghan JM, Morrow CP, Tattersall MHN (eds): Gynecologic Oncology: Fundamental Principles and Clinical Practice. 2nd ed. London: Churchill Livingstone, 1992; 31–54.
5. Messing MJ, Gallup DG. Carcinoma of the vulva in young women. Obstet Gynecol. 1995; *86*: 51-4.
6. Wilkinson EJ. Premalignant and malignant tumors of the vulva. In: Kurman RJ, editor. Blaustein's pathology of the female genital tract. 5th ed. New York: Springer Verlag; 2002. 99-149.
7. Shea CR, Stevens A, Dalziel KL, Robboy SJ. The vulva: cysts, neoplasms and related lesions. In: Robboy SJ, Anderson MC, Russell P, editors. Pathology of the female reproductive tract. London: Churchill Livingstone; 2002. 35-74.
8. Beller U, Quinn MA, Benedet JL, Creasman WT, Ngan HY, Maisonneuve P. Carcinoma of the vulva. FIGO 26th Annual Report on the Results of Treatment in Gynecological Cancer. Int J Gynaecol Obstet 2006; 95 Suppl 1:S7-27.
9. Afandy Ahmad El, Soliman H, Sherbiny Magdy El, Elkasem Hatem A. Groin recurrence in patients with early vulvar cancer following superficial inguinal node dissection. Journal of the Egyptian National Cancer Institute.2013; 25.(3): 121-24.
10. Hay DM, Cole FM. Primary invasive carcinoma of the vulva in Jamaica. J Obstet Gynaecol Brit Commwlth.1969; 76:821-30.
11. Way S.Malignant Disease of the Vulva. London: Churchill Livingstone, 1982.
12. Rothschild F: 1912 Inaugural dissertation, Freiburg. Quoted by Taussig FJ: Diseases of the Vulva. New York: Appleton, 1931, 142.
13. Piura B, Rabinovich A, Cohen Y, Friger M, Glezerman M. Squamous cell carcinoma of the vulva in the south of Israel: a study of 50 cases. Journal of surgical oncology.1998; 67(3), 174-181.
14. Gyenwali D, Khanal G, Paudel R, Amatya A, Pariyar J, Onta S. R. Estimates of delays in diagnosis of cervical cancer in Nepal. BMC women's health.2014;14(1): 29.
15. Krupp PJ Jr: Invasive tumors of vulva: Clinical features, staging and management. In Coppleson M, Monaghan JM, Morrow CP, Tattersall MHN (eds): Gynecologic Oncology: Fundamental Principles and Clinical Practice. 2nd ed. London: Churchill Livingstone, 1992;479–491.