

Carcinosarcoma of oral cavity: A unique but rare entity

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Abstract

Background and purpose: Carcinosarcoma is an extremely rare biphasic tumor of oral cavity with both malignant epithelial & mesenchymal components. It has carcinomatous as well as specific differentiated sarcomatous elements with hallmarks of malignancy.

Methodology: This is a prospective study of three patients admitted in a tertiary care teaching institutions of Kolkata between June 2014 to May 2015 with a histopathological diagnosis of carcinosarcoma of oral cavity.

Result & Discussion: Age of patients varied between 42 to 68 years. All patients were male and all of them had primary lesion within oral cavity. All the patients had recurrence (one immediately after surgery and two after radiotherapy) and were treated with palliative therapy.

Conclusion: Carcinosarcoma is a rare and aggressive neoplasm with poor survival outcomes. The study concludes surgery as primary modality of treatment and radiotherapy as adjuvant or palliative.

Keywords: Carcinosarcoma, Pseudosarcoma, Spindle cell carcinoma.

Learning points

1. Prognosis of carcinosarcoma is very much dependent on the site of origin.
2. Surgery with optimum preoperative planning and adjuvant radiotherapy remains the mainstay of treatment.
3. The role of chemotherapy is not clear, however, it may reduce the chance of recurrence or metastasis of primarily sarcomatous tissue.

Introduction

Carcinosarcoma is an extremely rare biphasic tumour of oral cavity with both malignant epithelial and mesenchymal components. It has carcinomatous as well as specific differentiated sarcomatous elements with hallmarks of malignancy [1]. Carcinosarcoma of the tongue, lower gingival and floor of the mouth have been reported in the literature. Most of the reported cases originated from major salivary glands; larynx, pharynx, nasal cavity and esophagus being the other common sites. Prognosis of the disease is very much dependent on the site of origin. These aggressive tumours were first described by Virchow in 1864 [2] and Batasakis [3] classified these tumours in three distinct categories –

1. Pleomorphic (spindle cell) carcinoma - biphasic epithelial malignancy with uniphasic or mixed metastases.
2. Carcinoma with pseudosarcomatous stroma - uniphasic epithelial

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malignancy with a benign yet atypical stromal reaction or epithelial malignancy with post- irradiation stromal & epithelial anaplasia, metastases may be uniphasic or mixed.

3. Carcinosarcoma - heterologous malignancy with uniphasic or mixed metastases.

Histogenesis of these complex neoplasms is not clear. Probably they arise out of differentiation of primitive blastic mesenchymal cells that can mature and produce tumours of multiple differentiations [4]. Therefore, they have the potency to spread rapidly to epithelial tissues of lungs, urinary tract, breast and uterus. We report three cases of carcinosarcoma of oral cavity who were managed by multimodality approach in a tertiary care teaching hospital of Kolkata.

Materials and Methods

All the patients of carcinosarcoma of oral cavity presented in our institute (B.R. Singh Hospital and Research Centre, Eastern Railways, Kolkata) over a period of 12 months between June 2014 to May 2015, and were studied prospectively. Details of the cases were reviewed from hospital records of General Surgery, Surgical Oncology and pathology departments. Detailed history, clinical examination formats, imaging like CT scan of head and neck region, chest x-ray, orthopantomogram, metastatic work up and biopsy reports were studied. Parameters like age, sex, site of tumour, primary modalities of treatment, adjuvant treatment, recurrence, follow up and survival data were collected from the Hospital Registry. Histochemistry was performed in all patients for carcinoma and sarcoma panel with special reference to AE1, Vimentin and EMA (Epithelial Membrane Antigen). Patients were followed up monthly for 3 months & thereafter 3 monthly.

Observation and Results

Total three patients were studied with age ranging from 42 to 68 years. All patients were male and all of them had primary lesion within oral cavity - one patient had growth in buccal mucosa extending to lower lip with bony invasion in central arch of mandible, one patient had growth in left lower gingivobuccal sulcus involving lower lip with huge fungating mass below left angle of mouth and one patient had growth in buccal mucosa involving left lower gingivobuccal sulcus with huge fungating mass below left jaw. None of them received neo-adjuvant chemotherapy, all

underwent surgery as a primary modality of treatment. One out of three patients had microscopically positive margin who was treated with re-resection. Two patients received adjuvant radiotherapy and one patient received adjuvant chemotherapy followed by radiotherapy. All the patients had recurrence (one immediately after surgery and two after radiotherapy) and were treated with palliative therapy. Two patients died within one year from the time of diagnosis due to lung metastasis (One after 6 months and one after 11 months). The third patient lost to follow up after 15 months. **(Table 1)**

Review of Literature

Carcinosarcoma is a rare tumour expressing biphasic features (mesenchymal and epithelial) with distinguished hallmarks of malignancy. Virchow first coined the term "Carcinosarcoma" instead of its previous names, pseudosarcoma, spindle cell carcinoma and sarcomatoid carcinoma [4]. Carcinosarcomas of uterus, vagina, lungs, oral cavity, larynx, thyroid, urinary tract and esophagus have been reported. Amongst head and neck region, it commonly involves salivary glands, larynx, oral cavity, hypopharynx, pyriform fossa, sinonasal tract and oropharynx. Largest landmark study on carcinosarcoma of oral cavity was done by Bataskis [3] on 111 patients and Patel [5] using the surveillance, epidemiology and end results (SEER) database.

The exact histogenesis of carcinosarcoma is not known. Two theories have been postulated-the first one proposes multiclonal origin arising from two or more stem cell lines, and the second theory proposes monoclonal origin from a single totipotent stem cell that differentiates in separate epithelial and mesenchymal cell lines [6]. However, the second theory has been supported in most of the immunohistochemical studies. However, many more theories like origin from embryonal rests of epithelial and mesenchymal tissues and tumour de-differentiation have also been found in literature. Histopathologically these tumours are characterized by primitive mesenchymal blastic differentiation into multiple cell lines although true sarcoma remains the main cell line with high mitotic index, cellular pleomorphism, dysplasia and lack of transition zone [7]. On immunohistochemistry epithelial components can be easily identified by cytokeratin and mesenchymal components can be identified by vimentin, S-100, actin, desmin etc (Figure 1 and 3).

Primary surgery followed by adjuvant radiotherapy

Table 1: Detailed particulars of the patients involved in the study.

S. No.	Age	Sex	Primary Site	NACT	Primary Modality of Treatment	Staging	Adjuvant Therapy	Recurrence / Metastasis / Death
1	65	M	Buccal Mucosa + Lower Lip + Bony Invasion in Central Arch of Mandible	No	Surgery	p T4a N2b Mx	CT + RT	Yes (Death After 6 Months)
2	42	M	Lt Lower Gbs + Lower lip + fungating mass below left angle of mouth	No	Surgery	pT4a N1 Mx	RT	Yes (Death After 11 Months)
3	68	M	Buccal Mucosa + Lt lower Gbs + fungating mass below left jaw	No	Surgery	pT4a N1 Mx	RT	Yes (Lost Follow Up After 15 Months)



Figure 1: 1st Patient.



Figure 3: 3rd patient.



Figure 2: 2nd Patient.

remains the cornerstone of treatment for carcinosarcoma. Multimodality therapy is prescribed because of the aggressive and infiltrating nature and course of the disease [8]. Some researchers think that definitive radiotherapy is of no use, however, some researchers consider it a viable option in very advanced inoperable cases. Re-resection and adjuvant radiotherapy is the standard treatment regime for margin positive cases. Adjuvant radiation has been proven to be beneficial in patients with proven nodal metastasis [2]. The role of chemotherapy is not clear in either neo-adjuvant or adjuvant settings, however, it may reduce the chance of recurrence or metastasis of primarily sarcomatous tissue. Conservative surgeries or non-surgical treatment modalities are functionally preservative but have poor prognosis. Till now there is no set treatment protocols for carcinosarcoma.

Carcinosarcoma is known to have grave prognosis. Lethality has been as high as 60% overall and 42% in 30 months as per literature [9]. The outcome has been reported to be influenced by gross morphology and size of the tumour, location, carcinomatous differentiation, depth of invasion and stage of the disease. Local recurrence and metastasis are common. Metastasis is most common in lungs.

Conclusion

Carcinosarcoma is a rare and aggressive neoplasm with poor survival outcomes. The study concludes surgery as primary modality of treatment and radiotherapy as adjuvant or palliative. Re-excision should be offered in patients with positive margin. A standard treatment guideline is highly

solicited for this lethal entity.

Consent/Ethical Approval

Written informed consent was obtained from the patients for publication of this study and accompanying images. A copy of the written consent is available for review by the journal's Editor-in-chief.

Conflict of Interest

The authors declare that they have no competing interests.

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Author's Contributions

All authors approved and equally contributed to the preparation of this final manuscript.

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