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SARCOMATOID CARCINOMAS OF HEAD AND NECK: OUR EXPERIENCE WITH REVIEW OF LITERATURE

Oncology

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ABSTRACT

Sarcomatoid carcinoma of head and neck, a subtype of squamous cell carcinoma is a unique and rare neoplasm, composed of both malignant epithelial and mesenchymal components. A series of 20 patients with Sarcomatoid carcinoma is presented, along with an analysis of potential prognostic factors, outcome following treatment, and patterns of failure. Details of patients were reviewed from hospital records of surgical, radiotherapy and pathology departments.

KEYWORDS

Head and Neck, Sarcomatoid Carcinoma, Squamous Cell Carcinoma

INTRODUCTION

Sarcomatoid carcinoma of head and neck, a subtype of squamous cell carcinoma is a unique and rare neoplasm, composed of both malignant epithelial and mesenchymal components. Sarcomatoid carcinoma (also known as pseudosarcoma, carcinosarcoma ,pseudosarcomatous squamous cell carcinoma, pleomorphic carcinoma, and spindle cell carcinoma) is a highly malignant tumor characterized by dual malignant histologic differentiation of the epithelial component consisting of a focally squamous cell carcinoma and a mesenchymal component having a sarcomatoid stroma. Carcinosarcoma may arise in any squamous epithelium of the body, however its occurrence is extremely rare in the sinonasal cavity. These tumors have now been proved to be monoclonal, evolving from conventional squamous carcinoma with dedifferentiation associated with sarcomatoid transformation. They pose a significant diagnostic challenge to the pathologist with remarkable morphological and immuno histochemical overlap with other benign and malignant spindle cell tumors .An accurate diagnosis of these tumors is essential as they vary in their clinical management and outcome. This study is an overview of the spectrum of a large number of sarcomatoid carcinomas arising in all head and neck mucosal sites to evaluate their clinicopathologic, detailed morphological and immunohistochemical characteristics and problems in differentiating them from other spindle cell tumors commonly occurring at these sites.

METHODOLOGY:

A series of 20 patients with Sarcomatoid carcinoma is presented, along with an analysis of potential prognostic factors, outcome following treatment, and patterns of failure. Details of patients were reviewed from hospital records. Patient evaluation consisted of history, physical examination, CECT 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. H &E stained slides of all the cases were assessed. All reports with Sarcomatoid differentiation on histopathology were sent for immunohistochemistry for AE1, Vimentin and EpithelialMembraneAntigen (EMA) markers.

RESULTS:

Total of 20 patients was considered. Patients were in the age group of 32 to 76 years. Out of these 17 were males and 3 female. Sixteen out of 20 patients had primary lesion in the oral cavity, two in oropharynx and two in the larynx. Thirteen patients underwent upfront surgery, one patient had partial response to neoadjuvant chemotherapy and subsequently underwent surgery, three patients had disease progression on chemotherapy and three patients received radiotherapy. Three patients out of 14 patients who underwent surgery had microscopic positive surgical margins. In both these patients complete re-excision of surgical margin was done. All the patients who underwent surgery received adjuvant treatment, out of which ten patients received adjuvant radiotherapy and four patients received adjuvant chemoradiotherapy. There were six recurrences in the series, two after radiotherapy and four after surgery.These patients were referred for palliative chemotherapy and all died in the course of 1year from the time of diagnosis.3 patients with disease progression on chemotherapy were considered for palliative chemotherapy. 3 patients were lost to follow up after their primary treatment. Remaining patients i.e. 8 patients are still following up and are disease free. The longest follow up in the series is 48 months and the shortest follow up is 7months.

DISCUSSION.

Spindle cell carcinoma is an uncommon tumour accounting for 3% of SCC [1]. In the past SpCC has been thought to be a collision tumour between carcinoma and sarcoma. Virchow was the first person to introduce the term carcinosarcoma for this group of lesions. Kettle E. H., in the beginning of 20th century described that many authors were of the opinion that the spindle cell component is actually epithelial cells being transformed into mesenchymal cells. Numerous hypotheses regarding the histogenesis of SpCC have been proposed. Three dominant pathogenetic theories have been proposed: the tumor 1) represents a "collision tumour" (carcinosarcoma), 2) is a squamous cell carcinoma with an atypical reactive stroma (pseudosarcoma), or 3) is of epithelial origin, with "de-differentiation" or transformation to a spindle cell morphology (sarcomatoid carcinoma) [3]. Recently, the third hypothesis has been supported by following evidences: their occurrence in the exact sites that normally have squamous epithelium and a preponderance of carcinomas rather than sarcomas; a superficial location; a polypoid appearance; the direct continuity and smooth transition of the spindle cells with areas of squamous epithelium; immunoreactivity with epithelial antigens; a dual expression of epithelial and mesenchymal differentiation with double labeling techniques in some neoplastic spindle cells; and the presence of epithelial only, sarcomatous only, or a duality of expression in metastatic deposits from laryngeal sarcomatoid carcinoma [4]. The mean age of diagnosis of Spindle cell carcinoma is 57 years. However it can be diagnosed in younger age group and very old age group (range 29 - 93 yr). As far the gender predilection is concerned most authors reported that there is equal frequency in both males and females. Some studies have reported a male predilection. Site predilection for SpCC are the lower lip, tongue and alveolar ridge or gingival although most tumours in head and neck region occur in the larynx [6]. Radiation, trauma, tobacco use or alcohol consumption seemed to play a role in etiological factors [7]. Spindle cell carcinoma can demonstrate varied histopathological appearance from case to case or within different areas of the same tumour tissue. The cells may appear like epithelial cells or may appear as atypical mesenchymal cells to add to the confusion. A histopathologist using a light microscope may find it extremely difficult to assess the case as it may appear as a carcinoma as well as a sarcoma. The tumour generally consists of fascicles of anaplastic spindle cells with considerable number of mitotic figures [8]. Differential diagnosis includes a number of benign and malignant tumours, such as fibromatosis, nodular fasciitis, reactive epithelial proliferations, squamous cell carcinoma, fibrosarcoma, malignant fibroushistocytoma, leiomyosarcoma, rhabdomyosarcoma, malignant peripheral nerve sheath tumour, mesenchymal chondrosarma and malignant melanoma [4]. The treatment in case of SpCC has been debated. Many are in the opinion that wide excision which includes

radical dissection along with metastasis is adequate. Some authors are of the opinion that surgery alone is not sufficient in that radiotherapy is mandatory along with surgery. Unfortunately 30% of all oral cases, most cases were ending fatally within one year. This is similar in connection with the high grade SCC. Presence of metastasis signals a poor prognosis as per study conducted by Ellis and Corio and this applicable in cases of oral cavity [9]. In a study out of total 15 patients of SpCC of oral cavity and oropharynx receiving surgery, recurrence rate was 73.3% and metastasis rate 33.3% was reported [10]. Immunohistochemical characterization of tumour cells using antibodies to keratin, vimentin, and S-100 protein is very helpful in differentiating SpCC from true spindle cell sarcoma, melanoma and malignant myoepithelioma.IHC analysis of our cases show co-expression of cytokeratin (CK) and vimentin to various degrees. In our study Pan cytokeratin has been used which includes wide range of cytokeratin antibodies.The largest landmark study on this topic has been that of Bataskis[2] where he studied 111 patients of carcinosarcoma. 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. In another Canadian article by Berthlet[3]et al. 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. Gorsky et al. 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 chemoradiotherapy and died 8 year later of leukaemia .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 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. This study is in consensus with studies of Bataskis[2], Berthlet et al[3], Gorsky et al[4] etc .Surgery appears superior to radiotherapy in management of primary disease, irradiation constitutes an acceptable alternative for inoperable patients or those with sinonasal tumours. It also indicates the benefit of adjuvant radiotherapy in cases of advanced disease, with positive surgical margins and patients with nodal metastasis. Prognosis of the disease is generally poor and distant metastases are more often than conventional squamous cell carcinoma. It poses a diagnostic dilemma when confronted with malignant tumor with spindle cell morphology in the head and neck region and it's more aggressive behavior as compared to classical squamous cell carcinoma warrants surgical interventions with wider surgical margins[5,6,7].

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 criterion as what is resectable for one surgeon may not be the case for another. 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 be settled with randomized controlled trials with larger number of patients. Rarity of the condition is the major obstacle

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