

# Diseases of the vulva- a clinicopathological study

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## Abstract

**Background:** Vulval lesions may present in a variety of ways ranging from asymptomatic to chronic disabling conditions. The differential diagnosis may include a wide range of diseases. This study was undertaken to find out the distribution and type of diseases affecting the vulva. **Materials and Methods:** All the vulval lesions encountered in the histopathology section for a duration of 3 years were included in this retrospective study. **Results:** A total number of 35 cases of vulval lesions were encountered in the study. The age group of patients ranged from 21 to 86 years. Majority of the women had itching or mass as a presenting complaint. Out of the 35 cases, 28 cases ( 80% ) were found to be benign and 7 cases ( 20 % ) were found to be malignant. Bartholin's cyst was the commonest benign lesion which was found in the reproductive age group. Squamous cell carcinoma was the next common lesion in our study which was seen in older age groups. **Conclusion:** The vulval lesions are one of the most neglected especially by the Indian women. Since the vulva can harbour lesions starting from a benign bartholin's cyst to aggressive malignancies, more importance should be given to the timely detection, diagnosis and appropriate treatment of these lesions.

**Key words:** vulva, squamous cell carcinoma, lymphangioma circumscriptum, lichen sclerosis

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## INTRODUCTION

The vulva, a small part of the external genital tract, is also a part of one of the the largest organs of the body namely the skin. For this reason most of its disorders are those of the skin modified only by site. In the last few years, interest in vulval diseases has greatly increased. Vulval lesions may present in a variety of ways ranging from asymptomatic to chronic disabling conditions. All of these conditions may present with itching, fissuring, bleeding or dyspareunia. The differential diagnosis may include a wide range of diseases. The diagnosis and management of chronic vulval disease requires attention

to several issues. Psychological factors are usually important, because women worry about malignancy, impaired sexuality and self image, fertility and sexually transmitted diseases. Hence, the histopathological study and the correct diagnosis is of utmost importance for the appropriate management of the patient. This study was undertaken to find out the distribution and type of diseases affecting the vulva

## MATERIALS AND METHODS

Tissue samples of all the vulval lesions which included biopsies, vulvectomy specimens and excision specimens received in the pathology department for a duration of 3.5 years from January 2009 to June 2012 were included in the study. The clinical details and HandE stained sections of all the cases were reviewed.

## RESULTS

A total number of 35 cases of vulval lesions were encountered in the study. The age group of patients ranged from 21 to 86 years. Majority of the women had itching or mass as a presenting complaint. Out of the 35 cases, 28 cases ( 80% ) were found to be benign and 7 cases ( 20 % ) were found to be malignant. Distribution

of all the lesions is shown in the table 1. Bartholin's cyst was the commonest benign lesion which was found in the reproductive age group. Size varied from < 1 cm to upto 4 cms. The most common lining epithelium encountered in our study was a transitional type of epithelium. Out of the 15 cases, 20% of cases were chronically infected. Squamous cell carcinoma was the next common lesion in our study which was seen in older age groups (45- 80 years). These women presented with ulcer or nodule in the vulva. (fig 1A). Four cases of invasive squamous cell carcinomas were seen out of which one showed in situ component and microinvasion (Fig 1B). One had associated Paget's disease. One case of lymphangioma circumscriptum was seen which presented as growth over genitalia (fig 1C). Histopathology showed numerous dilated lymphatics in the superficial and papillary dermis. (Fig 1D). One case of Pyoderma gangrenosum presented with painful ulcer in the genitalia since two weeks associated with fever, joint pain and ulcer in the mouth. Examination revealed a solitary ulcer with slough present over left labia majora extending to labia minora. Purulent discharge was also present. (fig 2A). Histopathology of Pyoderma gangrenosum, angiofibroma, and septal panniculitis is shown in fig 2B, 2C, 2D.

## DISCUSSION

Vulva can encompass a wide range of lesions. Till date, to the best of our knowledge, there are no studies done which include all the vulval lesions in one. However, individual case reports and case series of individual lesions have been reported. Among the benign lesions, Bartholin's cyst was the most common lesion seen in the younger (reproductive) age group between 20 – 40 years, which is consistent with the study done by Sosnik *et al* in which the mean age was found to be 39.4 years<sup>1</sup>. Bartholin cysts are formed when a Bartholin duct is blocked causing a fluid filled cyst to develop. These can grow from a size of a pea to the size of an egg or even larger at times and can be mistaken for edema.<sup>2</sup> Fibroepithelial polyp was the next common benign lesion followed by Lichen sclerosus et atrophicus. Lichen sclerosus (LS) is a complex chronic inflammatory skin disease with genetic, physiologic and environmental factors that influence the clinical phenotype and disease outcome. Vulvar lichen sclerosus is usually a clinical diagnosis. In the early stages of the disease the diagnosis can sometimes be difficult.<sup>3</sup> The classic histological features of LS are a thinned epidermis with hyperkeratosis, a wide band of homogenized collagen beneath the dermo-epidermal junction and a lymphocytic infiltrate beneath the homogenized area.<sup>4</sup> Two cases of Lichen sclerosus were seen in our study both presenting in post menopausal age group. In a study done by

Leighton and Langley, the mean age for Lichen sclerosus was found to be 57 years.<sup>5</sup> We also had cases of Lymphangioma circumscriptum, Pyoderma gangrenosum, Angiofibroma, Condyloma acuminata, Vulval keratosis, Septal panniculitis and Hemangioma of the vulva. All these lesions have been reported in the vulva in individual case reports<sup>(6,7)</sup>. Lymphangioma circumscriptum is a rare disorder with only 66 cases reported in English language literature till 2005.<sup>8</sup> It is a benign disorder of lymphatic channels that protrude above the surface of skin resembling blisters. It can occur as either a congenital abnormality as a developmental defect of lymphatics in deep and subcutaneous layers, or it can occur as a result of lymphatic obstruction of any cause, including chronic edema, tumor, radiation therapy and surgery.<sup>9</sup> The malignant lesions of vulva were more common in the elderly age group (61 – 80 years) and presented with growth in the vulval region. Echeverri *et al* have reported a similar age incidence<sup>10</sup>. Squamous cell carcinoma was the only malignancy in our study. Individual cases of Paget's disease, vulval intra epithelial neoplasia and adenocarcinoma metastasis to the vulva were also seen. Vulvar squamous cell carcinoma (SCC) accounts for approximately 3–5% of all gynaecological malignancies and 1% of all carcinomas in women, with an incidence rate of 1–2/100,000.<sup>11</sup> There are two different types of vulvar SCC with their own associated premalignant lesions. The most common type occurs in elderly women and leads to mostly differentiated keratinising SCC, in a background of lichen sclerosus (LS) and often differentiated vulvar intraepithelial neoplasia (VIN).<sup>12</sup> The second type of vulvar SCC consists of mainly non-keratinising carcinomas and primarily affects younger women.<sup>13</sup> Extramammary Paget's disease (EMPD) is a rare condition with only a few hundred cases reported in the world medical literature.<sup>14</sup> EMPD represents 6.5% of all cutaneous Paget's disease and affects predominantly patients between 50 to 80 years of age with peak incidence of 65 years.<sup>15,16</sup> The vulva remains the most frequently involved site with 65% of EMPD located in this area.<sup>17</sup>

## CONCLUSION

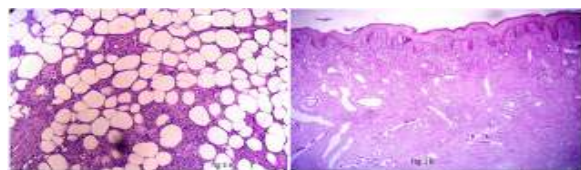
The vulval lesions are one of the most neglected especially by the Indian women. The relevant material regarding these lesions are scattered throughout the literature of various specialities including dermatology, gynaecology and pathology. Since the vulva can harbour lesions starting from a benign Bartholin's cyst to aggressive malignancies, more importance should be given to the timely detection, diagnosis and appropriate treatment of these lesions.

**Table 1: DISTRIBUTION OF LESIONS**

S NO	LESION	NO. OF CASES	%
1	BARTHOLIN'S CYST	15	45
2	SQUAMOUS CELL CARCINOMA	4	11.4
3	VULVAL INTRAEPITHELIAL NEOPLASIA	1	2.85
4	PAGET'S DISEASE	1	2.85
5	LICHEN SCLEROSUS	2	5
6	ADENOCARCINOMA METASTASES	1	2.85
7	PYODERMA GANGRENOSUM	1	2.85
8	LYMPHANGIOMA CIRCUMSCRIPTUM	1	2.85
9	HEMANGIOMA	2	5.71
10	CONDYLOMA ACUMINATUM	1	2.85
11	FIBROEPITHELIAL POLYP	3	8.57
12	ANGIOFIBROMA	1	2.85
13	VULVAL KERATOSIS	1	2.85
14	SEPTAL PANNICULITIS	1	2.85


**Figure 1(A) Figure 1(B)**

**Figure 1(C) Figure 1(D)**

**Figure 2(A) Figure 2(B)**

**Figure 3(A) Figure 3(B)**

#### Legends for figures

**Figure 1A:** Squamous cell carcinoma presenting as ulceroproliferative growth

**Figure 1B:** Microscopic appearance of squamous cell carcinoma. HandE, 10X

**Figure 1C:** Lymphangioma circumscriptum presenting as multiple vesicles

**Figure 1D:** Microscopy of Lymphangioma circumscriptum - Multiple lymphatic channels in the superficial dermis. HandE, 10X

**Figure 2A:** Pyoderma gangrenosum

**Figure 2B:** Microscopy of pyoderma gangrenosum-Ulceration of the epidermis with extensive neutrophilic infiltrate. HandE, 10X

**Figure 3A:** Microscopic appearance of septal Panniculitis. HandE, 10X

**Figure 3B:** Microscopic appearance of cutaneous angiofibroma. HandE, 10X

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