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RESEARCH ARTICLE

RARE CASE OF PROXIMAL EPITHELOID SARCOMA OF VULVA IN A YOUNG ADULT – CASE REPORT AND REVIEW OF LITERATURE

***Jayashree Natarajan**

Kidwai Memorial Institute of Oncology, India

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ABSTRACT

Vulval tumors are rare disease and account for 4% of gynaecological malignancies. (Newman and Fletcher, 1991) These rare tumors are disease of elderly age group. Primary vulval sarcomas account for 1-3% of vulval malignancies (Newman and Fletcher, 1991; Disaia *et al*., 1971; Curtin *et al*., 1995). The most common subtype is leiomyosarcoma of vulva. Few subtypes of vulval sarcomas are seen in adolesents and young adults. One of such rare type vulval sarcoma is epitheloid sarcoma. The epitheloid sarcoma variety of soft tissue sarcoma is seen in proximal and distal extremities of body. The proximal type tumors are found to be more aggressive with poor prognosis. The mean age of presentation is 36 years, with patients as young as 17 years are also been reported. We report a case of proximal type of epitheloid sarcoma of vulva with multiple recurrence of tumor managed with surgery and chemotherapy.

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INTRODUCTION

21 years old unmarried girl presented with complaints of swelling in left vulva since one year, gradually increasing in size with no associated pain or discharge.She had consulted gynaecologist for the same complaints 7 month before. She was advised Ultrasound of the vulval and groin region. Ultrasound of vulvalregion showed a 2.6x2.4x2.4cm lobulated mass in subcutaneous plane in left side mons. Ultrasound of the mass showed no calcification with minimal cystic degeneration.On colour Doppler significant neovascularity with prominent vascular pedicle on medial side seen. Ultrasound of inguinal nodes showed tiny lymphnodes with normal echotexture. FNAC of the mass was diagnosed as epidermal cyst. Patient was counselled about the benign nature of the mass and reassured. Since the mass was gradually increasing in size patient reported again. Patient offered no complaints of pain, white discharge per vaginum, menstrual irregularities, altered bowel or bladder habits. Repeat USG showed mass measuring 5.3x3.0x4.5cm hypoechoic lesion with lobulated margins, seen mainly in subcutaneous plane, with peripheral areas of necrosis, internal vascularity with arterial flow signal seen. Underlying pubic bone, uterus, both ovaries were found normal. Bilateral inguinal lymphadenopathy with fat stranding on left side more than right side seen. MRI pelvis showed well defined lobultedmass heterogeneously enhancing in the subcutaneous plane in the pubis on left side measuring 53x45x40mm in size, extending inferiorly in the proximal labium, slightly hypointense on T1, hyperintense on T2 with multiple septation. It showed multiple flow voids noted with in the lesion suggestive of arterial flow, few enlarged left inguinal lymphnodesseen.Patient underwent left vulvar mass excision with lymphnode dissection. Intraoperatively, a firm mass under the skin over the left side of mons pubis, extending from the labia majora, to the root of the thigh of size approximately 4x5cm. Myxomatous degeneration was present. Lymph nodes were dissected and sent for Histopathological examination showed fibroadipose and muscular tissue infiltrated by a tumor composed of cells arranged in sheets and lobules. Cells were showing round vesicular nuclei with prominent nucleoli and moderate eosinophilic cytoplasm. Few of these cells showed abundant densely eosinophilic cytoplasm. Scattered mitosis observed. Stroma showed myxoiddegeneration. Lymphnodes showed metastatic tumor deposits. The impression by the first pathologist was high grade sarcoma, features suggestive of malignant peripheral nerve sheath tumor with extensive areas of rhabdoid differentiation. Review with second pathologist confirmed high grade sarcoma with areas of myxoid and epithelioid differentiation vulva. On immunohistochemistry tumor was positive for vimentin, AE1/AE3, weakly positive for EMA, focally positive for CD34 and negative for desmin, myoglobin, SALL4, S-100, INI 1. Immunohistochemistry

****Corresponding author: Jayashree Natarajan,***
Kidwai Memorial Institute of Oncology, India

confirmed the diagnosis of epitheloid sarcoma of the vulva proximal type. Patient developed swelling in the left inguinal region 2months after surgery. She underwent excision of left inguinal soft tissue tumour with inguinal block dissection. Microscopy of dissected specimen showed o.1cm residual tumor microscopically with reactive nodes. Patient received six cycles of chemotherapy with Ifosfamide and Adriamycin. After chemotherapy patient was asymptomatic and was on follow up for the following 8 months. On follow up she was found to have multiple firm to hard nodules on the left side vulva, left labia was pedunculated and hanging loose. Small nodule felt in the lower one third of vagina. Patient underwent wide local excision of the vulvallesion, 5x4cm mass excised with 1cm margin all around. The histopatholgy confirmed recurrent mass of epitheloid sarcoma of proximal type. Patient was counselled regarding the nature of the disease and further managements. Options for second line chemotherapy versus observation explained. Patient opted for observation. She is on followup for last 5 months after her last surgery, surviving with the disease for 42 months since the time she noticed and 36 months from her initial consultation.

FNAC of the mass and benign diagnosis of epidermal cyst was offered, which delayed the management for 8-9months by then the disease had increased in size with spread to groin nodes. These tumors has been reported in patients aged between 17-80 years. The soft tissue sarcomas of vulva including the epitheloid variant most commonly present with swelling in the external genitaliasome times patients may present with itching in external genitalia, rarely with secondary changes in skin like ulceration or bleeding from the tumor. The median age for presentation of epitheloid sarcoma is 36.4years. The earlier reported cases of epitheloid sarcoma are extra genital involving the distal upper extremities (58– 74.5%), particularly the hand and the wrist area, followed by the distal lower extremities (15%), proximal lower extremities (12%), proximal upper extremities (10%), the trunk (3%), and the head and neck (1%). (Hasegawa *et al*., 2001) The involvement of vulva by primary epitheloid tumor is rare, with reported cases less than ~ 30. The proximal type of primary epitheloid tumors of vulvaare found to be more aggressive with very poor prognosis compared to extra genital lesions. (Ubright *et al*., 1983)

Table 1.

Tumor type	Vimentin	Cytokertin	EMA	s-100	CD31	Desmin	CD 34	Others
PES	p	p	P	N	N	P/N	P/N	
LCS	P	N	N	P				P-Langerin
MS	P	N	N	N	N	N	N	P-CD10
EMPNST	P/N	P/N	P					
MM				P				P-gp100 (HMB-45)
LMS	Fascicle of elongated tumor cells, with blunt shape, cigar shaped atypical nucei.					N	N	
Undifferentiated carcinoma	PES – does not Involve over lying dermis or cutaneous adnexae, absence features of glandular or squamous differentiation						N	

P-positive; N-negative; PES-primary epitheloid sarcoma, LCS-langerhan cell sarcoma, MS- mammary sarcoma, EMPNST- epitheloid malignant peripheral nerve sheath tumor; MM- malignant melanoma; LMS –leiomyosarcoma

Table 2.

Score	0	1	2	3
Tumor differentiation score (A)		Resembling normal adult mesenchymal tissue, eg, Well differentiated liposarcoma	Sarcomas for which the histologic typing is certain, eg, biphasic synovial sarcoma, myxoidliposarcoma	Embryonal sarcomas, undifferentiated and sarcomas of undoubtful tumor type
Mitosis count per 10 high power fields (B)		0-10	11-19	>20
Tumor necrosis score (C)	No necrosis	<50% necrosis in exmined tumor surface	>50% of examined tumor surface	

Trojani score =A+B+C
Grade 1=2,3
Grade 2=4,5
Grade 3=6,7,8

DISCUSSION

Epitheloid sarcoma a soft tissue tumour first described by enzinger in 1970 (Enzinger, 1970), followed by the description of vulval epitheloid sarcoma by Piver *et al*. in 1972. (Piver *et al*., 1972) This aggressive tumour is a disease of adolesents and young adults. This aggressive metastatic rapidly recurring tumor is fortunately a rare tumor. In his initial description of this tumor Enzinger found this tumor to be misdiagnosed as benign pathologies like granulomatous inflammation, nodular tenosynovitis, nodular fasciitis, chronic ulcer or inflammation etc The most common malignant diagnosis given for this tumour were synovial sarcoma, fibro sarcoma, malignant melanoma, angiosarcoma etc. Hence high level of suspicion is needed to reach the right diagnosis of epitheloid sarcoma. In our reported patient also at her initial visit she had undergone

Histologically the tumor consists of nodules of polygonal epithelioid and spindle cells, or contains both (Behranwala *et al*., 2004). These cells are lightly eosinophilic, often with intercellular collagen, and sometimes with hyalinization. The growth pattern of epithelioid cells is a pseudogranulomatous proliferation with broad, undulating collars around central, relatively acellular or necrotic areas. The tumor may show chronic inflammatory cells infiltration misleading to the diagnosis of a inflammatory or infective pathology (Enzinger, 1970). On immunohistochemistrytumor shows positivity for vimentin, cytokeratin, epithelial membrane antigen, negative for S100, CD31, some cases positive for Desmin, CD34. (Lin *et al*., 2012; Petrillo *et al*., 2011; Hasegawa *et al*., 2001) The major immunohistochemistry difference with the other tumors are given in Table 1.

Malignant rhabdoid tumor resembles proximal type epitheloid tumor in its immunohistochemistry, but carries a very aggressive coarse. (Argenta *et al*., 2007) Both tumors has been studied for their genetic origin, INI1 sensitivity of tumors has been studied to differentiate. (Kim *et al*., 2012)

EORTC soft tissue sarcoma grading system given below in Table 2

In various studies for vulval sarcomas the prognostic factors favouring good outcome were size of the primary tumor less than 5cm, low grade histology,and complete resection. inadequate resection margins was a important prognostic factor for recurrence. Hence resection of tumor with adequate surgical margin (defined 2cm by some authors) is the best way to manage these tumors with poor response to chemotherapy or radiotherapy. These aggressive tumors are found to recur locally in 70-77% patients (Ubright *et al*., 1983). The best management protocol with concensusis not available for this rare tumor. Wide local excision or radical vulvectomy with adequate margin will reduce the chance of recurrence. Wide local excision with excision of lymphnode dissection has been postulated as the standard of care by some authors (Kasamatsu *et al*., 2001). The groin node dissection as a routine even without evidence of disease may show microscopic disease, and may help to clear these disease or help todecide for the need for adjuvant treatment. (Kasamatsu *et al*., 2001; Royal College of Obstetricians and Gynaecologists, 2006) The role of lymphadenectomy when the imaging does not show involvement is also not clear. Some authors prefer lymphadenectomy for these tumors since the lymphnodes are common site of recurrence. The role of adjuvant chemotherapy or radiotherapy is also controversial. In a study reported by Curtin *et al*. (Kasamatsu *et al*., 2001) among 24 women with gynecologic sarcomas, 7 women had low grade sarcomas. They were followed up after primary surgery without adjuvant therapy, only one out of the seven recurred. The most frequent metastatic site is the lung (51%), followed by lymph nodes (34%), scalp (22%), bone (13%), brain, liver, and pleura (EJAartsen, 1994). Ofthese, themostcommoninitialsiteswere lymph nodes (48%), followed by lungs (25%) (Curtin *et al*., 1995). The advantage of treating these patients with adjuvant radiotherapy or chemotherapy is not clear the various studies reported shows local control of 40-60% but patient dies of metastatic disease. Hence early diagnosis is the key to better survival. The indolent asymptomatic nature of disease causes delay of atleast 6 months in diagnosis (Disaia *et al*., 1971; Ulutin *et al*., 2003) but the rapid rate at which the disease metastasise they become unmanageable killers. (Patrizi *et al*., 2013; Moore *et al*., 2002; Royal College of Obstetricians and Gynaecologists, 2006; Ong *et al*., 2012; Ulutin *et al*., 2003)

Conclusion

Primary epitheloid tumors of the vulva are rare tumors. The management outcome depends upon the diagnosis at the right time with timely intervention. Excision of the tumor with adequate margins is the minimum mandatory management intervention. Need for radical vulvectomy, adjuvant therapy not clear. Early diagnosis and intervention will help in prolonging tumor free survival.

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