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Sarcamatoid Carcinoma of the Hard Palate- A Rare Second Primary with Aggressive Behaviour: A Case Report

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ABSTRACT

Sarcomatoid carcinoma is an aggressive variant of squamous cell carcinoma. It occurs more commonly in previously irradiated cases of squamous cell carcinoma. The objective of this study is to develop an understanding of the importance of this aggressive and rare tumor of the head and neck, which has the potential of local invasion and distant metastasis.

Keywords: Sarcomatoid Carcinoma, Squamous Cell Carcinoma, Oral Cavity

*See End Note for complete author details

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INTRODUCTION

Sarcomatoid or spindle cell carcinoma (spCC) of the head and neck, a rare subtype of squamous cell carcinoma (SCC), is a biphasic neoplasm known for its more aggressive behavior compared to the classical SCC. Common sites in head and neck includes oral cavity followed by larynx, oropharynx / hypopharynx and maxilla. In oral cavity, it mostly affects gingivobuccal complex. It seldom presents as a palatal growth.[1]

CASE REPORT

Hereby we present a case of Sarcomatoid carcinoma which came as a second primary in the region of hard palate. Our patient was a 46-year-old male **(figure 1),** with history of smoking, alcoholism and pan chewing, with no known comorbidity.

He was a previously diagnosed and treated case of moderately differentiated squamous cell carcinoma (SCC) of the right lower gingiva, staged IVA (cT3N2b), in September 2022. He underwent neoadjuvant chemotherapy (1 cycle of cisplatin + 5 fluro uracil and 1 cycle of carboplatin + 5 fluro uracil) followed by right hemimandibulectomy and right modified radical neck dissection (levels 1 to 5) from Regional Cancer Centre, Thiruvananthapuaram. Reconstruction was done using a right pectoralis major myocutaneous (PMMC) flap.

Figure 1. 46 yr old patient with diagnosed and treated case of carcinoma lower GBS right with hard palate growth

Table 1. Timeline regarding brief summary of the case	
Diagnosed as case of squamous cell carcinoma –lower gingiva	august 2022
Underwent neoadjuvant chemotherapy followed by right hemimandibulectomy and right modified radical neck dissection	September 2022
Growth in the hard palate	March 2023
Diagnosed as sarcamatoid carcinoma hard palate	March 2023
Patient started on palliative chemoradiotherapy	April 2023
Underwent emergency tracheostomy	April 2023
Succumbed to death	May 2023

Postoperatively, he got adjuvant radiotherapy of 60 Gy in 30 fractions. He was on regular follow up for the first few months post surgery from RCC.

Six months following radiotherapy, the patient presented to our OPD with a painless growth on the hard palate with halitosis and dysphagia. Patient also had recurrent episodes of bilateral nasal block and occasional bleed from the oral cavity. On examination, grade 3 trismus was noted. A proliferative growth was seen arising from the hard palate extending into the right upper gingival area and posteriorly towards the oropharynx. The lesion was tender to palpation. There were multiple missing teeth in the oral cavity. There was no history of epistaxis, or nasal discharge. Flexible nasopharyngolaryngoscopy was done and it showed a proliferative growth involving the superior surface of soft palate extending downwards, obscuring the view of larynx & hypopharynx.

MRI of the neck **(figure 2, 3)** revealed a heterogeneously enhancing lesion involving the hard palate and soft palate, measuring 6.3 x 6 x 7.2 cm, with surrounding extension, laterally up to the subdermal aspect of face **(figure 2, 3),** invading floor of mouth and right retromolar region. Medially the lesion crossed the midline. Posteriorly involving retropharyngeal space, parapharyngeal space and para vertebral space with carotid artery encasement, and intracranial involvement. Superiorly the lesion extended into the pterygoid muscles, right maxillary sinus. In the nasopharynx, the lesion invaded the right torus tubarius and fossa of Rosenmuller. Inferiorly the lesion to the right valleculae and glosso- epiglottic fold. Metastatic cervical lymph nodes were noted involving the right level 1&2 regions.

Biopsy was taken from the growth and the histopathology report **(figure 4,5)** came as sarcamatoid carcinoma -malignant spindle cell neoplasm, which was P63 positive **(figure 6).** All other markers of S100, desmin, CD45, CD31, and CD34 were negative for the tumor cells.

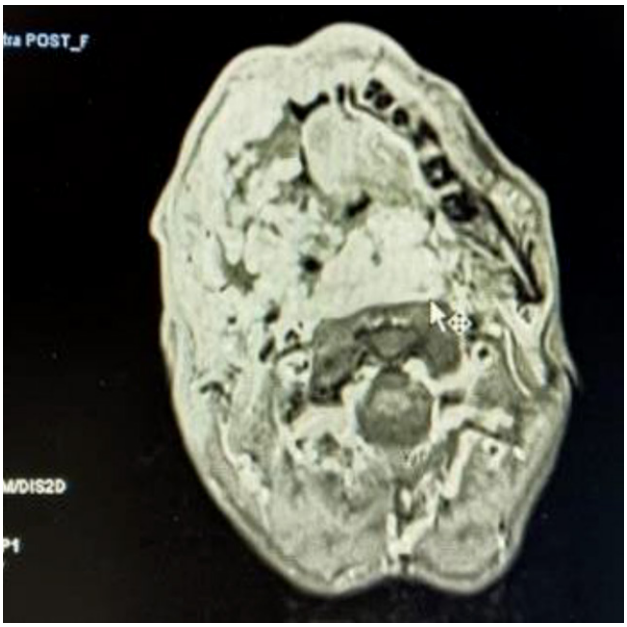

Figure 2. T1weighted MRI neck (axial) with contrast showing heterogeneously enhancing mass lesion in the area of hard palate and soft palate with extension

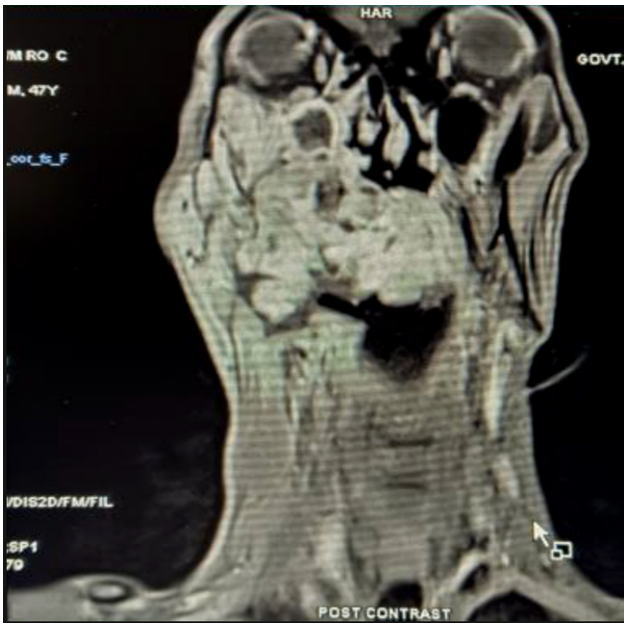

Figure 3. T1Weighted MRI neck with contrast (coronal) showing heterogenous;y enhancing lesion involving right oral cavity with carotid encasement

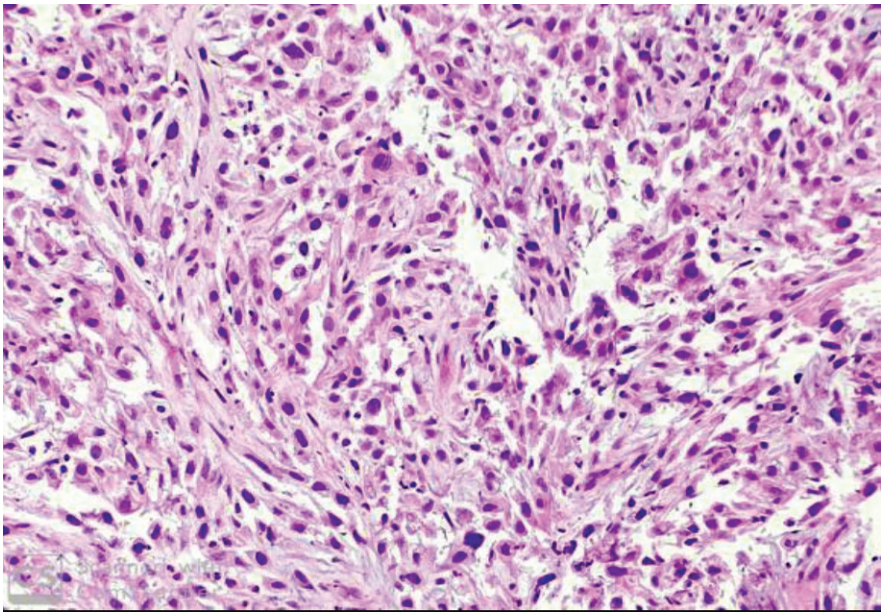

Figure 4. Low power view: malignant spindle cells

The patient then developed respiratory distress and stridor, underwent emergency tracheostomy. Due to the rapidly progressive and extensive nature of the disease, surgical intervention was unfeasible (due to carotid encasement and poor general health of the patient). He was subsequently started on palliative chemo radiotherapy with gemcitabine and 5 cycles of radiotherapy. Patient succumbed to death within 2 weeks following initiation of palliative treatment.

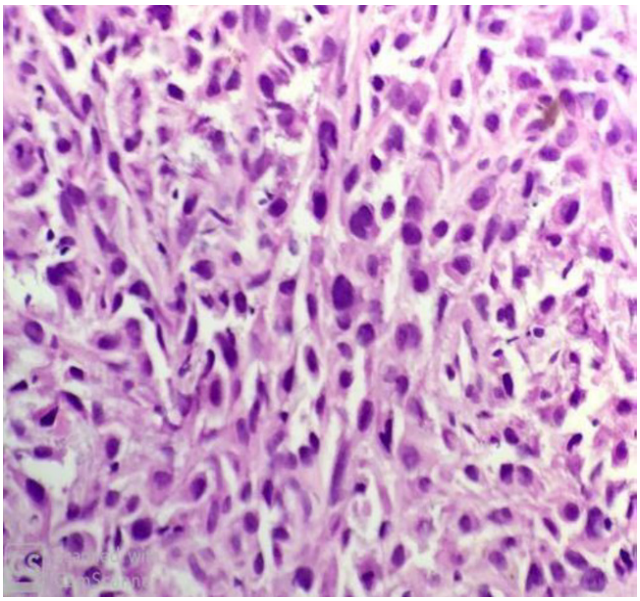

Figure 5. High power view: sarcomatoid tumor

DISCUSSION

The hard palate is a rarer site for primary malignancies and can have varying pathologies. The sarcomatoid variant of SCC is a rare biphasic type characterized by dysplastic surface squamous epithelium and a stromal component composed of invasive spindle cells.[2] It is also known as spindle cell carcinoma, psuedosarcoma, carcinosarcoma, collusion tumor or pleomorphic carcinoma. Spindle cell carcinoma (SpCC) is a variant of squamous cell carcinoma characterized by spindled or pleomorphic tumor cells that resemble a true sarcoma but are of epithelial origin.[3]

They comprises less than 1% of all head and neck malignancies and less than 3 % of all head and neck squamous cell carcinomas.[3] The larynx, especially the glottis, is the most frequent primary site, followed by the oral cavity, which includes the tongue, floor of the mouth, and gingiva. Less common sites include the hypopharynx, hard palate, oropharynx, paranasal sinuses, and nasal cavity.[4] It appears as an exophytic, ulcerated, and proliferative polypoid growth with shaggy exudates, although sessile, nodular, or endophytic forms have also been observed.[1]

Studies indicate that both the conventional SCC and its sarcomatoid components originate from a single stem cell. This aggressive variant is known for its invasive growth, high rates of local recurrence, and distant metastasis, all of which lead to a poor prognosis.[1] One hypothesis suggests that sarcomatoid carcinomas may arise from the transformation of a specific subset of poorly differentiated squamous carcinomas. Epithelial differentiation in the tumor shows considerable variability. The epithelial component experiences a gradual phenotypic shift, adopting a mesenchymal differentiation pathway that results in a spindle shape, loss of cellular polarity, formation of a mesenchymal matrix, and increased vimentin expression alongside a decrease in keratin expression.[5]

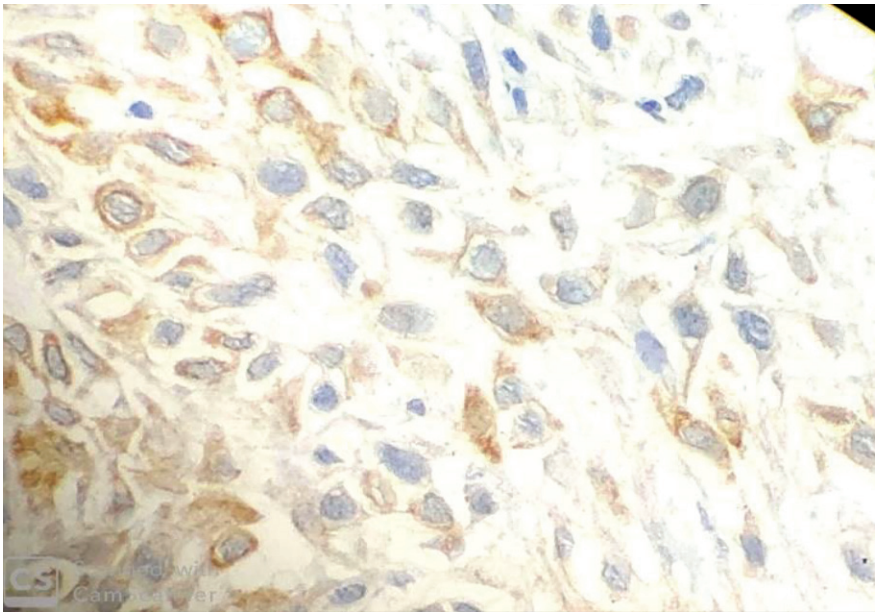

Figure 6. IHC showing P63 positivity

Sarcomatoid carcinoma can occur across a wide age range, with incidence increasing with age and a higher occurrence in males. Other risk factors include older age, alcohol use, any form of tobacco consumption, and prior radiation therapy.[5] The histological appearance can differ between cases and even within different areas of the same tumor. Epithelial cells are known to undergo a range of progressive phenotypic changes, gradually adopting characteristics of mesenchymal differentiation. This transition involves a shift to a spindle-shaped morphology, loss of cellular polarity, production of mesenchymal matrix components, and a molecular switch marked by the gain of vimentin expression and the loss of keratin. This phenotypic plasticity, reminiscent of epithelial-to-mesenchymal transition (EMT) seen in embryogenesis, is characterized by reduced intercellular cohesion, cell elongation, basement membrane degradation, connective tissue (collagen) production, and the capacity to invade the surrounding stroma.[6]

The cells may resemble epithelial cells or atypical mesenchymal cells. The tumor is composed of fascicles of anaplastic spindle cells.[7] The morphology of the spindle cells in sarcamatoid carcinoma cannot be just predicted by routine light microscopy but requires the use of immunohistochemistry (IHC).[7] Immunohisto-chemistry (IHC) can be helpful in diagnosing mucosal squamous cell carcinoma in the head and neck by identifying markers of epithelial differentiation, such as pan-CK, EMA, or p63.

Overall, the mortality rate associated with sarcomatoid carcinoma is higher than that of conventional squamous cell carcinomas. Sarcomatoid carcinoma has a worse prognosis, even with aggressive surgical treatment and accompanying adjuvant therapies.[5] The management is as complex and debated as its diagnosis. The most favored and effective treatment approach appears to be wide surgical excision, potentially accompanied by radical neck dissection. While many authors regard radiotherapy as largely ineffective, it is considered an acceptable option for patients who cannot undergo surgery, those with positive surgical margins, or those with nodal metastasis.[8]

Overall, 5 year survival rates are poorer as compared to conventional squamous cell carcinoma. Benniger et al[9] and Su et al,[10] in two retrospective studies, concluded that spindle cell carcinoma (SC) of the oral cavity exhibits more aggressive behavior than other forms. Despite aggressive treatment, the 5-year survival rate for splindle cell carcinoma remains lower compared to conventional squamous cell carcinoma (SCC), with SC showing a survival rate of 32% versus 45% for SCC. Reflecting this aggressive nature, a combined approach of surgery and radiotherapy was utilized in 63.6% of SC cases, compared to 40.4% in conventional SCC . Given that many SC cases are unresectable, palliative therapy often becomes the preferred treatment option.

Studies have shown that over half of patients with sarcomatoid carcinoma display abnormal expression of the PD-L1 protein, suggesting they may respond well to immune checkpoint inhibitors like Pembrolizumab. Consequently, these inhibitors, along with targeted therapies, have emerged as promising treatment options which can downstage the tumour and can make the tumour surgically operable.[11]

CONCLUSION

Sarcomatoid carcinoma of the head and neck, a subtype of squamous cell carcinoma, is a rare biphasic neoplasm (constituting less than 1 % of all head and neck cancers). The tumor simulates a true sarcoma but is of epithelial origin. Mucosal trauma, alcohol and tobacco consumption, prior radiation are considered as the etiological factors. They have poorer survival rates due to the invasive nature of the tumor and its metastatic potential as well as poor response to conventional radiotherapy. So, prompt diagnosis and treatment are necessary to address significant morbidity and mortality.

END NOTE

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