Case Report

Sarcomatoid Carcinoma: A Clinicopathological Profile of Two Cases with Diagnostic Emphasis

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Abstract

Sarcomatoid carcinoma (SC) is an unusual and aggressive variant of squamous cell carcinoma, which frequently recurs and metastasizes, and is associated with poor survival rate. For this reason, its accurate diagnosis is very important. It is considered to be a biphasic tumor made up of epithelial as well as spindle cell component, but of epithelial origin. The diagnosis often represents a clinicopathologic challenge, and immunohistochemistry plays a key role in the histopathological diagnosis. The reported cases in oral cavity are limited. Here, we present two cases of SC where the use of immunohistochemistry allowed us to achieve a conclusive diagnosis.

Keywords: *Immunohistochemistry, sarcomatoid carcinoma, squamous cell carcinoma*

Introduction

Sarcomatoid carcinoma (SC) has always been the focus of controversy regarding its histogenesis. Ambiguity surrounding the origin of SC is reflected in the inconsistent terminology used to designate this tumor in the past such as carcinosarcoma, pseudosarcoma, collision tumor, and pleomorphic carcinoma.[1,2] Over the years, many studies have provided substantial evidence that it is a biphasic tumor of monoclonal origin. It evolves from conventional squamous cell carcinoma by means of dedifferentiation associated with sarcomatoid transformation.[1-8] It most commonly involves larynx followed by other mucosal sites such as hypopharynx, nasal cavity, and oral cavity. Oral subsites frequently involved are buccal mucosa, gingiva, alveolus, and tongue.[1,2] SC poses as a significant diagnostic challenge due to morphological and immunohistochemical (IHC) overlap with other benign and malignant spindle cell tumors, especially when carcinomatous component is lacking.[7] It is essential to differentiate it from squamous cell carcinoma (SCC) or spindle cell neoplasms as it is associated with poorer prognosis, frequent metastasis, and higher recurrence rate. IHC has especially proved to be an important diagnostic tool in cases of dilemma.

Two case reports of SC along with their IHC profile are discussed here, with emphasis on diagnostic approach.

Case Reports

Case 1

A 65-year-old male reported with the complaint of a growth on the right lateral border of tongue which he noted 1 week back. No history of pain, burning sensation, pus discharge, or bleeding was present. The patient complained of chronic trauma from tongue biting due to sharp cusps. He had a habit of smoking 10–12 cigarettes/day for 42 years. The medical and family history of the patient was noncontributory. Submandibular lymphadenopathy was present on the right side which was mobile and nontender. On intraoral examination, a well-defined, irregular, pedunculated, reddish-pink growth of size 2.5 cm × 1.5 cm was present on the right lateral border of tongue. It was firm, smooth, compressible, and nontender on palpation. It was clinically diagnosed as traumatic fibroma, and surgical excision of the lesion was performed [Figure 1].

Histopathologically, atypical epithelial cells from overlying dysplastic epithelium were seen showing invasion in underlying stroma in the form of nests and cords at focal areas. Abundant epithelioid tumor cells were observed having vesicular nuclei and atypical mitosis. Pleomorphic

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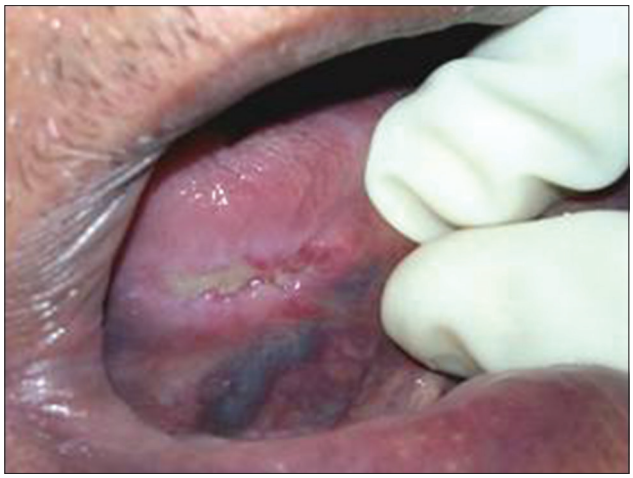

Figure 1: Intraoral photograph of Case 1 seen after incisional biopsy showed remaining part of the lesion on the right lateral border of tongue along with the biopsy wound in the center

cells varying from oval to spindled morphology having dense, hyperchromatic nuclei were intermingling with the epithelioid cells [Figure 2a and b]. Deeper muscle tissue also showed infiltration and degeneration. Inferior margin was positive for tumor cells.

On IHC tumor cells showed focal positivity for cytokeratin and diffused strong positivity for vimentin [Figure 3a and b]. Based on the pathological evidence, it was diagnosed as SC and further wider local resection of tongue was performed. Postsurgery, the patient underwent 33 cycles of radiotherapy 5 times a week. The patient has been on regular follow-up for 1½ year and shows no signs of recurrence.

Case 2

A 60-year-old female patient reported with a chief complaint of pain in the right lower back tooth region for 2 months. Pain was insidious in onset, dull and continuous in nature, nonradiating, aggravated on chewing, and relieved on medication. Later, a small growth also developed in the same region on the buccal mucosa which showed progressive increase in size. The patient visited a private practitioner where she was medicated for pain. She was wearing a fixed partial denture in the same region which she discontinued after the development of the lesion. Intraoral examination revealed a well-defined, irregular, and reddish-white growth approximately 1 cm × 1 cm × 1 cm in size on the right buccal mucosa. On palpation, it was firm with slight induration, sessile, fixed, and tender. A clinical diagnosis of fibroma was given and the growth was excised.

Histopathologic sections showed lesional tissue composed of pleomorphic spindled to ovoid cells with nuclear hyperchromasia and pleomorphism. Increased and bizarre mitosis was noted. Lesional cells were separated into small clusters by fine fibrous septae. Another bit showed hyperparakeratinized stratified squamous epithelium with dense stromal inflammatory cell infiltrate [Figure 2c and d]. Spindle cells showed strong immunopositivity for pancytokeratin and vimentin [Figure 3c and d]. Eventually, the lesion was diagnosed as SC on the basis of all the findings. The patient was lost to follow-up after few months.

Discussion

SC is a rare variant of SCC accounting for only 3% of all SCC cases in head and neck region.[1,2] Viswanathan *et al*. in their study of 103 cases of head and neck SCs reported an age range of 22–90 years (median 53 years), with male-to-female ratio as 3.7:1.[2] Thompson *et al*.[1] and Leventon and Evans[3] reported a higher mean age ranging from 63 to 66 years. A similar age involvement was noticed in our cases. The etiology has been linked to smoking, alcohol abuse, and previous history of radiation to affected area.[2] Thompson *et al*. observed that of 187 patients of laryngeal SC, 162 (87%) had smoking habit and 90 (65%) had a history of heavy alcohol consumption. Further, 17 (9.9%) patients had a history of radiation exposure among which 14 received it therapeutically due to prior SCC of larynx, pyriform sinus, or base of tongue.[1] The common oral subsites involved include buccal mucosa followed by alveolus, tongue, and palate.[2] Our cases showed a similar site predilection for buccal mucosa and tongue. Clinical presentation of SC may vary from superficial lesions (ulcerative or exophytic) to invasive (endophytic) lesions. SC presents as a polypoidal mass in 80%–90% of the cases.[1-4] Among the present two cases, one patient had a prior history of cigarette smoking and both cases clinically presented as a polypoidal mass [Table 1].

SC has biphasic histologic pattern proven to be originating from monoclonal dedifferentiation of conventional SCC.[1-5] The fraction of cells showing spindle cell morphology is highly variable in these tumors. According to Viswanathan *et al*., epithelial differentiation was evident on morphology in 48 (46.6%) cases, on IHC in 34 (33%) cases, and no epithelial differentiation was seen in 21 (20.4%) cases.[2] Both the cases described here showed admixture of epithelioid and spindle cell features. Cellular pleomorphism and atypical mitoses were prominent features in both the cases. The presence of definable squamous cell carcinoma (*in situ* or invasive) with atypical stromal component aids in legitimate diagnosis, but its absence may impede the same.[7] Such lesions can be easily confused with mesenchymal malignancies. Here, both cases showed invasion by the dysplastic overlying epithelium on histopathologic examination.

IHC was employed to rule out other lesions and confirm the diagnosis. According to most of the studies, SC shows positive immunoexpression for both cytokeratin and vimentin. Epithelial cells undergo progressive phenotypic

Table 1: Clinicopathological data of both cases

Age/sex	Habit	Site	Clinical presentation	Cell morphology	IHC profile	Follow-up	Recurrence	Mortality
65/male	Present	Tongue	Polypoidal	Epithelioid + spindle	PanCK+, VIM+	Yes	No	-
60/female	Absent	Buccal mucosa	Polypoidal	Epithelioid + spindle	PanCK+, VIM+	No	-	-

PanCK: Pancytokeratin; VIM: Vimentin; IHC: Immunohistochemistry

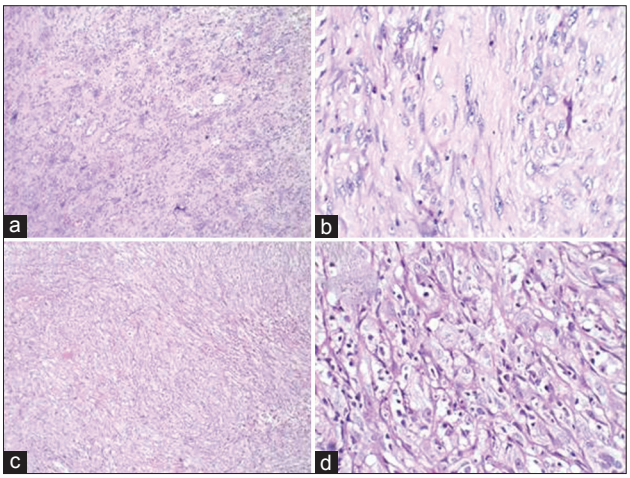

Figure 2: Histopathological sections of Case 1 showed (a) abundant epithelioid tumor cells intermingling with pleomorphic cells having oval to spindled morphology (H and E, ×10). (b) Epithelioid cells showed vesicular nuclei and atypical mitosis (H and E, ×40). Histopathological sections of Case 2 showed (c) mixed cellular features having both epithelial and spindle cell morphology (H and E, ×10). (d) Pleomorphism, nuclear hyperchromasia and increased and bizarre mitosis was observed (H and E, ×40)

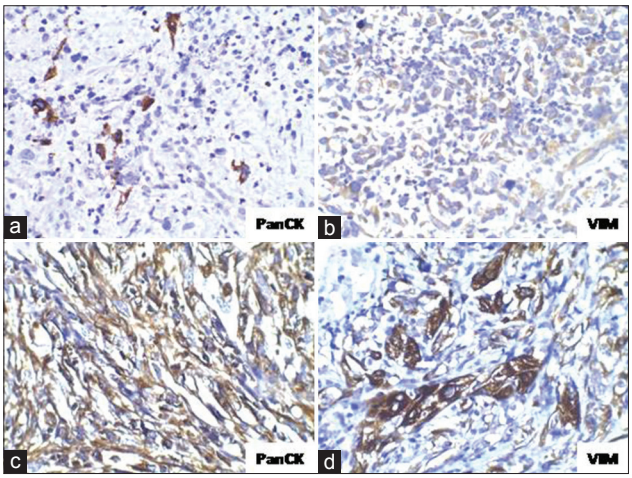

Figure 3: Immunohistochemical staining with antibody against pancytokeratin and vimentin in (a) Case 1 showed focal positivity for pancytokeratin and (b) diffuse strong positivity for vimentin (×40). (c) Case 2 showed diffuse strong positivity for pancytokeratin and (d) strong positivity for vimentin (×40)

changes as during embryogenesis, acquiring mesenchymal features such as spindle shape, loss of cellular polarity, and change in intermediate filament from keratin to vimentin. This interconversion of epithelial to mesenchymal cell aids in invasion into the stroma.[1,2,8] However, it is important to remember that SC should not be ruled out on the basis of positive reaction for vimentin and negative reaction for cytokeratin in tumor cells, as expression may vary with degree of metaplastic transformation. The diagnostic approach of Ro *et al*. which requires either an identifiable squamous component or evidence of epithelial differentiation in the spindle cell component (by electron microscopy or immunohistochemistry) should be essential criteria for a definitive diagnosis of SC.[8] Both cases showed diffuse strong immunopositivity for vimentin. However, focal immunopositivity was observed in Case 1 and diffuse immunopositivity in Case 2 for pancytokeratin. IHC was proved to be of paramount importance in diagnosing SC. Hence, the use of IHC is advocated in cases of spindle cell lesions, epithelioid malignancies, or both in combination. In cases of doubt, IHC helps substantiate the diagnosis.

Leventon and Evans[3] and Zarbo *et al*.[4] observed that prognosis was better when lesion is exophytic as compared to the deep infiltrating lesions. Recurrence rate ranges from 20% to 25% and distant metastasis is not uncommon.[1,3] Among the 20 cases followed up by Leventon *et al*., they reported a mortality rate as high as 45%; all deaths were seen in invasive SC lesions.[3] Viswanathan *et al*. managed to follow up 39 patients with average follow-up time of 8 months. They reported local recurrence in 8 (20.5%) patients, distant metastasis in 2 patients, and death in 3 (5.8%) patients.[2]

Conclusion

SC continues to remain a diagnostic dilemma for the pathologists. It varies largely in its pathogenesis, clinical behavior, and prognosis as compared to conventional SCC. That is why an unerring diagnosis is important which further leads to correct patient management.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

References

1. Thompson LD, Wieneke JA, Miettinen M, Heffner DK. Spindle cell (sarcomatoid) carcinomas of the larynx: A clinicopathologic study of 187 cases. Am J Surg Pathol 2002;26:153-70.
2. Viswanathan S, Rahman K, Pallavi S, Sachin J, Patil A, Chaturvedi P, *et al.* Sarcomatoid (spindle cell) carcinoma of the head and neck mucosal region: A clinicopathologic review of 103 cases from a tertiary referral cancer centre. Head Neck Pathol 2010;4:265-75.
3. Leventon GS, Evans HL. Sarcomatoid squamous cell carcinoma of the mucous membranes of the head and neck: A clinicopathologic study of 20 cases. Cancer 1981;48:994-1003.
4. Zarbo RJ, Crissman JD, Venkat H, Weiss MA. Spindle-cell carcinoma of the upper aerodigestive tract mucosa. An immunohistologic and ultrastructural study of 18 biphasic tumors and comparison with seven monophasic spindle-cell tumors. Am J Surg Pathol 1986;10:741-53.
5. Balercia G, Bhan AK, Dickersin GR. Sarcomatoid carcinoma: An ultrastructural study with light microscopic and immunohistochemical correlation of 10 cases from various anatomic sites. Ultrastruct Pathol 1995;19:249-63.
6. Choi HR, Sturgis EM, Rosenthal DI, Luna MA, Batsakis JG, El-Naggar AK, *et al.* Sarcomatoid carcinoma of the head and neck: Molecular evidence for evolution and progression from conventional squamous cell carcinomas. Am J Surg Pathol 2003;27:1216-20.
7. Batsakis JG, Rice DH, Howard DR. The pathology of head and neck tumors: Spindle cell lesions (sarcomatoid carcinomas, nodular fasciitis, and fibrosarcoma) of the aerodigestive tracts, part 14. Head Neck Surg 1982;4:499-513.
8. Ro JY, Chen JL, Lee JS, Sahin AA, Ordóñez NG, Ayala AG, *et al.* Sarcomatoid carcinoma of the lung. Immunohistochemical and ultrastructural studies of 14 cases. Cancer 1992;69:376-86.