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Sarcomatoid Squamous Cell Carcinoma of Vulva: A Very Rare Entity - Case Report

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Abstract: ***The Sarcomatoid Squamous Cell Carcinoma (SCC), showing both epithelial and mesenchymal features, has been very rarely reported in the vulva. It is a rare variant of SCC with prominent spindle cell morphology. Till date less than 20 cases have been reported so far. It is also known as pseudosarcoma, spindle cell carcinoma or carcinosarcoma. Disease specific survival is worse, compared to non-spindle Cell morphology. Currently, most common treatment modality is surgical removal with or without adjuvant radiotherapy. Systematic reporting of these tumours is needed to plan future treatment protocols.***

Keywords: Vulval Cancer, Sarcomatoid Squamous Cell Carcinoma, Pseudosarcoma, Spindle Cell Carcinoma, Carcinosarcoma

1. Introduction

Vulval cancer is the fouth most common malignancy of female genital tract, mostly seen in elderly women. [1] The Sarcomatoid Squamous Cell Carcinoma (SCC), showing both epithelial and mesenchymal features, was first reported by Steeper et al in 1983. [2] It is reported at many anatomical sites like skin, oral cavity, larynx, breast, lung and oesophagus, but it is a very rare finding in vulva. [3,4] Till date less than 20 cases have been reported so far.

It is also known as pseudosarcoma, spindle cell carcinoma or carcinosarcoma. [2, 5] Spindle cell are thin, slender, cancer cells, which have atypical epithelial characteristic and have gained ability to infiltrate the underlying stroma and metastasize. They infiltrate stroma and can occur as single cells or as cords of cells. Disease specific survival is worse, compared to non-spindle Cell morphology. [6]

2. Case Report

A 62-year-old multiparous woman presented to our department in June 2022, with history of exophytic growth in pubic region. On examination, we found a 3.5 x 2 x1.5 cm sized exophytic nodular mass with irregular surface on the right Labia Majora. The mass did not cross midline, and urethral meatus, clitoris were also not involved, the mass was not adherent to deeper tissues or any bony structure, bilateral palpable inguinal lymph nodes were present. (Fig -1)

Figure 1: Vulval Growth

A punch biopsy was taken from the vulval mass and histo-pathological examination (HPE) was suggestive of undifferentiated spindle cell sarcoma. Fine needle aspiration cytology (FNAC) from bilateral inguinal lymph nodes was suggestive of chronic lymphadenitis thus ruling out nodal spread. The findings of CECT Scan of pelvis showed no pelvic lymph node enlargement.

We did a wide local excision of the lesion with bilateral superficial inguinal dissection. On pathological analysis, all resection margins and base were negative (0.5 cm superior, 0.5 cm inferior, 0.5 cm medial, 0.5 cm lateral and 0.5cm from base).

The HPE report showed spindle cell variant of SCC. Pleomorphic spindle cells arranged in storiform and fascicular pattern interspersed by small nests of atypical cells displaying squamous differentiation in the form of round to oval pleomorphic cells with vesicular chromatin, Prominent nucleoli and moderate amount of eosinophilic cytoplasm. At places, dysplastic stratified squamous epithelium is seen, which is invading the underlying zone in the form of small

nests and scattered singly. Intra-cytoplasmic keratinization is noted. Atypical mitosis is also seen. (Fig 2 & 3)

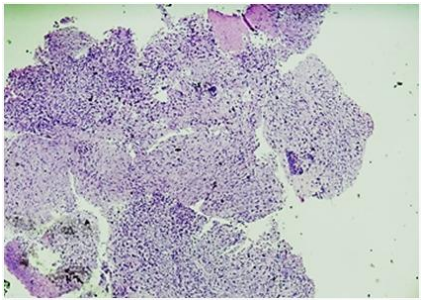

Figure 2: HPE (10x)

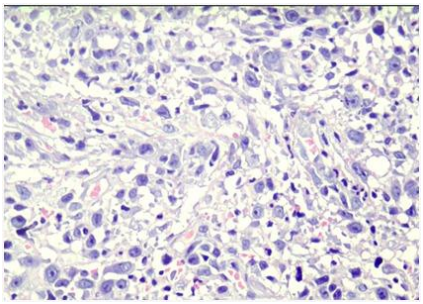

Figure 3: HPE showing spindle cells with sarcoma cells

Immuno-histo-chemistry (IHC) was confirmatory for Sarcomatoid SCC, The tumor cells are positive for CK, P40, P16, SMA and negative for S100, CD34, DESMIN Ki-67~70%.

The patient was discharged with no postoperative complication and adjuvant Radiotherapy was given. Currently after one and half year of follow-up, there is no evidence of local or systemic recurrence.

3. Discussion

The spindle cell or sarcomatoid variant of SCC, has been rarely reported in the vulva. It is a rare variant of SCC with prominent spindle cell morphology.

Mostly the sarcomatoid cells arise from metaplastic transformation of malignant epithelial cells (‘‘conversion’’ theory). Epithelial to mesenchymal transition is the biologic process through which a malignant epithelial cell, undergo progressive change to a mesenchymal cell. Loss of epithelial features like intercellular junctions, intercellular adhesion and contact inhibition with gaining of mesenchymal features, is accompanied with increased cell motility and great capacity for invasion and metastasis. This theory is supported by embryological, ultrastructural, immunohistochemical, molecular, and in vitro evidence. [6,7]

This cancer is mostly seen in post-menopausal women. The age of presentation ranges from 54 to 89 years, with a mean age of 75 years. In most cases, the tumour forms polypoid ulcerated masses ranging from 3 cm to 12 cm on the vulva. The presenting symptoms of vulval SCC are pruritus, leucorrhoea and mass. Predisposing factors include pre-existing vulval lichen sclerosus, epithelial hyperplasia and lichen planus. [8] The differential diagnosis include epithelioid sarcoma, synovial sarcoma, leiomyosarcoma, amelanotic melanoma, and dermatofibrosarcoma protuberans of the vulva. [5]

Vulval SCC with spindle cell morphology appears to be aggressive tumour type. This cancer type has a worse prognosis than conventional vulval SCC and an increased risk of lymph node metastases at the time of diagnosis. [1,5] In most cases, treatment includes wide local excision or radical vulvectomy with inguinal-femoral lymph node dissection in advanced cases. Adjuvant Radiotherapy can be given in high grade tumours, positive lymph node metastasis and close margins of resection to decrease possibility of loco-regional relapse. Primary treatment with Radiotherapy or Chemo-radiation leads to early relapse and poor overall survival. [3,5]

4. Conclusion

Sarcomatoid SCC being rare faces diagnostic difficulties, and has poor survival so systematic reporting of these tumours is needed to plan future treatment Protocols. Currently, most common treatment modality is surgical removal with or without adjuvant radiotherapy.

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