CASE REPORT

Sarcomatoid Carcinoma of Tongue: A Case Report

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Abstract

Sarcomatoid carcinoma (SC) is an unusual and aggressive variant of squamous cell carcinoma, which frequently recurs and metastasizes; for this reason, the right diagnosis is very important. It is a biphasic tumor consisting of epithelial and mesenchymal components. The diagnosis often presents a clinicopathological challenge solved only by immunohistochemistry. Because of different behavior and treatment response, there is a need to diagnose this entity accurately for better management and therapeutic intervention. In the present article, we report a case of SC of the tongue in a 65-year-old male who presented with a polypoidal growth over the lateral border of his tongue with a short history of 1 month. Immunohistochemical expression of cytokeratin was strongly positive in the epithelial component and focally in the spindle cell component. The spindle cell component showed a strong positivity for vimentin.

Keywords: Carcinosarcoma, Sarcomatoid, Squamous cell carcinoma.

International Journal of Head and Neck Surgery (2020): 10.5005/jp-journals-10001-1388

Introduction

Sarcomatoid carcinoma (SC) represents a rare variant growth pattern of squamous cell carcinoma (SCC) in which the spindle epithelial cell resembles a sarcoma on histological examination. It is also known as pseudosarcoma, carcinosarcoma, sarcomatoid SCC, or collision tumor.[1] This variation in nomenclature reflects the divergent interpretation of the sarcomatoid component as reactive or neoplastic, mesenchymal or epithelial. However, the WHO has placed this disease entity under malignant epithelial tumors as a variant of SCC.[2] The clinical course is considered aggressive with a high incidence of metastases.[3]

Case Description

A 65-year-old male presented with a polypoidal growth over the lateral border of his tongue since 1 month (Fig. 1). Magnetic resonance imaging revealed a well-defined lesion with altered signal intensity seen along the lateral aspect of anterior two-third of the right side of tongue. The lesion crossed the midline to involve the adjacent left side of tongue as well as most anterior part near the tip. Biopsy of the lesion revealed features of SCC—moderately differentiated type. A wide local excision of the tongue along with supraomohyoid dissection was thus done and sent for histopathologic examination. The cut surface of the tumor was solid, gray-white, and homogenous. Microscopic examination showed a tumor comprising sheets and nests of malignant squamous cells along with areas of atypical spindle cells arranged in interlacing fascicles and bundles. The tumor was seen infiltrating the underlying muscle, fat, and perineurium (Fig. 2). On immunohistochemistry, the spindle cells were diffusely positive for vimentin and focally for CK and p63 (Fig. 3) and negative for SMA, S100, and desmin. Thus, a diagnosis of SC was put forward and patient was referred for chemoradiotherapy. Consent has been obtained by the patient for publication of this case.

Discussion

Sarcomatoid carcinoma of the head and neck is a biphasic neoplasm first described by Virchow in 1865.[4] Although the etiology and clinical profile of this tumor are similar to SCC, they differ in their biological behavior and response to treatment. Sarcomatoid carcinoma typically affects men in sixth to seventh decade and is associated with smoking and alcohol consumption. It has been described as a more aggressive tumor with little response to radiotherapy in contrast with SCC. Usually, spindle cell area greater than 30% is considered a criterion by some authors for labeling the tumor as SC.[4]

In the region of the head and neck, these tumors are seen usually in the larynx and rarely in the oral cavity reportedly accounting for less than 1%. Very limited literature is available on the localization of SC on the tongue. Many terms, including pseudosarcoma, SC, collision tumor, carcinosarcoma, pleomorphic carcinoma, and polypoid carcinoma, have been applied. This reflects the divergent interpretation of the histogenesis of the spindle cell component. But it has been proved now that both components have a monoclonal origin, and the mesenchymal component would represent an undifferentiation of the squamous component. Evidence supporting this is the direct continuity and smooth transition of the spindle cells with areas of squamous epithelium and a dual expression of epithelial and mesenchymal antigens on the spindle cells.[2,5]

Sarcomatoid carcinoma often creates a diagnostic dilemma owing to its varying morphology. The diagnosis may be

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How to cite this article: Agrawal U, Kaur M, Narula V. Sarcomatoid Carcinoma of Tongue: A Case Report. Int J Head Neck Surg 2020;11(4):75–77.

Source of support: Nil

Conflict of interest: None

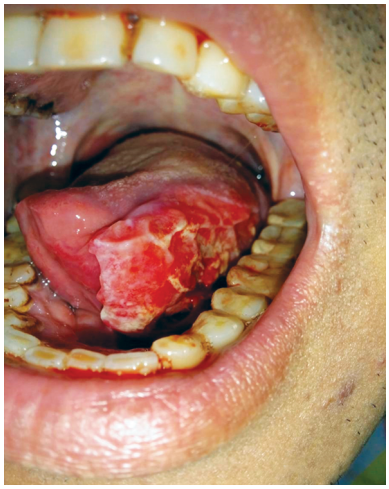
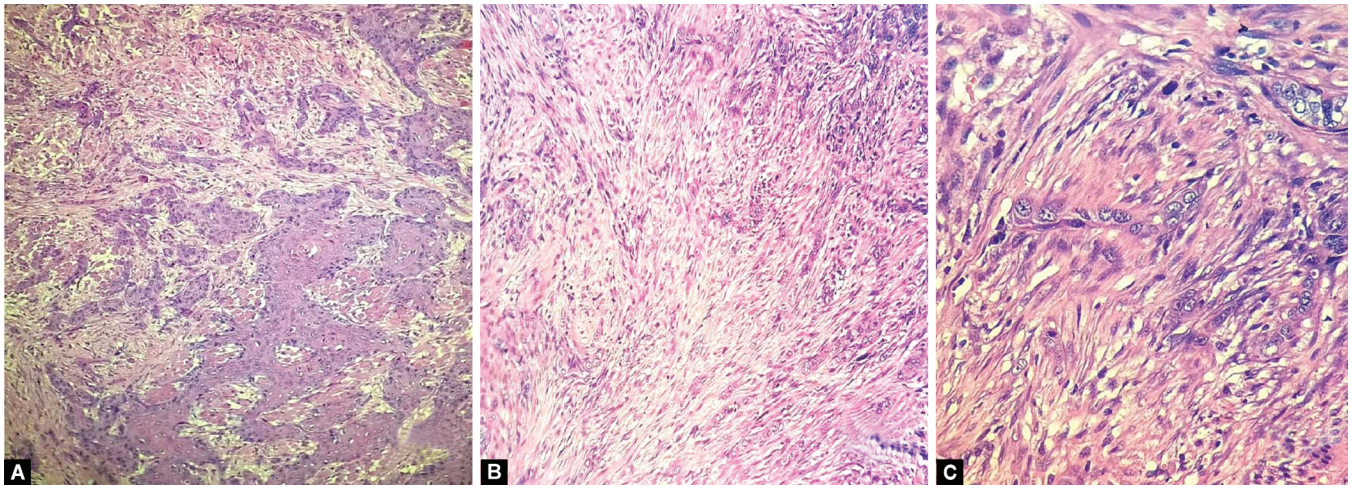


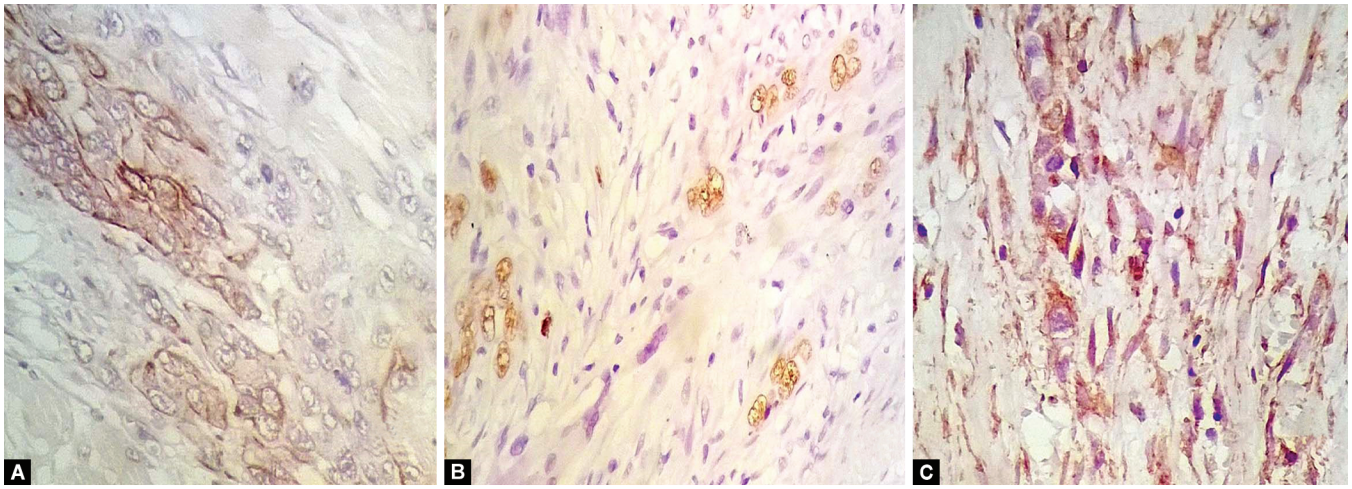
Fig. 1: Ulceroproliferative lesion seen in the tongue involving the anterior right and left side

exceptionally challenging when the SCC component is not obvious. A predominant spindle cell pattern raises the differential diagnoses of various benign and malignant tumors, such as fibromatosis, nodular fasciitis, reactive epithelial proliferations, fibrosarcoma, malignant fibrous histocytoma, leiomyosarcoma, rhabdomyosarcoma, malignant peripheral nerve sheath tumor, mesenchymal chondrosarma, and malignant melanoma.[6,7] An epithelial component should therefore be exigently searched for in such cases. Infiltrating SCC may be very focal, requiring multiple sections for demonstration.

In the present case, it was found that the spindle cell component was strongly positive for vimentin and focally positive for cytokeratin and p63 indicating the squamous origin. The vimentin positivity reflects that these spindle cells are squamous cells with true mesenchymal metaplasia. It has been proved that the epithelial cells acquire a mesenchymal pathway of differentiation metamorphosing to a spindle shape by undergoing a loss of cellular polarity, intercellular cohesion, and production of the mesenchymal matrix. These features also add to the invading property of the



Figs 2A to C: Hematoxylin and eosin-stained sections from tumor tongue show: (A) Areas of squamous cells arranged in sheets and nests infiltrating the lamina propria and muscle; (B and C) Spindle-shaped tumor cells in bundles with some multinucleated giant cells



Figs 3A to C: Immunohistochemistry for: (A) Pan-CK showed cytoplasmic and membranous expression in the tumor cells; (B) p63 was found expressed in the nuclei of the tumor cells; (C) Vimentin was present in the cytoplasm of tumor cells indicating mesenchymal nature of the cells

tumor and gain in vimentin expression.[6,7] In this process, keratin positivity may be scant, difficult to demonstrate, or even absent as was seen in the present case. Thus, the aggressiveness of these tumors can be attributed to all these factors.

Disease progression in SC is reported to be characterized by recurrences and metastases.[8] Surgical resection with adequate margins and neck dissection is accepted as the best treatment of choice in the oral cavity and is aimed at controlling local and distant recurrence. The role of radiotherapy is debatable as these tumors are known to show resistance. The role of chemotherapy in the treatment of SC is not explained. Factors associated with good prognosis are smaller size, superficial location, and absence of previous irradiation.[5,6] Our patient had undergone hemiglossectomy, and his condition was monitored with no recurrence to date.

The SC is a rare and aggressive variant of the SCC with a disputed histogenesis. Despite its challenging character, clinical history and presence of the squamous cell component or determination of epithelial nature of spindle cells would guide the pathologist to the diagnosis. Finally, it should be remembered that pure sarcomas of the head and neck are extremely rare and therefore spindle cell carcinoma should always be considered during evaluation of the polypoid lesions of this region containing spindle cells.[2] The role of immunohistochemistry is quintessential in establishing the epithelial nature of the spindle cells, which is fundamental for the diagnosis and an appropriate clinical management.

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