Online Journal of Gynecology, Obstetrics and Maternity Care
ISSN: 2642-0163

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Case Report *Volume 6 Issue 1*

Malignant Extrarenal Rhabdoid Tumour of Vulva - A Very Rare Case Report

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Received Date: November 08, 2024; **Published Date:** November 21, 2024

Abstract

Vulval sarcomas account for only 1-3% of all vulval malignancies. Common vulval sarcomas described are Leiomyosarcoma, Angiomyxoma, fibrohistosarcoma. Malignant extra renal rhabdoid (ERT/MRT) of vulva is very rare and very aggressive neoplasm with very poor prognosis. Median survival reported in studies is 9 months to 61 months. Only 14 cases have been reported till date in English literature. We are reporting the 15th very rare case of a 31 years young female with primary vulval extra renal rhabdoid tumour of vulva who presented with tender vulval swelling with a short duration of 15 days.

Keywords: Malignant Extra Renal Rhabdoid Tumour of Vulva (MRT/ERT); Immunohistochemistry; Malignant Rhabdoid Tumor

Abbreviations

MRT: Malignant Rhabdoid Tumour; ERT: Extra Renal Tumour of Vulva; IHC-Immunohistochemistry; CT: Computerized Tomography; MRI: Magnetic Resonance Imaging; NCI: National Cancer Institute Nagpur India.

Introduction

Extrarenal malignant rhabdoid tumor (EMRT) highly aggressive neoplasm. Although they are known to occur during infant and childhood, they can occur in adults [1]. Malignant rhabdoid tumor of kidney was first reported in 1978. Though it was initially thought to be rhabdomyosarcomatoid variant of wilms tumor, it was subsequently recognized as separate clinico pathological entity. Most MRT occur in infancy and are aggressive in nature, majority of patient depart this life in short period of time. Malignant extrarenal rhabdoid tumor were described in various extrarenal sites like soft tissue, skin, gastro intestinal tract, liver, urogenital tract, paraspinal and paratesticular area and CNS [2]. MRT commonly occur in deep axial location like neck or paraspinal region. Microscopically tumor consist of diffuse proliferation of rounded or polygonal cells with eccentric nuclei, prominent nucleoli and glassy eosinophilic cytoplasm containing hyaline line inclusion bodies arranged in sheets or nests. These characteristic "rhabdoid cells" are also present in certain other carcinomas like renal cell carcinoma and soft tissue sarcomas, like synovial sarcomas, extra skeletal myxoid chondrosarcoma and leiomyosarcoma [3]. MRT of vulva is very rare and aggressive neoplasm of uncertain histogenesis [4]. On line search yielded 14 reported cases till

now in English literature, probably this is the 15th reported case. Presenting herewith a case report of young female who presented with vulval swelling in one hospital with pain of 15 days duration, diagnosed as Bartholins abscess, posted for surgery but during surgery mass was firm and pus could not be drained thus biopsy was taken and unexpectedly the report turned out to be malignant rhabdoid tumour of vulva with a grave prognosis. In spite of surgery, chemotherapy patient succumbed to death within nine months of diagnosis due to extensive metastasis.

Case Report

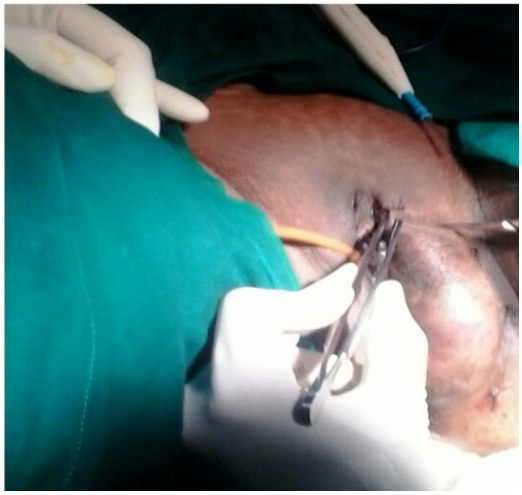
A 31 years married woman presented to one of the gynaecologist with history of swelling over vulval area left side since 15 days, with pain on same side since 5 days. The swelling increased in size in 15 days from a small boil to lemon size. Patient was para 1, with normal vaginal delivery and last child birth was 2.5 years back. She was menstruating regularly. Her personal and family history was not contributory. Patient had her normal delivery with the same gynaecologist. Her general and other systemic examination was normal. On local examination she had vulval swelling was noted on left side 3/3 cm tender, skin over it was inflamed, thus they made clinical diagnosis of? bartholins abscess left vulva,?? vulval abscess. She was investigated and posted for surgery incision and drainage. During surgery they found that mass was firm and no pus/ fluid was drained thus they took biopsy from the mass and sent it for histo pathological examination. The HP diagnosis came as epitheloid sarcoma of vulva and patient was referred to national cancer institute for the further management. When patient came to NCI she was examined again. Patient and family members were very anxious and mentally disturbed due to unexpected diagnosis. She was clinically stable. Her local examination revealed left vulval swelling of 7/7 cm red, inflamed skin. There was no groin or other lymphadenopathy. Swelling had increase in size from 3 cm to 7/7 cm in 7 days. She was investigated further with complete blood counts, liver function test, renal function test, coagulation profile, HIV, Hepatitis b , HbSAg, LDH , ultra sound whole abdomen and chest X ray was advised all the reports were within normal limits. CT scan of abdomen and pelvis was done which revealed thickened vulval wall with soft tissue density lesion. MRI was suggestive of multilobulated large neoplastic lobulated mass along left vulva displaying midline towards right side, extending posteriorly along posterior side of left ischio pubic ramus and ischiorectal fossa. Bilateral inguinal lymph nodes were noted but were small in size and not significant. The tumour rapidly increases in size from 2/2 cm to 7/7 cm in 15 days.

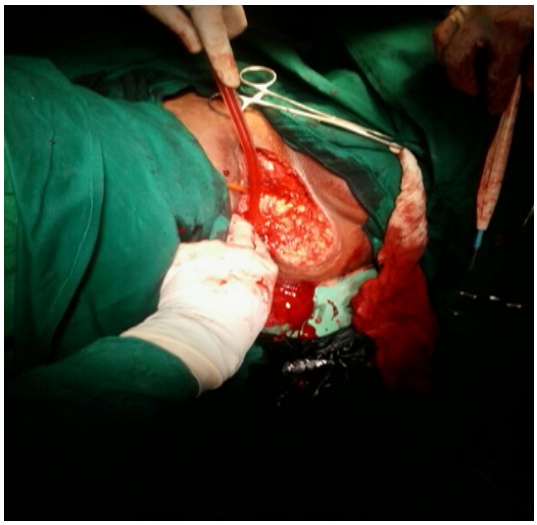
The final clinical diagnosis was discussed in detail with patient and relatives. All modalities of treatment were discussed and prognosis was explained to relatives and counselling was done. Patient was posted for surgery-radical vulvectomy with wide local excision with unilateral groin dissection. Under spinal and epidural anaesthesia after catheterizing urinary bladder. There was a vulval mass of 7/7/7 cm on left side, hard in consistency. Skin over mass was showed erythematous patches. Clitoris was free. Posteriorly mass was reaching up to anterior anal margin. Per speculum, per vaginal and per rectal examination was normal. Radical hemivulvectomy was done with left groin dissection. Lymphatic tissue with gland of Cloquet was removed and sent for histo pathological examination. Urinary catheter was placed for 10 days and drain was placed at groin dissection site for 4 days. Post-operative period was good and wound healing was good.

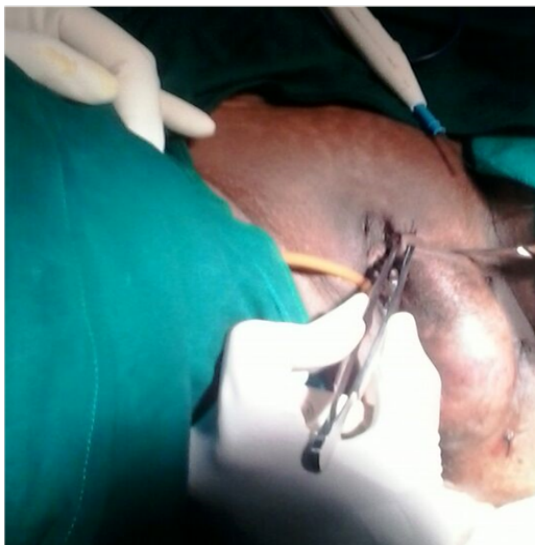
The histopathological report showed high grade malignant lesion located in dermis and sub cutaneous plane. Tumour cells were arranged in diffuse sheets with little cohesiveness. Individual cell showed characteristic eosinophilic cytoplasm imparting a rhabdoid phenotype. On IHC tumour cells were focally positive for EMA and CK whereas negative for MYOD, Desmin. Tumour cells nucleus showed loss of INI expression, all features were suggestive of Extra renal rhabdoid tumour of left vulva. One out of 8 lymph nodes showed metastases with perinodal extension.

Patient was advised and started with chemotherapy 5 cycles and was advised radiotherapy. Patient had recurrence of the disease locally and later systemic with extensive lung metastases within five months of diagnosis. Palliative treatment was continued but patient deceased within nine months of initial diagnosis.

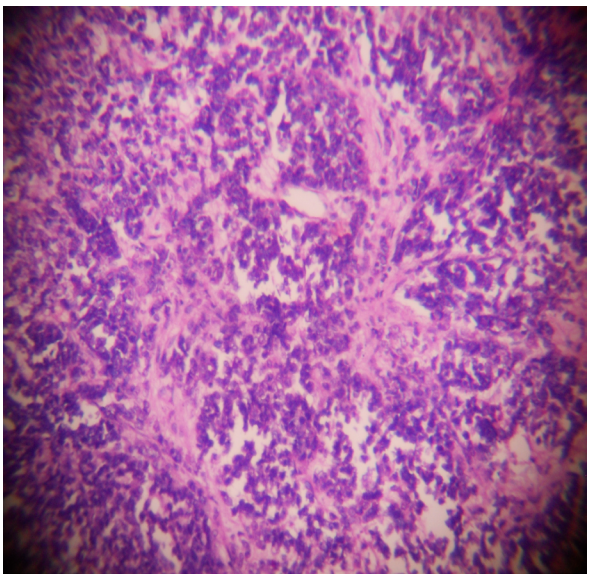
Figure 1: Vulval Swelling before Surgery as Described.

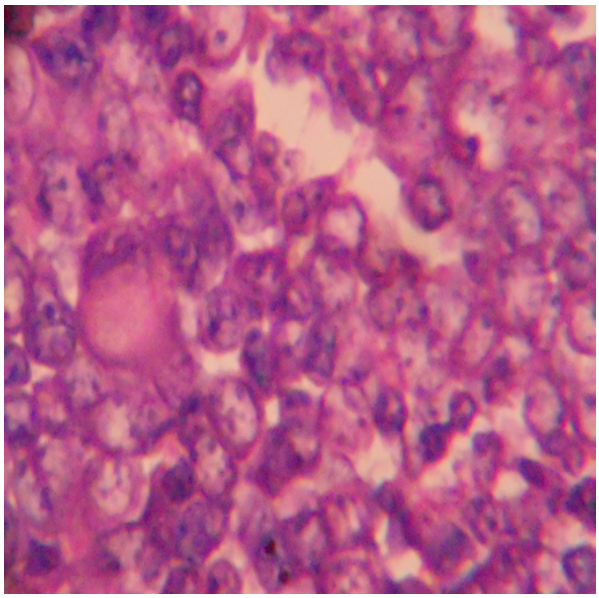






Figures 2, 3 & 4: Intraoperative Hemivulvuvectomy as Described.





Figures 5 & 6: Histopathology Microphographs as Describe.

Discussion

Extra renal rhabdoid tumour of vulva is very rare and aggressive gynaecological neoplasm with median age of 39 years. Patients usually presents with short history of palpable mass, labia majora is most frequently affected site. Beckwith and Palmer described malignant rhabdoid tumour in kidney as a variant of Wilms tumour in 1978. Extra renal forms occur in variety of sites including soft tissue, para spinal, para testicular site, colon, tongue, prostate, thymus, heart, vulva and uterus and central nervous system [5,6]. Till now in English literature only 14 cases have been reported and

we are reporting 15 th case of extra renal rhabdoid tumour of vulva. In contrast to other vulval malignancies like squamous cell carcinoma ,majority of cases of MRT's of the vulva are located on labia majora, other reported sites are vulva, mons pubis and clitoris. Most of these vulval tumor are noted in the age group of 30-50 yrs., youngest patient being 19yrs and oldest 63yrs of age [7,8]. most of them are of reproductive age group. In literature 5 had vulval mass on right side, 6 had on left side, unspecified in 2 and near mons pubis in 2 cases, in our patient it was on left side. Most of these reported MRT of vulva presented as painless, firm to hard circumscribed mass of 2 to 10 cm size in our patient presentation was with 3/3 cm mass. Pain was present in 5 out of 14 cases, in our patient also pain and mass were presenting complaints. Lymphatic spread was rare in most of other sarcomas, compare to more noted in vulval carcinomas.

Diagnosis was often confirmed by some form of incisional biopsy. This is necessary to provide sufficient material for IHC. Most MRT express vimentin, followed by EMA, cytokeratin and CD99. They do not express desmin [1] and they also show loss of nuclear expression of INI 1 protein. . In our case histopathology showed high grade malignant lesion in dermis and sub cutaneous plane and tumour cells were arranged in sheets with little cohesiveness. Individual cell showed characteristic eosinophilic cytoplasm imparting arachnoid pheno type. On IHC tumour cells were focally positive for EMA, CK & negative for MYOD1 & Desmin, features classically suggestive of extra renal rhabdoid tumour of left vulva.

Prognosis of ERMT in generally poor and their course tend to be assessed for loco regional and distant spread of the tumor prior to treatment planning. Treatment usually consists of wide local excision or hemivulvectomy depending upon the local tumor extent. . Radiological evaluation including CT chest should be done to assess locoregional and distant spread of tumour prior to treatment planning. PET scan might be useful to stage the disease.

Surgery usually includes grain node dissection. The post resection defects can be primarily closed, grafted or myocutaneous flaps used. Prognosis depend on size, depth of invasion, histological types, margins status after resection [1]. Chemotherapy or radiotherapy may be considered as additional first line treatment options. A study conducted by Woldenetal in pediatric patients with extrarenal, extra cranial MRT suggest that radiotherapy along with chemotherapy and surgery has a potential to prolong survival with acceptable toxicity [9]. The prognosis of ERMT is generally poor and their course tends to be aggressive, although surgery is the first line of management. There is no consensus that exist regarding adjuvant therapies. Chemotherapy and radiotherapy did not show to be effective in controlling the recurrent or metastasis diseases in all reported cases [1,5,6]. The reported median survival for patient with vulva MRT is around 9 months, the longest reported survival being 61 months. Local recurrence within a year are common and the appearance of distant metastases is almost invariably rapidly fatal. Most common site of metastasis is lung, chemotherapy or radiotherapy do not appear to be effective in controlling recurrent or metastatic diseases [8,10].

Conclusion

Malignant extra renal rhabdoid tumour of vulva is very rare gynaecological tumour of vulva with very aggressive nature. Presentation usually is short history of vulval swelling with pain and can be misdiagnosed initially clinically as vulval abscess. Vulval biopsy with histopathology and immunohistochemistry is the key to diagnosis. Most of the patients are in reproductive age with Mean age of presentation is 39 years. Surgery remains primary modality of treatment. Prognosis is poor.

Conflict of Interest

The authors disclose no conflict of interest

Financial Support

NIL

Acknowledgment

We express our sincere gratitude to the management of National cancer institute Nagpur (Maharashtra) India for their support and encouragement

Ethical Committee Approval

Taken, Patients and relatives' consent was taken for publication.

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