

Case Report

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Unveiling the enigma: A rare case of spindle cell carcinoma of the tongue with a comprehensive literature review**Renu Singh¹; Arun Kumar Yadav^{1*}; Tabassum Samani¹; Deepa Rani¹; Lalit Raj²; Ashok Kumar Arya¹**¹Department of Radiation Oncology, Sarojini Naidu Medical College, Agra 282002, Uttar Pradesh, India.²Department of Pathology, Sarojini Naidu Medical College, Agra 282002, Uttar Pradesh, India.***Corresponding Author: Arun Kumar Yadav**Department of Radiation Oncology, Sarojini Naidu
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Abstract

Spindle cell carcinoma with <1% incidence in the oral cavity is a rare and aggressive malignancy. They are usually biphasic and affect males more than females in the sixth and seventh decades, with a mean age of 58 years. The tongue, buccal mucosa, and gingiva are the three most typical sites for spindle cell carcinoma in the oral cavity. These tumours typically present as painless, slow-growing masses that are often mistaken for benign lesions, leading to delayed diagnosis and treatment. Due to their aggressive nature, early detection and intervention are crucial for improving patient outcomes. While spindle cell carcinoma may have a low incidence rate in the oral cavity, its rarity does not diminish the seriousness of the disease or the need for timely detection and treatment. Additionally, the fact that these tumours are often mistaken for benign lesions underscores the importance of raising awareness and vigilance among healthcare providers and patients. The index patient had a history of risk factors such as alcohol usage, chewing tobacco, smoking, or no prior head and neck radiation. In addition to the prevalent variables, there are a lot of different factors that could be the cause of this tumour. To better understand and raise awareness of this uncommon cancer, we have described that case here to shed light on potential risk factors and warning signs.

Keywords: Sarcomatoid; Oral cavity; Malignancy; Aggressive tumour.**Introduction**

A rare type of squamous cell cancer, known as sarcomatoid (spindle cell) carcinoma, is characterized by a biphasic tumor composition of carcinomatous and sarcomatous elements [1]. It is additionally referred to as collision tumour because epithelial and spindle cells originate from separate stem cells, pseudo-sarcomatous carcinoma, and pseudosarcoma [2]. Although the mean age at presentation is 58 years, it is crucial to keep in mind that a wide range of 14-87 years indicates significant variability

in age at presentation [1]. It affects men more frequently than women. Even though the larynx may be a prevalent location for tumours, cancers of the oral cavity and oropharynx can still occur and should not be disregarded when making a diagnosis or developing a treatment strategy [2]. Seldom does it originate from the tongue. Smoking, consuming alcoholic beverages, and preceding head-neck radiation exposure are risk factors. A poorly differentiated form of squamous cell cancer is called sarcomatoid carcinoma and acts aggressively. Despite numerous studies utilizing immunohistochemistry, electron microscopy,

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and genetics, the exact histogenesis of sarcomatoid carcinoma remains a subject of debate. The motivation behind creating this case report stemmed from the patient in the index case, who had a rare history despite the presence of a highly aggressive tumor in an unusual location. It is essential to investigate a broad range of additional factors such as radiation therapy, pre-existing SCC, smoking, alcohol consumption, and tobacco use as common contributors to the development of this tumor. As a result, the case was explained to us in order to determine the many tumours causes and to raise awareness of this uncommon cancer.

Case history

A 34-year-old male presented in OPD in May 2022 with proliferative growth over the tongue for 4 months. He had history of tobacco chewing and smoking bidi for past 5 years. Clinical examination showed an exophytic growth on the right anterolateral border of the tongue with haemorrhagic areas visible with grade 3 trismus (Figure 1). Contrast enhanced computed tomography of face and neck depicted large heterogenous enhancing soft tissue lesion measuring 6.3 x 3.5 cm involving anterior two-thirds of tongue; right half of posterior third of tongue with right hyoglossus involvement with right level II, III lymph nodes involvement. The Punch biopsy revealed spindle cells as shown in Figure 2 suggestive of sarcomatoid carcinoma. Immunohistochemistry showed p53 (Figure 3) and vimentin positivity and was negative for cytokeratin (Figure 4). The patient did not show any distant metastasis in the metastatic workup. The clinical staging of T3N0M0 was made. The patient was deemed unresectable disease due to posterior extension of disease and underwent two cycles of Adriamycin and cisplatin-based chemotherapy followed by concurrent chemoradiation on a Telecobalt-60 machine with a dose of 70 Gy/35#/2Gy per fraction. The patient completed treatment in November 2022. The patient tolerated treatment well with Grade II skin reaction, Grade II mucositis, and Grade II dysphagia.

The patient was kept on follow-up monthly. The last follow-up by the patient was in February 2023.

Discussion

Sarcomatoid carcinoma is characterised by spindle-shaped cells and has a more aggressive behaviour compared to typical squamous cell carcinoma. This type of cancer is often biphasic, diagnosed at a later stage, and has a poorer prognosis [1,2]. Establishing the diagnosis in general and in this particular case was exceedingly challenging, as the squamous cell carcinoma component was not readily apparent. Studies show that sarcomatoid carcinoma mainly affects men in their sixth or seventh decade of life. There is a strong correlation between alcohol use and smoking [3]. Contrary to expectations, the index patient was younger than the mean age. In their study, Oktay et al. reported a case of spindle cell carcinoma of the tongue in a patient with a history of smoking and alcohol use. The patient also had a recurring lesion, providing valuable insights into sarcomatoid carcinoma [4]. However contrary to findings in [4]. Which did not record any positive lymph nodes here patient had positive lymph nodes at right level II and Level III. The larynx, tonsil, skin, and throat are the most prevalent sites of spindle cell carcinoma in the head and neck region [5]. One of the uncommon places where spindle cell carcinoma can form



Figure 1: Shows a smooth, polypoidal mass with a slough covering attached over anterior two-third and right lateral border of tongue, along with areas of haemorrhage on its lateral surface.

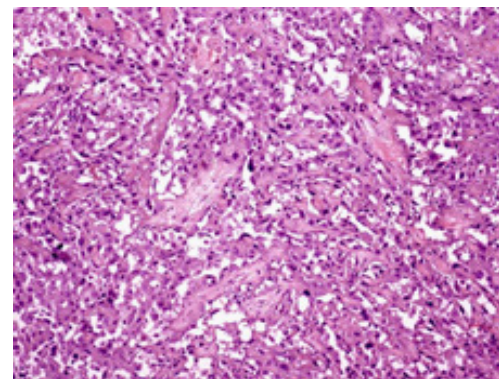


Figure 2: Haematoxylin and eosin (H&E, 40X) stain exhibits hypercellularity, giving the appearance of sarcomatous fascicles of spindle cells.

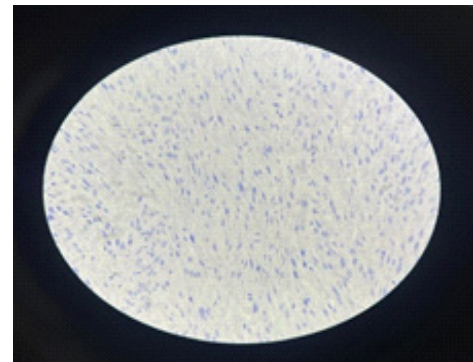


Figure 3: Immunohistochemistry showing p53 positivity.

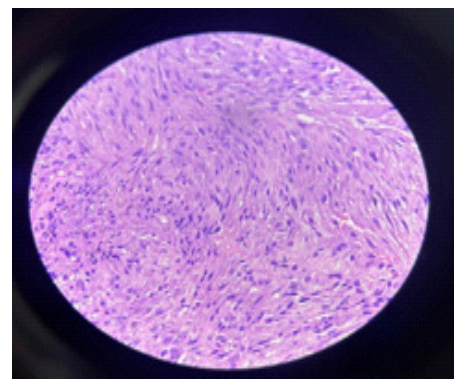


Figure 4: Immunohistochemistry showing negative for cytokeratin.

is the tongue, as it did in our case. Like in our case, these tumors typically grow quickly. According to histology, the epithelial component frequently merges with the mesenchymal component, which usually makes up the majority of the tumor [1]. Although distant metastasis is less common, regional lymph node metastasis has been found in up to twenty-five percent of patients [3]. In our situation, there were tumours in level II and III lymph nodes. According to Biradar et al. (2008), there was no involvement of lymph nodes, and the patient had a history of alcohol misuse, tobacco use, poor oral hygiene, and prior ionising radiation therapy to the tongue. The patient in this case report did not have any history of prior ionising radiation exposure [6]. Consequently, any genetic component or other etiological element might have also played a role. The treatment protocol for spindle cell carcinomas remains more or less similar to that for squamous cell variants. Surgery is the mainstay of treatment but radiation plays a pivotal role once the disease is deemed unresectable, inoperability, or in post-operative settings if margins are positive and or multiple lymph node involvement [1,7,8]. Surgery is seen as the best course of action in the oral region. It is unclear what part chemotherapy plays in the management of sarcomatoid cancer. A recurrence rate of up to 75% has been available in literature which demands closer follow-up of patients. Small size, superficial disease, lower stage of disease, and lack of prior radiation therapy are the parameters that offer a better prognosis [8].

Conclusion

Further research is needed to better understand the pathophysiology of sarcomatoid cancer of the tongue and its clinical behaviour to improve diagnosis and treatment outcomes. The lack of studies on oral spindle cell cancer highlights the importance of raising awareness about this rare form of cancer among healthcare professionals and researchers. Exploring potential risk factors or genetic mutations associated with sarcomatoid cancer of the tongue could provide valuable insights into its aetiology and possible prevention strategies. Long-term follow-up studies on patients diagnosed with sarcomatoid cancer

of the tongue could help clarify the prognosis and identify any potential recurrence patterns that may influence treatment decisions. Collaborative efforts among oncologists, pathologists, and researchers are crucial for advancing our knowledge of oral spindle cell cancer and developing more effective therapeutic approaches for patients affected by this rare disease. Furthermore, if malignant spindle cells are observed, a differential diagnosis of this rare entity should also be considered.

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