Carcinosarcoma of Upper Aerodigestive Tract: A Case Series

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Introduction

Carcinosarcoma is a biphasic tumour composed of both malignant epithelial and mesenchymal components. The initial description of this type of entity was given by Virchow in 1864 [1].

Batsakis [2] described three different categories of this tumour namely:

Pleomorphic (spindle cell) carcinoma: Biphasic epithelial malignancy. Metastasis may be uniphasic or mixed.

Carcinoma with pseudosarcomatous stroma: Uniphasic epithelial malignancy with a benign yet atypical stromal reaction or epithelial malignancy with post irradiation stromal and epithelial anaplasia. Metastasis from either are epithelial only.

Carcinosarcoma: Heterologous malignancy. Metastasis may be uniphasic or mixed.

Materials and Methods

All patients of carcinosarcoma of upper aero digestive tract presented in our institute over a period of 20 months i.e. from January 2010 to August 2011 were studied retrospectively.

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Details of patients were reviewed from hospital records of surgical, radiotherapy and pathology departments (Table 1). Patient evaluation consisted of history, physical examination, computed tomographic scan of head and neck, chest x ray, endoscopic examination and biopsy. Various parameters like age, gender, site of tumour, primary modality of treatment, adjuvant treatment, recurrence and survival were considered. All reports with spindle cell differentiation on histopathology were sent for immunohistochemistry for AE1, Vimentin and Epithelial Membrane Antigen (EMA) markers. Staging was done according to 2010 American Joint Committee on Cancer (AJCC) cancer staging manual [3] (Figs. 1, 2, 3 and 4).

Patients were followed up monthly after suture removal for 3 months and three monthly thereafter. As carcinosarcoma is a rare neoplasm, our institute does not have any fixed treatment protocol for the same and patients are referred for surgery, radiotherapy or neoadjuvant chemotherapy at the discretion of the treating clinician.

Also the article reviews 3 out of very few studies on this neoplasm.

Observation and Results

Total of 11 patients were considered. Patients were in the age group of 45 to 68 years with mean age of 58.18 years. Out of 11 patients 9 were males and there were 2 females.

Six out of 11 patients had their primary lesion in the oral cavity, two in oropharynx, two in larynx and one patient presented as metastasis of unknown origin of neck where primary lesion could not be identified on triple endoscopy and CT scan. Neoadjuvant chemotherapy was given in one patient of oral cavity [4] however the tumour did not respond to it and the patient was later operated. Seven patients underwent upfront

Fig. 1 Clinical photograph (Histopathology showed carcinosarcoma)

surgery including the one with neoadjuvant chemotherapy. Four patients received radiotherapy as the primary treatment. Ten patients in the series had stage 4 disease and remaining one patient had stage 3 disease. Two patients out of seven patients who underwent surgery had microscopic positive surgical margins. In both these patients complete re excision of surgical margin was done. All the seven patients who underwent surgery received adjuvant treatment. Six patients received adjuvant radiotherapy and one patient received adjuvant chemo radiotherapy.

There were three recurrences in the series, two after radiotherapy and one after surgery. These patients were referred for palliative chemotherapy and all died in the course of 1 year from the time of diagnosis. Two patients were lost to follow up after their primary treatment. Remaining patients i.e. six patients are still following up and are disease free. The longest follow up in the series is 18 months and the shortest follow up is 2 months.

Discussion & Review of Literature

The largest landmark study on this topic has been that of Bataskis [2], where he studied 111 patients of carcinosarcoma. However his study was more confined to the biological

Fig. 2 Low power view: Carcinosarcoma

Fig. 3 Immunohistochemistry slide showing AE 1 positivity

behaviour of the tumour and did not elaborate much on the management of the disease.

In his series of 111 patients 13 patients had primary disease of sinonasal tract, 7 had primary disease in the pharynx and remaining majority of 65 had disease in larynx. The time of survival in these three groups was 6–30 months, 8 months and 4–24 months respectively from the time of diagnosis. 27 presented with nodal metastasis at the time of initial presentation. 36 of 102 determinate cases died as a consequence of their malignancy. The term determinate was used to refer to patients in whom death was directly related to the neoplasm and excludes mortality due to unrelated causes.

In another Canadian article by Berthlet et al. [1] overall survival, disease free survival and local control above clavicles were studied. There were 17 patients in the study with median follow up of 29 months with median survival of 32 months. The study concluded overall survival advantage for patients with early stage disease, extra laryngeal presentations and patients treated with surgery. Study also highlighted optimum therapy for the disease as surgery appears superior to radiotherapy, irradiation constitutes an acceptable alternative for inoperable patients or those with sinonasal tumours. It also indicates the benefit of adjuvant radiotherapy in cases with positive surgical margins and patients with nodal metastasis. The study also supported the role of primary radiotherapy as a laryngeal preservation procedure with surgery reserved for failures of radiotherapy.

Gorsky et al. [5] studied 139 patients of head and neck and intra oral soft tissue sarcomas and reported only one case of carcinosarcoma in their series. The patient had the disease in base of tongue and was treated with chemo radiotherapy and died 8 year later of leukaemia.

Apart from these articles there are number of anecdotal case reports on carcinosarcoma [6–8] of various sites in oral cavity however there is no set treatment guideline.

In our case series mean age of the patients was 58.18 years which is much below the western data of about 70 years. Most common site of presentation was the oral

Fig. 4 Immunohistochemistry slide showing Vimentin positivity

cavity which is again different from the western data where laryngeal carcinosarcomas are more common. We had no patients presenting early in the course of the disease, all our patients were in stage 3 or 4 of the disease. Nine out of 11 patients i.e. 81.8% in our series presented with positive neck

nodes whereas Bataskis [2] reported only 24% of patients with nodal disease.

Also Berthlet et al. [1] highlights the importance of adjuvant radiotherapy in margin positive cases and in our series surgical re excision was done to take care of positive

Table 1 Details of patients included in the series

Serial No.	Age (years)	Gender	Primary site	Neo adjuvant treatment	Primary modality of treatment	Staging	Adjuvant treatment	Recurrence
1	45	Male	Lower Alveolus	Nil	Surgery	pT4aN1R0(Stage 4a)	CT+RT	Nil
2	54	Female	Lower GBS	Chemotherapy	Surgery	ypT4aN1R0(Stage 4a)	RT	Nil
3	60	Male	Base Of Tongue	Nil	RT	cT4aN2(Stage4a)	Nil	Recurrence
4	55	Male	RMT+BM	Nil	Surgery	pT3N2bR1 (Stage 4a)	RT	Nil
5	65	Male	RMT+Soft Palate	Nil	RT	cT4bN1(Stage4b)	Nil	Lost to follow up
6	68	Male	Base Of Tongue	Nil	RT	cT4bN1(Stage4b)	Nil	Recurrence
7	64	Male	Central Arch	Nil	Surgery	pT3N0R0 (Stage 3)	RT	Nil
8	60	Male	Supraglottis	Nil	RT	cT3N2a (Stage 4a)	Nil	Lost to follow up
9	58	Male	Glottis	Nil	Surgery	pT2N1R0 (Stage 4a)	RT	Nil
10	55	Female	Lower Alveolus	Nil	Surgery	pT4aN0R1 (Stage 4a)	RT	Nil
11	56	Male	MUO Neck	Nil	Surgery	pN2b	RT	Recurrence

surgical margins. However these patients received radiotherapy with respect to their overall disease stage.

Conclusion

Due to small number of patients in each subgroup the inferences derived may not be statistically significant. Oral cavity seems to be a more common site in Indian patients with a younger age of presentation. As with any other malignancy relatively higher stage of the tumour is encountered in our part of the world.

The authors are in complete agreement with other studies that surgery being superior to radiotherapy as primary treatment, irradiation constitutes an acceptable alternative for inoperable patients or those with sinonasal tumours. However operability is a very subjective criteria as what is resectable for one surgeon may not be the case for another [4]. In light of available studies, surgery followed by adjuvant radiotherapy is the best treatment modality for patients of carcinosarcoma of upper aerodigestive tract.

We emphasize on the role of re-excision in patients with positive surgical margins. Nonetheless, these issues can only

be settled with randomized controlled trials with larger number of patients. Rarity of the condition is the major obstacle.

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