VULVAR SARCOMATOID CARCINOMA– A CASE REPORT OF AN UNUSUAL VARIANT AT AN UNUSUAL SITE

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ABSTRACT

BACKGROUND

Sarcomatoid squamous cell carcinoma (SSCC) is a well-recognised tumour in various sites of the body although its incidence is very low in lower female genital tract. Currently, there are only 17 published cases of vulvar sarcomatoid carcinoma in English literature. In this case report, we describe the histopathological and immunohistochemical characteristics of a vulvar sarcomatoid carcinoma. It is a tumour of extreme rarity and severe prognosis requiring a multidisciplinary approach for management, which should be included in the differential diagnosis of spindle cell tumours of lower female genital tract.

KEYWORDS

Sarcomatoid Squamous Cell Carcinoma, Sarcomatoid Carcinoma, Carcinosarcoma, Vulva.

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BACKGROUND

Sarcomatoid squamous cell carcinoma (SSCC) is a rare malignancy of vulva. The diagnosis of this variant is crucial because it differs from classical squamous cell carcinoma in terms of having advanced stage at presentation, poorer prognosis, aggressive course with short survival span and early recurrence following treatment. Here, we report a case of vulvar tumour having admixture of squamoid epithelial and spindle shaped sarcomatoid cells. To establish whether we were dealing with a single tumour or not, immunohistochemical characterisation of the sarcomatoid cells was performed using a panel of antibodies specific for both epithelial and mesenchymal differentiation. In this report, we also present a review of previously published cases.

Case Report

A 40-year-old lady came with a history of mass in vulvar region, which was rapidly increasing in size. The mass was associated with bleeding. Clinical examination revealed a 5 x 5 cm ulcerated, polypoidal growth in the vulva. Excisional biopsy of the mass was done. Cut section of the tumour was grey-white with extensive areas of necrosis and haemorrhage. Microscopic examination revealed a poorly differentiated tumour with sarcoma-like features. A radical vulvectomy with bilateral inguinofemoral lymph node dissection was performed later.

Histopathological examination showed a poorly differentiated tumour similar to the first excisional biopsy specimen. The tumour had both epithelial and mesenchymal features. There were regions of squamous cell morphology interspersed with spindle cells. Areas of haemorrhage and necrosis was noted. Immunohistochemistry was performed with markers like keratin, vimentin and HMB-45 to know the histogenesis of tumour. Spindle shaped sarcomatoid cells showed positivity for both keratin and vimentin. The tumour cells were negative for HMB-45 stain, thus ruling out the possibility of amelanotic melanoma.

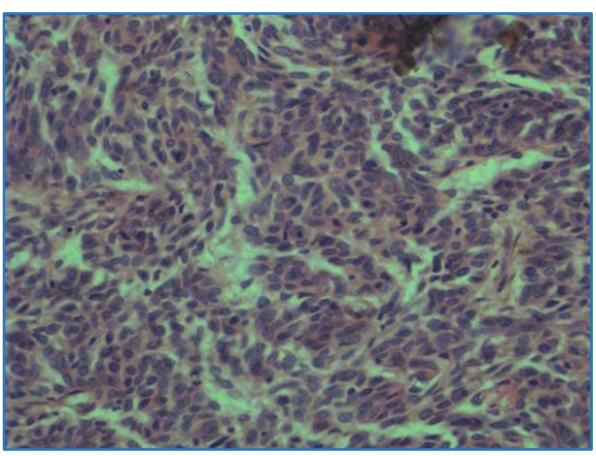

Figure 1. Round to Polygonal Squamoid Cells blending Imperceptibly with Sarcomatoid Spindle Cells

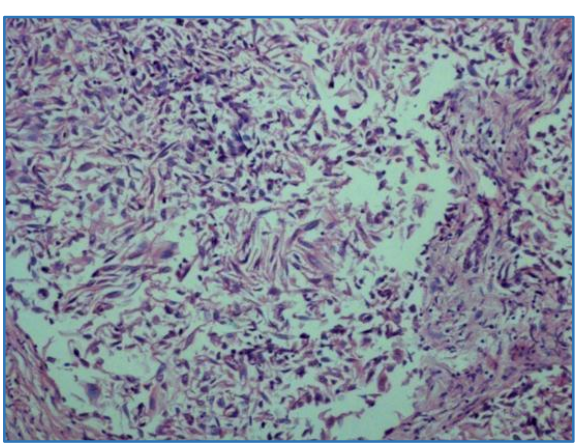

Figure 2. Sarcomatoid Cells having Elongated Nuclei and Scanty Eosinophilic Cytoplasm

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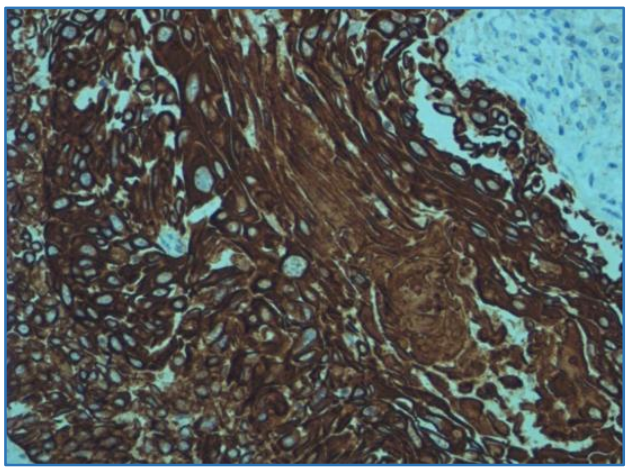

Figure 3. Tumour Cells showing Immunopositivity for Keratin

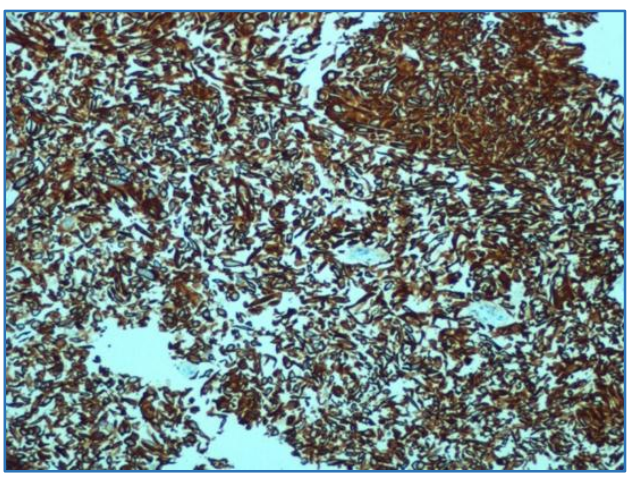

Figure 4. Tumour Cells showing Immunopositivity for Vimentin

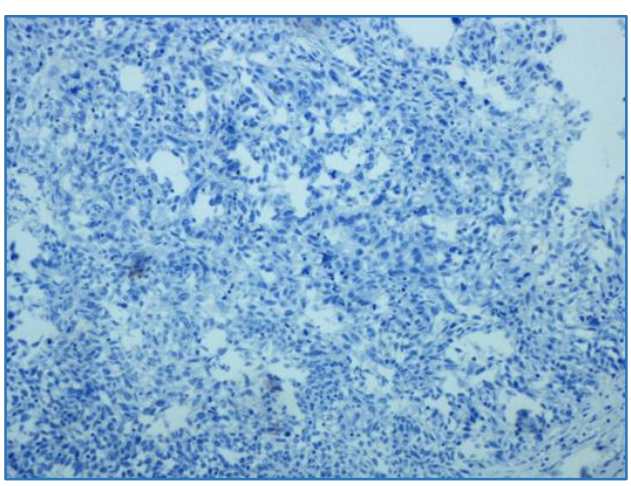

Figure 5. Tumour Cells showing Immunonegativity for HMB-45

DISCUSSION

Vulvar carcinomas are generally rare and constitute only about 5% of all genital tract malignancy. Squamous cell carcinomas are the most common primary malignant tumours of the vulva. Verrucous carcinoma, adenoid squamous carcinoma, basal cell carcinoma and sarcomatoid or metaplastic carcinoma are some of the unusual histological variants of vulvar squamous cell carcinoma.[1] Sarcomatoid squamous cell carcinoma is a well-documented tumour in various sites of the body like respiratory tract,[2,3] gastrointestinal tract,[4,5,6] urogenital tract,[7,8] thyroid,[9] skin,[10] breast.[11] Since these tumours consist of admixtures of both classical squamoid and spindle shaped sarcomatoid cells, different terms such as carcinosarcoma, metaplastic carcinoma, pseudosarcoma, sarcomatoid carcinoma, spindle cell carcinoma are used to describe them.[12]

The incidence of sarcomatoid squamous cell carcinoma in female genital tract is very low. There are currently 16 cases of cervical, 17 cases of vulvar and 5 cases of vaginal SSCC reported in the English literature to date. One of the possible reasons for this is, sarcomatoid component is underrecognised or underreported while signing out the histopathology report.[13]

Risk factors for SSCC of the female genital tract are not completely known. HPV infection, high risk sexual behaviour, immunosuppression and radiation exposure are assumed to be the most likely causes of SSCC.[13]

Many hypotheses have been proposed to explain the pathogenesis and aggressiveness of SSCC. Among these most accepted theory is that there is a transformation from the squamous cell carcinoma component into a spindle cell cancer. Parallel immunohistochemical, molecular and ultrastructural characteristics of SSCC and squamous cell carcinoma support this theory. Another theory is that spindle cell component and the squamous cell component arise concurrently from distinct stem cell lines.[13]

Fatigue, anaemia, pelvic pain, pelvic pressure, constipation, bloating, weight loss and loss of appetite are the early symptoms of SSCC. The most common symptom of vulvar carcinoma is a vulvar mass which is rapidly expanding and bleeds on touch. This tumour has unusual metastatic sites, one such is the skin.[14]

SSCC lesions of the vulva are generally described as being ulceroproliferative, friable, of polypoid configuration with haemorrhagic and necrotic areas. In general, the histopathological diagnosis of SSCC rests upon demonstration of a malignancy with round to polygonal squamoid cells, merging with spindle shaped sarcomatoid cells.[13]

The most problematic aspect of the differential diagnosis of SSCC of the female genital tract is its differentiation from malignant Mullerian mixed tumour (MMMT).[15,16] The differentiating feature between these two is the carcinomatous and sarcomatous components of MMMTs lack the characteristic merging of the carcinomatous and sarcomatoid elements which is always seen in SSCC. Other differential diagnoses include leiomyosarcoma, fibrosarcoma, rhabdomyosarcoma, malignant fibrous histiocytoma, spindle cell melanoma and metastatic transitional cell carcinoma. The correct recognition of SSCC in such cases depends on the identification of a component of squamous cell carcinoma and also immunopositivity of the spindle cells for cytokeratin.

The immunohistological features of SSCC is very characteristic and help in differentiating this tumour from various sarcomas. Spindle shaped tumour cells of this variant of squamous cell carcinoma are positive for markers associated with both epithelial and mesenchymal differentiation i.e., they co-express cytokeratin and vimentin. These results suggest that sarcomatoid looking cells are derived from a metaplastic alteration of the malignant squamous component. Moreover, since Frank et al showed

that during embryogenesis, cells that express vimentin can be derived from keratin positive cells, vimentin positivity may reflect a reversion to an embryonic pattern, thus justifying the aggressive clinical behaviour of tumour.[17]

Electron microscopic studies show prominent granular elements in the cytoplasm and well formed to poorly formed desmosomal junctions. Thus, electron microscopic studies also point towards epithelial origin of spindle shaped sarcomatoid cells.[18]

SSCC of vulva are staged according to FIGO system used for staging vulvar squamous cell carcinoma.[13]

In early stage disease, lesions are usually treated with radical surgery followed by radiation therapy and chemotherapy. Size, margin status and local tumour biology are the factors which dictate the need for radiotherapy. In more advanced stages, patients presenting with such an extensive disease require surgery which is usually done on a palliative basis, if indicated. These tumours are usually treated with concurrent chemotherapy and radiation.[13]

Positive inguinal lymph nodes are the single most important prognostic factor. The other factors include age, nuclear grade, depth of the tumoural invasion, tumour ploidy. Surgical resection borders are associated with local relapse and recurrence rate is elevated when it is less than 1 cm.[14]

CONCLUSION

SSCC is an extremely rare tumour with a very aggressive course. Due to rarity of this variant, no specific guidelines are available for the management as of now. We believe that larger case series on this variant might help to determine the true biological meaning of the entity and treat the tumour efficiently.

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