J. of Obst. & Gyn. of India Vol. 51, No. 4 : July/August 2001 Pg 167-171

# Vulval Dystrophy: An Experience in a Regional Cancer Center

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**Summary**

A review of fifty-five cases of vulval dystrophy is presented. The majority of cases were seen in the fifth and sixth decade. The most common and distressing symptom was pruritis vulva seen in 39 (71%) women. Sixty four percent of the women were postmenopausal. Labia majora was the commonest site of involvement seen in 47 (85.45%). The most frequent histologic variant was hyperplastic dystrophy. Topical corticosteroid was the commonest medical therapy instituted. Surgery was done for resistant, intractable cases and cases with moderate to severe atypia. Simple vulvectomy was the commonest surgical procedure performed. Subjective as well as objective response was better in cases where lichen sclerosis was the histological variant. Progression to malignancy was not noted in any case during follow up. In conclusion, histological diagnosis is essential at the outset. Medical therapy is the mainstay of treatment. Long term surveillance is recommended to know the potential for malignancy.

## Introduction

There are several lesions of the female genital tract, which at one time or another were presumed to be premalignant but have not endured the test of time. One such example is vulval dystrophy. At one time it was so highly regarded as to have resulted in what presently would be considered excessive surgery. Currently vulvectomy is no longer recommended as primary therapy. There is uncertainty about vulval dystrophy and its relation to the development of carcinoma. The risk of developing carcinoma in vulval dystrophy varies from 1% to as high as 25% (Langly et al 1951, Woodruff and Beans 1963, Jeffcoate TNA 1966, Kaufman et al 1974, Hart WR et al 1975, Meyrick-Thomas et al 1988,). This study reviews the clinico-pathological correlation and the current management options for vulval dystrophy.

## Patients and Methods

This is a study of fifty-five cases of vulval dystrophy carried out over a period of eighteen years from 1980 to 1998, at The Gujarat Cancer and Research Institute, Ahmedabad. All patients with white lesions of vulva were enrolled in the study. Detailed history was elicited. Thorough clinical examination was done and patients were subjected to multiple vulval biopsy. Once biopsy reports were available they were grouped to the classification outlined in Table 1. (ISSVD 1976) They were treated with local application of ointments and followed up regularly. Surgery was advised only if there was no improvement with topical application, symptoms were intractable and in patients with varying degree of atypia. During follow up the subjective and objective response were documented. Detailed local examination was done to assess the regression of lesion.

Repeat vulval biopsy was performed to know the potential for malignancy.

**Table I**
**Classification of Vulval Dystrophy**

| Microscopic features. |
|---|
| 1. Hyperplastic dystrophy |
| A. With atypia |
| B. Without atypia |
| 2. Lichen scleros |
| 3. Mixed dystrophy |
| A. With atypia |
| B. Without atypia |

### Analysis and Discussion

Fifty-five patients with vulval dystrophy were evaluated and grouped according to classification in Table I. (ISSVD 1976).

### Clinical features

Most of our patients were in the age group of 41 to 60 years. (Table II) The age ranged from 8 to 85 years and median was 54 years. We had one child of eight years with lichen sclerosis. Lichen sclerosis has been noted in patients as young as 6 months of age. A recent series described 15 cases in girls. (Jones 1991).

The symptomatology of vulval dystrophy is depicted in Table III. The commonest symptom was itching vulva, which constitutes 70% of the cases. Next commonest symptom is leucorrhoea seen in 38% of women and only 33% of patients presented with white lesion of vulva. Dysuria is seen in 27% of patients. Five patients presented with bleeding per vaginum. Three patients were asymptomatic. Each of the patients may have more than one symptom. Heart et al (1975) also noted that pruritis as the most common symptom occurring in 68% of cases. The duration of symptom in our series varies from 7 days to 15 years. The median was one year. More than 50% of the women were postmenopausal. This finding was similar to others. (Hart et al 1975) Only 33% of them were premenopausal.

**Table III**
**Spectrum of Symptoms**

| Symptoms | Number of patients | Percentage |
|---|---|---|
| Itching vulva | 39 | 70.91 |
| White lesion | 18 | 32.73 |
| Leucorrhoea | 21 | 38.18 |
| Dysuria | 15 | 27.27 |
| Bleeding PV | 5 | 9.09 |
| Asymptomatic | 3 | 5.45 |
| Others | 7 | 14.55 |

The specific anatomic sites of involvement are listed in Table IV. Majority of our patients had multiple lesions and more than one anatomic site of involement. In the order of decreasing frequency, they included the labia majora (85%), labia minora (71%), fourchette (27%), paraurethra (25%), clitoral (22%), anus (9%) and perianal (5%). Extra-genital cutaneous lesion was identified on left thigh in one woman. Whereas Hart et al (1975) in his series found labia minora (68%) is the commonest anatomic site involved. The importance of noting the specific anatomic site of involvement lies in the fact that the perianal area was the most likely to show malignancy. (Lavery 1984)

**Table IV**
**Specific Site of Involvement**

| Site | Number of patients | Percentage |
|---|---|---|
| Labia Majora | 47 | 85.45 |
| Labia minora | 39 | 70.91 |
| Fourchette | 15 | 27.27 |
| Paraurethral | 14 | 25.45 |
| Clitoral | 12 | 21.82 |
| Anus | 5 | 7.09 |
| Perianal | 3 | 5.45 |
| Others | 1 | 1.82 |

### Pathologic findings

As shown in Table II hyperplastic dystrophy is the most common histologic variant of vulval dystrophy, which constitutes 41.81%(23) of cases. In six of these

**Table II**
**Age-Wise Distribution of Vulval Dystrophy**

| Age (Years) | Hyperplastic dystrophy No atypia | atypia | Lichen sclerosis | Mixed Dystrophy No atypia | atypia | Unclassified | Total |
|---|---|---|---|---|---|---|---|
| <20 | - | - | 1 | - | - | - | 1 |
| 21-40 | 4 | 2 | 7 | 1 | - | 1 | 14 |
| 41-60 | 9 | 4 | 9 | 3 | - | 5 | 25 |
| 61-80 | 4 | - | 3 | - | - | 1 | 7 |
| 81> | - | - | 1 | - | - | - | 1 |
| Total | 17 | 6 | 21 | 4 | - | 7 | 55 |

patients varying degree of atypia was found. Another 21 patients were diagnosed as having lichen sclerosis. In four patients mixed dystrophy was noted. In four patients who refused to have vulval biopsy and in three others where the histopathology was non-specific were grouped as unclassified. (Others) Kaufman et al (1974) in his series had one hundred and eighty cases of vulval dystrophy, 61 were hyperplastic dystrophy, 48 were lichen sclerosis, 18 were mixed dystrophy and one patient was unclassified. He noted varying degree of atypia in 6 cases of hyperplastic dystrophy and in 3 cases of mixed dystrophy. Squamous hyperplasia within the background of lichen sclerosis (mixed dystrophy) constitutes a distinct group at higher risk of developing invasive cancer and hence more liberal use of biopsy is recommended in such patients (Elliot 1988).

### Treatment

**Table V**
**Medical Therapy**

| Type of therapy | Number of patients | Percentage |
|---|---|---|
| Topical steroid | 42 | 76.36 |
| 5 Flurouracil | 6 | 10.91 |
| Testosterone | 1 | 1.82 |
| Only symptomatic | 6 | 10.91 |

Topical therapy is the mainstay of treatment. The various types of topical medical therapy instituted are listed in Table V. Corticosteroid cream application was the commonest medical therapy instituted (76%). 11% of the patients received 5 flurouracil. One patient received testosterone. 10% of patients received only symptomatic therapy for itching vulva. Four patients of vulval dystrophy with mild to moderate atypia who refused surgery were treated with topical 5 flurouracil. Mahmud et al (1992) reported symptomatic improvement and reduction of lesion size in 80% of lichen sclerosis and 89% of squamous hyperplasia in patients treated with testosterone and corticosteroid respectively.

**Table VI**
**Surgical Therapy**

| Type of surgery | Number of patients | Percentage |
|---|---|---|
| Simple vulvectomy | 4 | 7.27 |
| CO2 laser | 2 | 3.64 |
| Simple vulvectomy with Vaginal hysterectomy | 1 | 1.82 |

In our series five patients underwent simple vulvectomy, out of which one patient underwent concomitant vaginal hysterectomy for uterovaginal prolapse. Two patients underwent laser vapourisation of vulva. Two patients of hyperplastic dystrophy with atypia who did not respond to topical 5-flurouracil were taken up for simple vulvectomy. One of them underwent repeat surgey with removal of distal urethra for intractable symptoms. Three of our patients with lichen sclerosis who did not respond to topical steroid therapy were subjected to simple vulvectomy. Recurrence rate after surgery varies from 39-78% (Woodruff 1963, Jeffcoate 1966). Due to high incidence of recurrence, vulvectomy is contraindicated in the absence of significant atypia. Surgical therapy is advocated only when medical therapy fails or symptoms are intractable.

**Table VII**
**Associated Conditions**

| Associated condition | Number of patients | Percentage |
|---|---|---|
| Carcinoma cervix | 8 | 14.55 |
| Hypertension | 4 | 5.66 |
| Prolapse uterus | 4 | 5.66 |
| Ischemic heart disease | 3 | 3.77 |
| Choriocarcinoma | 1 | 1.82 |
| Carcinoma vault | 1 | 1.82 |
| Carcinoma endometrium | 1 | 1.82 |
| Behcet syndrome | 1 | 1.82 |
| Mullerian agenesis | 1 | 1.82 |

There are certain gynaecological and non-gynecological disorders associated with vulval dystrophy. (Table VII) Eleven cases (20%) had been treated for a malignant neoplasm before the diagnosis of vulval dystrophy was made. Eight cases had history of prior irradiation to the pelvic area. These tumors include eight cases of carcinoma cervix. There was one case of choriocarcinoma, carcinoma of vaginal vault and carcinoma endometrium.

### Response to therapy and follow-up information

Table VIII demonstrates the response to treatment. In 40 cases information was available. Subjective as well as objective response was assessed during follow up. 31 out of 40 cases showed partial subjectie response. Complete alleviation of symptoms was observed in 3 cases of lichen sclerosis. One interesting observation was that symptom relief was 100% and was rapid in cases of lichen sclerosis. Our observation was similar with Kaufman (1974). In about 7 cases no subjective response was seen. Partial regression of lesion was documented in 12 cases, fifty percent of these patients had lichensclerosis.

Table VIII
Length of Follow UP

| Duration of follow up | Hyperplastic dystrophy No atypia | Hyperplastic dystrophy Atypia | Lichen sclerosis | Mixed dystrophy No atypia | Mixed dystrophy Atypia | Others | Total |
|---|---|---|---|---|---|---|---|
| <1 | 13 | 5 | 18 | 4 | - | 6 | 46 |
| 1-2 | 3 | - | 1 | | - | 1 | 5 |
| 2-3 | 1 | - | 1 | - | - | - | 2 |
| 3-5 | - | - | - | - | - | - | - |
| 5-10 | - | - | 1 | - | - | - | 1 |
| 10-20 | - | 1 | - | - | - | - | 1 |

Table IX
Response to Therapy

| Type of lesion | Subjective response None | Subjective response Partial | Subjective response Complete | Objective response None | Objective response Partial | Objective response Complete |
|---|---|---|---|---|---|---|
| Hyperplastic dystrophy | | | | | | |
| Without atypia | 1 | 9 | - | 7 | 3 | - |
| With atypia | 1 | 4 | - | 4 | 1 | - |
| Lichen sclerosis | 1 | 13 | 3 | 11 | 6 | - |
| Mixed dystrophy | | | | | | |
| without atypia | 1 | 2 | - | 2 | 1 | |
| with atypia | - | - | - | - | - | - |
| Unclassified (Others) | 2 | 3 | - | 4 | 1 | - |

Follow up in vulval dystrophy will enable to determine the response to therapy and the risk of development of carcinoma. The length of follow up period ranged from one month to seventeen and half years. Here out of 55 cases 46 cases were followed for less than one year. Six cases followed upto one year. Five cases up to two years and two cases up to 3 years. The long-term follow up was seen in two cases. One was for 10 years, another for 17 and half years. During follow up none of our patients were found to have malignancy. In Kaufman (1974) series 46 cases were followed for less than one year, 36 patients were followed for 1 to 5 years and 26 cases observed for 5 to 19 years. Out of 110 only one was found to have invasive carcinoma of vulva.

One of the most controversial points regarding vulval dystrophy is that of its malignant potential. The predisposition of patients with vulval dystrophy to development of carcinoma can be reliably estimated by follow up studies of large group of patients. To date very few such series have been reported as highlighted in Tabe X. The risk varies from 1% to 25% according to different authors. In addition in several instances it is questionable whether lichen sclerosis actually preceeded the carcinoma or was found in association with it. (Barker and Gross 1962) However in our series we have not seen any patients developing carcinoma during their surveillance.

Countless vulvectomies have been performed on the basis of preventing the subsequent development of carcinoma previously. A finding of atypical epithelial activity on biopsy usually foreshadows the likely development of carcinoma. Currently vulvectomy is

Table X
Long term risk of development of carcinoma in vulval dystrophy

| Authors | Number of patients | Patients who subsequently Developed carcinoma of vulva(%) | Follow up time years |
|---|---|---|---|
| 1. Langly et al (1951) | 122 | 1 | 5 |
| 2. Barker and Gross (1962) | 42 | 2 | 7-10 |
| 3. Woodruff and Beans (1963) | 80 | 25 | 8 |
| 4. Jeffcoate (1966) | 216 | 0.9 | 3-45 |
| 5. Kauffman et al (1974) | 110 | 0.9 | 5-19 |
| 6. Hart et al (1975) | 92 | 1 | 12 |

reserved primarily for such findings on biopsy. Nevertheless it has to be recognized that vulvectomy for vulval dystrophy with atypia does not necessarily protect the patients from subsequent development of cancer as seen in the several series. (Jeffcoate 1966, Langly 1951, Woodruff and Beans 11963) Such happenings are accounted for the fact that cancer of the vulva is often a multifocal disease and the changes which precedes it are part of a widespread field change. Even though the affected skin is excised, adjacent area are liable to undergo similar change in the future. So even when vulvectomy is performed there remains a need to keep the patient under observation for an indefinite period of time. Following conservative medical regimen most of our patients remain asymptomatic. All patients however must be impressed with the necessity for regular, adequate follow up studies, even though they may remain free of symptoms. The overall risk of malignant progression may be small but surgical cure rates for early invasive cancers are excellent and early diagnosis allows use of modified approach to radical vulvectomy. (Gleeson 1995).

## Conclusion

- Multiple biopsy is indicated at the outset of the management of all women with vulval dystrophy. It aims to detect the presence of malignancy or atypical change, which may threaten cancer.
- Medical therapy is the mainstay of treatment of vulval dystrophy.
- Surgery is advocated only when medical therapy fails or patients having atypia, who are at risk of invasive cancer.
- Long term surveillance is recommended for all patients with vulval dystrophy even after vulvectomy.

## Acknowledgment

We thank the Director, The Gujarat Cancer and Research Institute, Ahmedabad for permitting us to take up and publishing this study. We would also like to thank Dr Nisha and Dr Vani Ramkumar for their help.

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