Carcinosarcoma of Head and Neck: Clinicopathological Characteristics, Treatment and Outcomes of Unique Neoplasm

Abstract

Background: Carcinosarcoma of the head and neck is a rare neoplasm with incidence of <1%. This is a biphasic tumor with epithelial and mesenchymal components. Rarity of this neoplasm is a major obstacle to establish proper guidelines. Only few case series and reports are available.

Aim: The aim of this study was to evaluate clinicopathological characteristics, treatment and its outcomes to contribute in part to the better understanding and awareness of this rare malignancy.

Material and Methods: Retrospectively data of histologically proven carcinosarcoma of head and neck from January 2013 to December 2015 was retrieved and evaluated for various parameters and also compared with both Indian and western data.

Results: Total of 5 patients of carcinosarcoma were registered within selected timeframe. The mean age of 44.8 years ranged from 27 years to 68 years. Male predominance was reported with M:F ratio 4:1. Oral cavity is the most common site of tumor followed by oropharynx. Surgery is the primary modality of treatment followed by adjuvant radiotherapy as per indication. Shortest follow up was 11 month and longest was 31 months. Only one patient was presented with recurrence.

Conclusion: The diagnosis of carcinosarcoma is challenging so histopathology along with immunohistochemistry is needed for appropriate diagnosis. Oral cavity seems to be the most common site with younger age group in the present study which is supported by other Indian studies. Future studies are needed to define the geographical variation with clinicopathological character as well as treatment protocol for this relatively rare entity.

Keywords: Carcinosarcoma; Head and neck; Young age; Treatment; Outcomes

Introduction

Carcinosarcoma is a rare (incidence of <1%) and unique variant of squamous cell carcinoma [1]. These tumors are biphasic and have been proved by many studies that it evolves from conventional squamous carcinoma with dedifferentiation associated with sarcomatoid transformation [2,3]. Most commonly it arises from larynx, nasal cavity, hypopharynx, esophagus, trachea, oral mucosa. It has a male predominance with mean age of presentation in sixth decades of life with history of smoking, alcohol abuse, and radiation exposure [4,5]. Immunohistochemistry along with routine histopathology is essential in establishing the diagnosis of spindle cell carcinoma. Here we are reporting five cases of spindle cell carcinoma in head and neck region to the better understanding and awareness of rare malignancy.

Aim

The aim of this study was to evaluate clinicopathological characteristics, treatment and its outcomes of patients of carcinosarcoma of head and neck to contribute in part to the better understanding and awareness of this rare malignancy.

Material and Methods

The data of patients of carcinosarcoma of head and neck registered in Department of Radiotherapy, Safdarjung Hospital, Delhi, India from January 2013 to December 2015 was retrieved retrospectively. The various parameters were evaluated and compared with both Indian and western data.

Observation and Results

Total of 5 patients of carcinosarcoma were registered. Patients were in age group of 27 to 68 years with mean age of 44.8 years. Four patients were male and one was female. Three out of 5 patients had primary tumor site in oral cavity, one in oropharynx and one patient had primary lesion in larynx. 60% of cases were Stage III and 20% each of Stage II and Stage IV. Three patients had upfront surgery followed by adjuvant radiotherapy and two patients received radical radiotherapy as primary treatment.
There was one recurrence after 1 year of completion of treatment. One patient was lost to follow up. Three patients were on regular follow up and disease free. Follow up period ranged from 11 to 31 months (Table 1).

### Table 1: Patients clinicopathological characteristics, treatment and outcomes.

| No | Age | Habit          | Gender | Presenting Complaints                  | Primary Site       | Treatment                                                                 | Staging                        | Follow up                  |
|----|-----|----------------|--------|----------------------------------------|--------------------|---------------------------------------------------------------------------|--------------------------------|----------------------------|
| 1  | 27  | Tobacco and Bidi | Male   | Swelling left tonsil and pain           | Left Tonsil        | Left Tonsillectomy f/b RT 66Gy/33#                                      | cT2N1M0 [Stage II]            | On Regular Follow Up 31 months |
| 2  | 27  | Tobacco and Bidi | Male   | Ulceroproliferative growth of buccal mucosa | Buccal Mucosa +RMT | Radical radiotherapy with chemotherapy 70 Gy/35#                            | cT3N0M0 [Stage II]            | Lost to follow up After treatment |
| 3  | 40  | Bidi           | Male   | Ulcer on right lateral border of tongue | Right Lateral Border Of Tongue | Surgical Excision With Neck Dissection                                   | cT3N1M0 [Stage III]          | On Regular Follow Up 11 months |
| 4  | 62  | Tobacco        | Male   | Difficulty and pain during swallowing | Supraglottis       | Radical radiotherapy with chemotherapy 70Gy/35#                            | cT3N2bM0 [Stage IVA]          | Recurrence                 |
| 5  | 68  | Tobacco and Bidi | Female | Swelling lower alveolus                | Lower Alveolus     | Surgery f/b Radiotherapy with chemotherapy 60Gy/30#                       | cT3N0M0 [Stage III]            | On Regular Follow Up 18 months |

### Discussion & Review of Literature

Carcinosarcoma are rare variant of squamous cell carcinoma [1]. It consists of epithelial and mesenchymal components hence diagnosis is challenging and complex. Monoclonal hypothesis has been proved by many studies that both components have the same monoclonal origin with transformation and dedifferentiation of epithelial cell to spindle cells supported by immunohistochemistry [2,3]. It usually occurs in males during 6th and 7th decades of life. Similar to other studies [4,5], male predominance was reported in this study with M:F ratio 4:1. Mean age was 44.8 years this is much below when we compared from western data of about 60 to 70 years. But Indian study by Viswanathan S et al. [2,3] showed mean age of presentation was 53 and 58.18 years respectively comparable to present study. Predisposing factors are smoking, alcohol consumption, previous radiation exposure [5]. All 5 patients were chronic tobacco chewer and bidi smoker. Commonly it arises from larynx, nasal cavity, hypopharynx, oral cavity is rare site of presentation [1]. In present study most common site of primary tumor was oral cavity contrary to western data. Here tobacco chewing is more common habit rather than smoking this might be the probable cause of increase incidence of oral cavity carcinosarcoma [5,6]. None of the patients had prior history of exposure to radiation. 60% patients were of stage III and 20% each of stage II and stage IV. One patient (20%) presented with nodal metastasis. The largest landmark study by Batakis et al. [3] showed nodal metastasis of 24%. Carcinosarcoma is treated same as squamous cell carcinoma of head and neck there is no different established guidelines for this relatively rare entity.

### Conclusion

Carcinosarcoma is a rare neoplasm with biphasic features, composed of both malignant epithelial and mesenchymal component. In this study oral cavity is the most common site of presentation with younger age group. Though the data is very small so the statistical significance could not achieved but supported by other Indian studies where the similar finding was observed. So future studies are needed to define the geographical variation with clinicopathological character as well as treatment protocol for this relatively rare entity.
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