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Spindle Cell Squamous Cell Carcinoma of the Tongue: An Uncommon Variant in an Unusual Location

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ABSTRACT

Spindle cell carcinoma (SpCC) is an uncommon variant of squamous cell carcinoma (SCC) found in the head and neck region. This neoplasm, which is classified as biphasic, comprises both carcinomatous and sarcomatous components, making it rare, accounting for only 2% to 3% of all SCCs. The World Health Organization (WHO) formally recognized SpCC as a distinct malignant epithelial tumor 7 years ago. The origin of the spindle cells within the tumor remains a topic of debate, with many considering them to be monoclonal epithelial neoplasms closely linked to squamous carcinoma cells. Due to its rarity, the histopathological diagnosis of SpCC is challenging. Immunohistochemistry helps confirm its epithelial nature, with both tumor components displaying reactivity for cytokeratin and vimentin. In this report, we highlight a case involving a middle-aged man diagnosed with this rare variant in an even more unusual anatomical location.

Keywords: Tongue, Biphasic tumor, Spindle cell carcinoma, Head & Neck, Squamous cell carcinoma

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Introduction

Spindle cell carcinoma (SpCC) of the tongue represents an infrequent and distinctive form of squamous cell carcinoma (SCC) in the head and neck region. It is a biphasic tumor, meaning it comprises both carcinomatous and sarcomatous elements. This rare tumor variant, 1st described by Virchow in 1865 [1], accounts for only 2-3% of SCC cases in the head and neck. SpCC is known for its aggressive progression, with a tendency for both local recurrence and distant metastasis. While it is more frequently observed in the upper digestive tract, particularly in the larynx, its occurrence in the tongue is exceedingly rare. The tumor can also affect other regions such as the nasal cavity, hypopharynx, trachea, and even the breast. SpCC tends to predominantly affect males, especially those in their 60s and 70s. Known risk factors include alcohol consumption, smoking, and radiation exposure. The clinical presentation often includes a large, pedunculated, and polypoid mass, sometimes complicated by surface ulceration [2-7].

The rarity of SpCC makes its diagnosis particularly challenging. The exact origin of the spindle cells remains debated, though it is commonly believed that monoclonal epithelial neoplasia gives rise to both carcinomatous and sarcomatous components. This process likely involves metaplastic changes in squamous carcinoma cells, with a single stem cell being responsible for both components [8]. Immunohistochemistry (IHC) plays a crucial role in confirming the tumor's epithelial nature. Both carcinomatous and sarcomatous components test positive for cytokeratin, vimentin, and epithelial membrane antigen (EMA), but do not show any reactivity for markers like S-100 or smooth muscle actin alpha [9, 10]. Unfortunately, due to its invasive nature, the prognosis for patients with SpCC remains poor [7].

In this report, we highlight a case involving a middle-aged man diagnosed with this rare variant at an even more unusual anatomical location.

Case report

A 35-year-old man, with a background of tobacco chewing and excessive alcohol intake, visited the hospital complaining of speech difficulties and considerable weight loss over the past six months. He had previously been diagnosed with tongue carcinoma five years earlier and had undergone wide local excision. On clinical evaluation, a 3 cm by 2 cm ulcerative growth was noticed on the left side of the tongue, accompanied by enlarged lymph nodes on both sides of the neck. Imaging via FDG PET CT revealed an ulcerative lesion with heterogeneous soft tissue density along the left lateral tongue.

On macroscopic examination, the lesion appeared as a solitary, firm, grey-white growth measuring 3.0 cm by 2.0 cm by 1.0 cm, with a margin 0.8 cm from the base. The sectioned surface displayed a firm, greyish-white texture with areas showing ulceration and hemorrhage, but no signs of necrosis **(Figure 1a)**. Histological analysis revealed extensive granulation tissue and a subepithelial region containing two neoplastic cellular types: one with spindle-shaped, pleomorphic cells arranged in fascicles, and the other with nests of squamous epithelial cells exhibiting marked pleomorphism, atypical nuclei, and scant cytoplasm. Prominent nucleoli and irregular chromatin were also noted, along with atypical mitotic figures. No lymphovascular or perineural invasion was observed **(Figures 1b and 1c)**. The tumor margins were clear of malignancy. Immunohistochemical staining confirmed the epithelial origin of the tumor, with positive staining for cytokeratin **(Figure 1d)**, vimentin, and p40 (focal nuclear positivity), while CD34 and smooth muscle actin alpha were negative. During the modified neck dissection, 10 lymph nodes were removed, all of which were free of tumor involvement. The final diagnosis was confirmed as a spindle cell variant of squamous cell carcinoma (SpCC), staged as rpT3N0Mx grade 3. The patient was recommended to undergo neo-adjuvant radiotherapy and is currently being followed up.

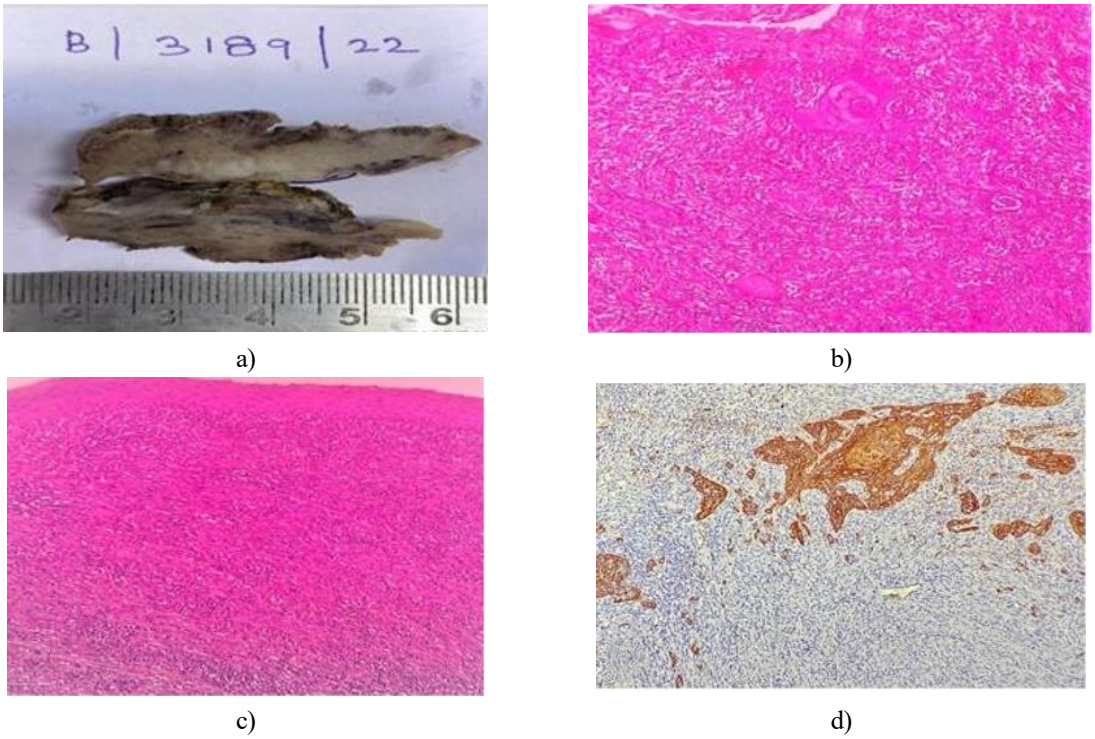

Figure 1. a) depicts the cut section of the ulceroproliferative growth of the tongue, which appears grey-white, b) a microscopic image showing the squamous epithelial cell component of the tumor, c) a microscopic image revealing the spindle cell component of the tumor, and d) immunohistochemical staining for CK 5/6 demonstrates positive results in the squamous epithelial cell portion, while the spindle cell area shows no reaction.

Results and Discussion

Spindle cell carcinoma (SpCC) is a rare tumor subtype, representing only 2-3% of all squamous cell carcinomas (SCC) found in the head and neck. This biphasic neoplasm contains both carcinomatous and sarcomatous elements. There is limited documentation of its occurrence in the tongue, particularly in younger patients. Various terms have been used to describe this tumor, including sarcomatoid carcinoma, carcinosarcoma, collision tumor, and pseudo-sarcoma. The histogenesis remains debated, although the monoclonal theory is widely accepted, which posits that both components of the tumor arise from a single stem cell through a process of metaplastic transformation or de-differentiation [8].

Typically, SpCC is most common in men between the ages of 60 and 80, with the larynx being the primary site of involvement [6, 7]. In the present case, however, the tumor arose in the tongue, a relatively rare location for SpCC, particularly in a young patient. Factors that may increase the risk of SpCC include tobacco use, poor oral hygiene, alcohol consumption, and prior radiation therapy to the affected area [2, 4, 7]. The patient in this case had a history of tobacco use and alcohol consumption, as well as a previous history of radiation therapy four years ago for carcinoma on the right lateral tongue.

Histologically, the tumor's mesenchymal component predominates and forms the bulk of the neoplasm. The characteristic spindling of the tumor cells is thought to result from alterations in cell adhesion molecules like e-cadherin, which leads to changes in the cytoskeletal network, including keratin [3, 4]. Immunohistochemical analysis in this case confirmed the diagnosis, as the spindle cells exhibited strong positivity for vimentin and focal positivity for cytokeratin, while the squamous cells were diffusely positive for cytokeratin. This suggests that the tumor's epithelial cells underwent metaplastic changes, leading to the loss of keratin expression [3].

According to the 2015 WHO classification for oral cavity neoplasms, SpCC is classified under the malignant epithelial tumors of SCC [3]. The progression of SpCC is often marked by recurrences and metastasis, with regional metastasis occurring more frequently than distant metastasis [2, 11]. Compared to conventional SCC, SpCC tends to have more aggressive behavior, resulting in a poorer overall survival rate [3, 6]. Factors influencing prognosis include tumor growth patterns, the extent of invasion, the presence of vascular involvement, and whether the disease has spread regionally or distantly, in addition to a history of radiation exposure [3, 6, 12].

Conclusion

The diagnosis of SpCC can be challenging due to its rare occurrence, and histopathological examination combined with immunohistochemical analysis is essential for confirming the diagnosis. Although the tongue is an uncommon site for this tumor, pathologists should always consider SpCC in the differential diagnosis when malignant spindle cells are observed. A comprehensive microscopic examination should be conducted to identify the epithelial component. Using ancillary diagnostic methods can clarify the diagnosis and assist in the development of a more effective treatment plan, ultimately improving survival outcomes for patients with oral squamous cell carcinoma.

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Conflict of Interest: None

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