Spindle Cell Carcinoma: A Report of a Rare Case in a 20-Year-Old Female Patient

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ABSTRACT

Spindle cell carcinoma (SpCC) also known as sarcomatoid carcinoma is a rare and peculiar biphasic malignant neoplasm that occurs mainly in the upper aerodigestive tract in the sixth to seventh decades of life with a male predominance. SpCC is an unusual aggressive variant of squamous cell carcinoma (SCC) that frequently recurs and metastasizes reinforcing the importance of its precise and early diagnosis. Herein we report a rare case of SpCC in an unusual location of maxillary alveolar ridge region in a 20-year-old female patient.

Keywords: Spindle cell carcinoma, Biphasic, Keratin, Vimentin and immunohistochemistry.


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INTRODUCTION

Spindle cell carcinoma (SpCC) is relatively a rare malignancy and a poorly differentiated variant of squamous cell carcinoma (SCC) with a more aggressive behavior. The most common site of origin in head and neck region is larynx and hypopharynx. It is rarely reported in the oral cavity. Despite several immunohistochemical, electron microscopic and genetic studies precise histogenesis of SpCC is still controversial. The incidence of this tumor in younger age group is yet considered to be rare and are sparsely reported in the literature. Hence we report a case of this rare tumor in a 20-year-old female patient with an aim to contribute a better understanding and awareness of this rare malignancy.

CASE REPORT

A 20-year-old female patient was referred to our oral medicine and radiology department for the evaluation of a nontender mass in relation to 27. The patient gave a history of extraction of 27, 2 weeks earlier following which she noticed a rapidly proliferating mass emanating from the post-extraction socket. Her medical history was unremarkable. There was no history of sinus-related pathologies or trauma to the maxilla. Extraoral examination revealed an obvious asymmetry (Fig. 1) on the left side of the face. Intraoral examination revealed an exophytic growth in relation to 27 measuring approximately 5 × 5 cm (Fig. 2). On palpation the surface was smooth, fibrous in consistency with well defined borders and palatal bone showed expansion in relation to 25, 24, 23, 22. The adjacent teeth 24, 25 and 26 were mobile. There were no palpable lymph nodes. OPG (Fig. 3) revealed an ill-defined radiolucency in relation to the socket of 27 involving the roots of 24, 25, 26 and the floor of maxillary sinus. Computerized tomography (Fig. 4) revealed a malignant mass eroding the walls of maxillary sinus, floor of the orbit and extending into the base of the skull. An incisional biopsy was performed. Microscopic examination revealed a biphasic tumor comprising a keratinized SCC and spindle cell type sarcomatous stroma diagnosing it as spindle cell carcinoma. Immunohistochemical markers such as pan CK, EMA and p63 were positive in cells composing the epithelial component and negative in spindle cells of the sarcomatous....
component. The spindle cells of the sarcomatous component were strongly positive with S100 whereas negative for vimentin, desmin and HMB-45. Treatment planning for a partial maxillectomy was abandoned due to the patient’s poor systemic health. The patient subsequently underwent palliative chemotherapy. However, this strategy was ineffective and the patient died.

DISCUSSION

SpCC is a malignant neoplasm characterized by both sarcomatoid proliferation of spindle shaped cells and SCC. Hence this entity can also be referred to plethora of names including pseudosarcoma, carcinosarcoma, pleomorphic carcinoma and SpCC.6 Recent studies states that the sarcomatoid component of SpSC could be a metaplastic alteration of SCC component.7 Some authors have emphasized that radiation or trauma can induce SpSC.6 But the patient we report here had no such history. Moreover, SpCC, like SCC affects primarily men in 6-7th decades and is strongly associated with smoking and alcohol consumption.1,4,5 This is in contrary to the present case where the patient is in the second decade and had no associated history of smoking or alcohol consumption.

In head and neck region, the most common localizations of SpCC are larynx, pharynx tonsil, oral cavity and skin.4 Oral tumors clinically present as a painful or painless polypoid exophytic masses expressing rapid growth or as a nonhealing ulcer.4 This is in accordance with the present case where the tumor expressed classical polypoid appearance with an extensive proliferation of the mass increasing in size from 5 × 5 (Fig. 2) to 10 × 10 cm (Fig. 5) within a duration of a month causing severe dysphagia.

Hsing-Hao Su1 et al8 in their study states that an overall 3-year survival rate of oral SpCC in the early stage group was 100%. But this is in contrary to the present case where the patient died within 2 months of diagnosis who had no nodal involvement and no distant metastasis. Here, the question arises, ‘Is the behavior and the survival rate of SpCC different from that of SCC?’ Leventon et al found that survival rate was related to the depth of invasion and in patients whose tumors invaded deeply the survival rate was low, whereas in those whose tumors were superficial, survival prospects were excellent.7

Alonso et al founded that SpCC demonstrated prominent local invasiveness, high angiogenic response, and a 90 to 100% incidence of lung metastases when inoculated subcutaneously into syngeneic mice.9 E cad is a calcium dependent intercellular adhesion molecule that is linked directly to α cat and indirectly to β cat through β cat in the cytoplasm. This cadherin and catenin complex is linked to actin filaments and plays an important role in the tight intercellular adhesion system. Recent studies have shown a possible rise in inactivation of E cad gene resulting in reduced expression of E cad and α cat in carcinomas. It is believed that a dysfunctional cadherin and catenin complex causes cells to shift in morphology from squamoid to a more spindled type lacking tight intercellular adhesion and
permits a more infiltrative and diffuse pattern of growth resulting in invasiveness and early distant metastasis.\textsuperscript{10}

To summarize SpCC of oral cavity is potentially more aggressive than SCC and seems to recur easily with early metastasize. Due to scanty studies in literature, its exact pathogenesis, clinical behavior, and long-term prognosis have not been well understood yet. Although it is difficult to predict the biologic behavior in every case, patients whose tumors are deeply invasive tend to have a poor prognosis, whereas those with early-stage tumors have an excellent prognosis. Hence it should be treated accordingly and the treatment should aim at controlling local and distant recurrence. Ampil reported combination of surgery and radiotherapy as a treatment of choice where radiotherapy might be helpful in improving local control.\textsuperscript{11} Considering the high angiogenic response of SpCC, patients with tumor stage $>$T2 may require chemotherapy to decrease the risk of distant metastasis or regional failure.\textsuperscript{10} To identify the recurrence early close postoperative follow-up is necessary.

**CONCLUSION**

SpCC can also occur in younger age group and a high index of suspicion is the prerequisite for an early diagnosis and a favorable outcome thereby providing a better life for the patients.

**REFERENCES**


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