CASE REPORT

Sarcomatoid carcinoma of the cervix with foci of malignant melanoma

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ABSTRACT

Introduction: Sarcomatoid squamous cell carcinoma (SSCC) is a rare malignancy of the cervix. Until date around eighteen cases of SSCC have been reported in the literature. It is an aggressive tumor with poor prognosis. The tumor usually presents at an advanced stage. Similarly, primary melanomas of the uterine cervix are rare tumors with not more than 60 cases reported in the world literature. It also has a poor prognosis. There is no reported case of sarcomatoid carcinoma with malignant melanoma. Here, we are presenting a rare case of cervical carcinoma with histopathology suggestive of SSCC with foci of malignant melanoma proven by immunohistochemistry study.
Case Report: The present case report is about a 42-year-old, Mrs. SR, $P_5L_3D_1A_1$, presented with the complaints of intermittent bleeding per vaginum since last 3 years and severe pain in the right lower limb since 1 month. On examination, there was a proliferative growth in the cervix. Her magnetic resonance imaging (MRI) showed cervical mass lesion with right parametrial extension invading the right ureter and bladder wall causing hydroureteronephrosis with contiguous bilateral pelvic nodes. There were multiple lesions in the left femur. Diagnosis of carcinoma of cervix International Federation of Gynecology and Obstetrics stage IIIB with distant metastasis was made. Histopathology report was suggestive of sarcomatoid carcinoma with foci of melanocytic melanoma. She was planned to be treated with palliative radiation.
Discussion: About 90% of cervical carcinomas are squamous cell carcinoma. Adenocarcinoma constitute about 3-4% of all cervical carcinomas. Other rare pathologies are lymphoma, melanoma, sarcoma and metastatic tumors. Our case is a rare combination of sarcomatoid carcinoma with foci of malignant melanoma of cervix.

Key Words: Cervical cancer, chemotherapy, malignant melanoma of cervix, radiotherapy, sarcomatoid carcinoma of cervix

INTRODUCTION

Cervical cancer is the most common cancer in the females in developing countries. The most common histopathology in cervical cancer is squamous cell carcinoma. Other varieties include adenocarcinoma, adenosquamous carcinoma, small cell carcinoma, clear cell carcinoma, carcinosarcoma. Sarcomatoid squamous cell carcinoma of the uterine cervix is an extremely rare histologic entity. Until date, only 18 cases of sarcomatoid carcinoma of the cervix have been reported in the literature.[1-3] The prognosis for women with cervical sarcomatoid carcinomas tends to be worse than that of squamous histology. Patients often present with more advanced stages and follow a very aggressive course of the disease. Primary malignant melanoma of the cervix constitutes a rare disease,[4-7] representing <2% of cases of malignant melanoma that affects the female genital tract.[4] Due to the rarity of the disease, no standard diagnostic and treatment approach is available. Here, we are presenting a rare case of sarcomatoid carcinoma of the cervix with foci of malignant melanoma.

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CASE REPORT

Mrs. SRD, 42-year-old, $P_5L_3D_1A_1$, came to our hospital on 16th July 2012 with complaints of irregular bleeding per vaginum since 3 years. She had severe pain in the right lower limb since 1 month, because of which she was unable to walk and restricted to the bed. On examination, her

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general condition was fair, she was anemic. She had right sided unilateral lower limb edema. There was no evidence of any melanocytic lesion on her body.

On local examination, she had proliferative growth on the cervix that bleeds on touch. Both anterior and posterior fornix was involved with infiltration of upper 1/3rd of the vagina. On per rectal examination the right parametrium was involved up to the pelvic side wall and half of the left para was involved; the rectal mucosa was free. A clinical diagnosis of cervical cancer stage III B was made. Cervical biopsy was taken. On cystoscopic examination, proliferative growth was seen on the posterior wall of the bladder. Biopsy was taken from the bladder growth for histopathological confirmation.

Her hemoglobin was 7.6 g%, liver and renal functions were within the normal limit. Chest X-ray was normal. Contrast enhanced magnetic resonance imaging (MRI) of pelvis showed large T2 hyperintense and T1 isointense cervical mass lesion with right parametrial extension. The mass was abutting the postero-lateral wall of the bladder with invasion of the right ureter, and contiguous bilateral pelvic nodes causing hydroureteronephrosis. There were multiple lesion in the left femur. MRI impression was Ca Cervix International Federation of Gynecology and Obstetrics (FIGO) IV-A with bone mets and bladder invasion [Figures 1-3]. Histopathology was suggestive of sarcomatoid carcinoma of the cervix. Immunohistochemistry was positive for cytokeratin, vimentin, HMB45. Hence the final histo-pathological impression was spindle cell carcinoma with aberrant melanocytic differentiation/malignant melanoma [Figures 4 and 5a-c]. The histology of bladder growth was similar to that of cervix.

The case was discussed in tumor board meeting with radiation and medical oncologists. In view of advanced stage and poor general condition of the patient, decision of only palliative radiation was taken. Single fraction of pelvic radiation with a dose of 10 Gy was given, covering the

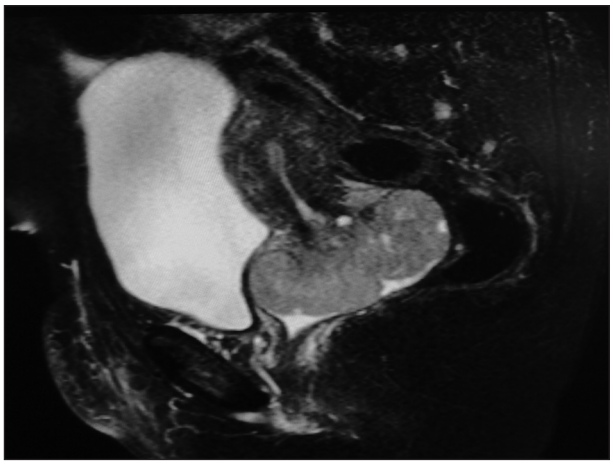

Figure 1: T1 Weighted magnetic resonance imaging showing large cervical mass

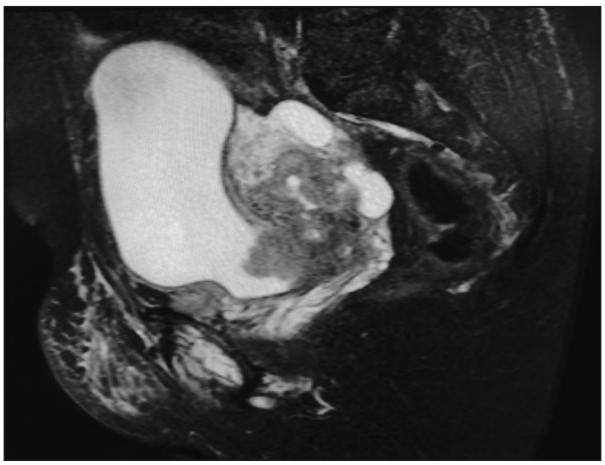

Figure 2: T1 Weighted magnetic resonance imaging showing invasion of the posterior wall of the bladder by the tumor growth

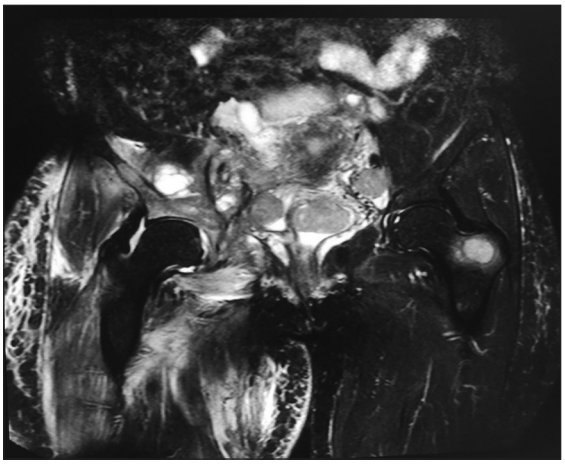

Figure 3: T2 Weighted magnetic resonance imaging showing multiple lesions in the femur suggestive of metastasis

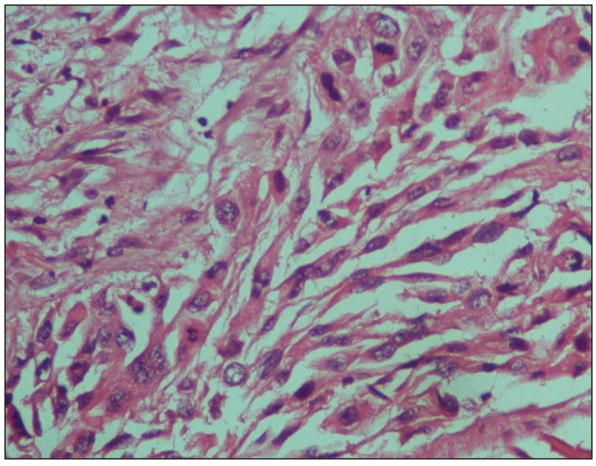

Figure 4: Magnification of tumor showing fascicles of short spindle cells (H and E, ×40)

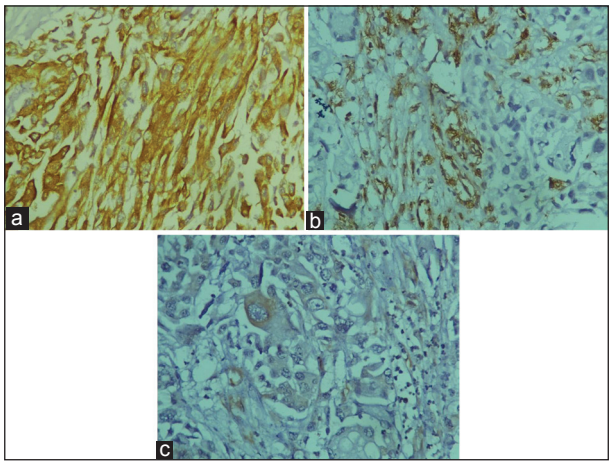

Figure 5: Immunohistochemical staining of tumor: (a) Cytokeratin, (b) vimentin, (c) HMB45 stain — focally positive

soft-tissue tumor and the bony metastasis. The unilateral lower limb edema was managed conservatively by keeping the left lower limb elevated. Patient didn't respond well to the treatment; she had residual disease and succumbed to her disease after 5 months of the diagnosis of the disease.

DISCUSSION

Cervical cancer is the most common cancer in the females in developing countries. The most common histopathology in cervical cancer is squamous cell carcinoma. Squamous cell carcinoma accounts for over 90% of cervical cancer. Adenocarcinoma constitutes about 2-4% of cervical cancer and other histological types, such as carcinosarcoma, lymphoma and sarcoma, account for the rest.

Sarcomatoid carcinoma of cervix (SCC) and malignant melanoma of the cervix both are very rare pathological entities of the female genital tract. In an English literature search for cervical sarcomatoid carcinomas, around 18 cases were found.[1-3] SCC has been described more frequently in the oral cavity, esophagus, pharynx, larynx and skin. Brown *et al*. in their study have reported the largest series of 9 cases of sarcomatoid carcinomas of the cervix with a median disease-free interval of 4.9 months (2-9.5 months). Maximum disease free interval was about 40 months. In this series, although all patients had a complete response to initial therapy, more than half of the patients had recurrences of the disease in <5 months after initial treatments. None of them responded to a second-line therapy.[1]

Nageeti and Jastania reported a case of SCC stage IIIB, treated with radiotherapy and chemotherapy. However, she didn't respond to the treatment and presented with multiple metastasis on follow-up after radiotherapy. She died after 6 months of her primary diagnosis.[3]

Due to the lack of data and recommendations, these patients have been treated according to the treatment guidelines set out for squamous cell carcinoma of the cervix. Radiation therapy (RT) remains the main modality of treatment. RT has been shown to reduce local recurrence and can salvage about 25% of the patients. In view of the poor survival data and the aggressive nature of the tumor, the addition of novel chemotherapeutic agents needs to be explored in this tumor.

Malignant melanomas are generally found in areas of skin exposed to the sun, but can also be present in non-exposed sites. Primary malignant melanoma of the cervix constitutes a rare disease,[4-7] representing <2% of cases of malignant melanoma that affects the female genital tract.[4] Due to that the cervix is an unusual site for this type of neoplasm, the FIGO staging system for cervical cancer is used,[8] rather than the Clark and Breslow scales because the FIGO staging system correlates better with the prognosis.

Norris and Taylor criteria are used[9] to distinguish whether it is a primary malignant melanoma of the cervix:

a. Presence of melanin in the cervical epithelium;
b. Absence of melanoma in another site of the body;
c. Presence of binding activity in the cervical epithelium near the lesion and
d. If metastatic disease is found, it should be according to the cervical carcinoma pattern.

Five-year survival after radical hysterectomy as only treatment is very low: <40% in stage I and 14% in stage II.[4,10] Average survival reported in the world literature of these patients ranges from 6 months to 14 years.

For primary melanoma of cervix surgical approach is the most usual-radical hysterectomy with or without pelvic lymphadenectomy and/or superior vaginectomy.[4,5] There is not enough information about the real role of negative margins in primary melanoma of the cervix. The primary surgery should have the purpose of obtaining negative margins, some authors recommend 2-cm margins as minimum.[11,12] Adjuvant pelvic RT is considered in the case of positive surgical resection margins, when the parametrium is involved, or when lymph nodes are found to be involved.[4,8,13] Adjuvant radiation may improve local control if the surgical margins are close.

No chemotherapy regimens have been reported that substantially may reduce the possibility of recurrence. Dacarbazine is used in advanced disease and it has been observed that up to 20% of patients may have response.[11,13] Combination chemotherapy with cisplatin, bleomycin and vinblastine or dacarbazine with vincristine and carmustine can provoke a better response than the use of solely dacarbazine.[4]

CONCLUSION

Reporting such cases might help clinicians to understand this entity of cervical cancer. We do believe that sarcomatoid carcinoma and melanoma should be considered as a high-grade carcinoma characterized by the development of early progression after initial therapy and failure to respond to second-line therapy, which is why a more aggressive approach at the initial presentation in the form of multimodality treatments should be considered.

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