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General Surgery

CARCINOMA CLITORIS: A CASE SERIES AND REVIEW OF LITERATURE

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ABSTRACT

Background: Approximately 4% of all gynecologic cancers in the United States are vulvar carcinomas. Although, malignancy is detected in almost every parts of vulva, the incidence differs. Histological carcinoma of clitoris was found to be different from squamous cell carcinoma of vulva. The localization of tumor, its size, status of lymph node and intensity and depth of invasion are some of the important prognostic factors.

Method: Present study was carried out in the Department of General Surgery tertiary care centre in India over a period of two years. In our series, 6 patients underwent surgery for clitoral mass. All the surgeries were performed by single surgical oncologist. All patients are still undergoing follow-up.

Results: The demographic parameters of the study were with the mean age of presentation as 58.5years. Majority of patients had a chief complain of growth or swelling in vulvar area. The mean duration of presenting symptoms were 3.1 months. The pre- operative biopsy of majority of patient showed squamous cell carcinoma and one patient had cellular angiofibroma. Various surgical procedures performed were wide local excision with primary closure or with the help of local rotational flaps. Inguinal nodal dissection was carried out in 3 patients. final histopathology of 4 patients was well differentiated squamous cell carcinoma and one each had moderately differentiated squamous cell carcinoma and cellular angiofibroma. The mean duration of follow up in the study is 12 months.

Conclusion: Clitoral carcinoma is a rare tumor encountered in day-to-day practice, even rarer in the younger age group of patients. There are different treatment strategies, such as surgical, radiation and chemotherapy. Management of such cases is a challenge for the surgeon due to late diagnosis and lack of well accepted protocol for management of such cases.

KEYWORDS :

INTRODUCTION:

Anatomically, Clitoris a significant part of vulva is located in the anterior region where labia minora meet. It forms a round visible part, the glans and a long shaft, covered by tissue of labia minora, in form of a hood or prepuce. The other anatomical parts of vulva are labia majora, labia minora, openings of vagina and urethra and perineal area. Due to the presence of a dense concentration of nerve endings, the function of clitoris is pleasure of sex and organisms[1].Any abnormality in clitoris could lead to sexual dysfunction related with arousal and/or orgasmic disorders[1].

Approximately 4% of all gynecologic cancers in the United States are vulvar carcinomas with an estimated 3490 new cases diagnosed in 2007 and an estimated 880 women will die from vulvar cancer[2]. Recently data shows a striking increase in the incidence of Vulvar Intraepithelial Neoplasia (VIN) as a precursor lesion of the undifferentiated form of invasive vulvar carcinoma over the last 30 years in New Zealand, United States, Norway and Austria[2,3,4,5]. In contrast, the overall incidence of invasive vulvar cancer has not changed in Austria[3], whereas in the US population, an increase of 20% between 1973 and 2000 was reported in the study.[6]

Although, malignancy is detected in almost every parts of vulva, the incidence differs. Bokhman*et al*.reported 73 lesions confined to vulva, 17 to labia majora and minora, 41 to the clitoris and 17 lesions to other structures[7]. In a series 50 patients, Popovic*et al*. found 42 lesions to be on the labia majora, 7 on labia minora, 1 each on clitoris and Bartholin`s gland[8]. Among 360 patients diagnosed with vulvar cancer, 40 cases were found to have clitoral cancer.[9]

Histologically carcinoma of clitoris was found to be different from squamous cell carcinoma of vulva. It shows large areas showing a transitional cell carcinoma pattern and foci of spindle and anaplastic cells, in addition to poorly differentiated non-keratinizing squamous cell carcinoma. The clitoral neoplasm was also found to be different in its cytokeratin profile from that described in invasive squamous cell carcinoma of vulva, especially with regard to the widespread staining for cytokeratin[10].The localization of tumor, its size, status of lymph node and intensity and depth of invasion are some of the important prognosticfactors.

OBJECTIVE:

Data has been published previously on vulval cancer along with different parts of involvement but there is scarcity of data on clitoral cancer. In view of the difference in histology of vulvar and clitoral cancer it was found important to make an attempt to review the literature on clinical presentation, including etiology, diagnosis, pathogenesis, treatment and follow up strategies of clitoralcarcinoma.

MATERIALS AND METHODS:

Present study was carried out in the Department of general surgery in Chirayu Medical College and hospital, Bhopal, India over a period of two years. In our series, 6 patients underwent surgery for clitoralmass. All the surgeries were performed by single surgical oncologist. All patients are still undergoing follow-up.

The patient's medical record, surgical notes, radiographs, and pathologic records were retrospectively reviewed. Written informed consent was obtained from all of the patients included in this study. Those patients who did not consent to participate in the study were excluded.

Initial assessment and diagnosis were made with the help of history, physical examination, Pap smear and biopsy. Local extent of tumor was assessed with CT scan/ MRI in all patients. Additionally, the basic essential work up was done in patients undergoing surgical excision of the tumor and reconstruction of the defects.

DATA COLLECTION:

Data were collected from patient records, pathologic reports and surgical notes. Afterwards, demographic data and clitoral cancer characteristics were extracted.

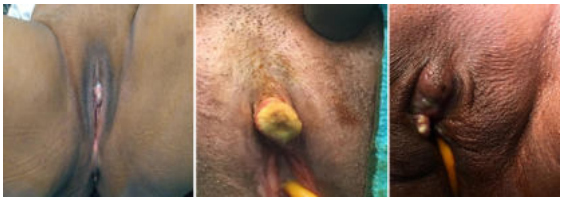
DATA ANALYSIS:

The following data were analyzed for each clitoral cancer: patent's age, clinical presentation, duration of symptoms, Pap smear, pre-operative biopsy, final histopathological diagnosis and follow up.

RESULTS:

The demographic parameters of the study were with the mean age of presentation as 58.5years. Majority of patients had a chief complain of growth or swelling in vulvar area. The mean duration of presenting symptoms were 3.1 months. The pre- operative biopsy of majority of patient showed squamous cell carcinoma and one patient had cellular angiofibroma. Various surgical procedures performed were wide local excision with primary closure or with the help of local rotational flaps. Inguinal nodal dissection was carried out in 3 patients. Final histopathology of 4 patients was well differentiated squamous cell carcinoma and one each had moderately differentiated squamous cell carcinoma and cellular angiofibroma. The mean duration of follow up in the study is 12months.

Figure A-C depicts pictographic presentation of clitoral carcinoma.



(A) (B) (C)

Table: Clinico-pathological parameters of various cases Clinical pictures

Patient	Age	Clinical complaints	Duration of complains	Pre- operative biopsy	Pap smear	Surgery performed	Final histopathology	Follow up
Case 1	58	Swelling Lump	3 months	Verracoidprolifera tionwith dysplasia	Atrophic vaginitis	WLE with primary closure	WDSCC	18 months
Case 2	75	Pain itching	1 month	NP	Senile vaginitis	WLE with primary closure	WDSCC	15 months
Case 3	60	growth	4 months	MDSCC	NP	WLE + LRF +B/L inguinal dissection	MDSCC	13 months
Case 4	63	Itching growth	2 months	WDSCC	Atrophic vaginitis	WLE +LRF +B/L inguinal nodal sampling	WDSCC	10 months
Case 5	65	growth	6 months	WDSCC	Senile vaginitis	WLE + Primary closure + LT. inguinal dissection	WDSCC	8 months
Case 6	30	swelling	3 months	Cellular angiofibroma	Inflammatory	WLE + primary closure	Cellular angiofibroma	8 months

DISCUSSION:

The vulvar cancer is normally diagnosed in later stages, due to ignorance of the symptoms by the patient, and due to embarrassment to reveal the personal details to a health provider.

The various etiological factors responsible for clitoral cancer are

A. Unprotected sex and infections from viruses and fungus,
B. Circumcision and infibulation,
C. Clitoromegaly, of several etiologies,
D. Priapism, due to erectogens or some pathological conditions,
E. Invasion from vulvar cancers (angiokeratoma, keratoacanthoma, angiofibroma, melanoma, neurofibromas),
F. Metastasis from ovarian tumors (Sertoli-Leydic and Stromal-Leydic cell tumors, gynandroblastoma),
G. Mixed gonadal dysgenesis, including gonadoblastoma,
H. Metastatis from adrenal carcinomas and adenomas,
I. Metastasis from squamous and transitional cell carcinomas of the bladder and pelvis
J. Genitourinary neurofibromatosis and (k. vascular tumors, includinghemangiopericytoma.[11]

Treatment options available are: a) clitoridectomy to treat clitoromegaly associated with malignancy, b) Clitoroplasty is performed to protect the neurovascular bundle and glans, C) Vulvectomy, either alone or in combination with dissection of inguinal lymph node with subsequent photon therapy is done for vulvar cancers with malignancy of clitoris d) Pelvic lymphadenectomy is done in case of inguinal node metastases e) CO2 laser is used to cure cancers of vulva affecting clitoris f) Laser vaporization is done for treatment of condylomata acuminate g) Sentinel node procedure is a safe treatmentoption[11].

Due to follow up treatment with the basic investigations and biopsy as the last modality for diagnosis, valuable time is lost because the cancer is growing masked by the seemingly normal investigation profiles. Patients with clitoral SCC have worse prognosis compared to patients without clitoral involvement, because of the deeper invasiveness of the primary tumors resulting in more groin metastases and its central location, involving bilateral lymph nodes. Thus, any suspicious lesion needs to be biopsied early for good prognosis of the patient.[12] prior to the availability of combination chemotherapy; mortality was very high even for stage 1 disease. Administration of combination chemotherapy with 5- Fluorouracil and cisplatin, has radically changed the mortality rate, and further increased the survival rate. However, because of high rate of local recurrence of vulvar SCCs, close long-term follow-up isnecessary.

Preventive Strategies

Genital Human Papillomavirus (HPV) infection is the commonest sexually transmitted infection worldwide. It is known fact that 90% of cases of anogenital warts are caused by HPV and HPV has been implicated in many of the gynecologic cancers, including those of vulva. Vaccines (quadrivalent (HPV subtypes 6,11,16 and 18) and bivalent (HPV subtypes 16 and 18) are available and licensed in some countries. Both vaccines show a more than 90% protection against persistent HPV infection for up to 5 years after vaccination [13]. The circumcision of clitoris is often related with inclusion of cysts and/or clitoromegaly, which might form the basis for clitoral malignancies. Hence, circumcision may be condemned by educating the masses who practice it. Furthermore the health care professionals should teach their patients about bad genital hygienic practices, such as the use of rougher toilet paper and/or keeping the partsdirty.[11]

CONCLUSION:

Clitoral carcinoma is a rare tumor encountered in day-to-day practice, even rarer in the younger age group of patients. The different etiological factors for clitoral malignancies are infective organisms, cysts, local invasion of cancers in the vicinity and metastasis from distant organs. The diagnosis of clitoral tumors includes certain biochemical measures; histological, surgical procedures and radiological diagnosis. There are different treatment strategies, such as surgical, radiation and chemotherapy. Management of such cases is a challenge for the surgeon due to late diagnosis and lack of well accepted protocol for management of such cases[14]. Religious, economic, and ethical factor should be considered along with counseling of the patient and her relatives regarding the risks and hazards of the disease, investigations, and the treatment options available.

ABBREVIATIONS:

CT: Computed tomography; MRI: Magnetic resonance imaging; SCC: squamous cell carcinoma; HPV: human papilloma virus.

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