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Sarcomatoid carcinoma: A rare biphasic variant of OSCC: A case report

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Abstract
Sarcomatoid carcinoma is a type of spindle cell neoplasm. These varieties of neoplasms comprise of a diverse collection of benign and malignant neoplasms. Sarcomatoid carcinoma, also known by various other names such as Spindle cell carcinoma, Lane's tumor, to name a few, is an exceedingly rare and aggressive variant of squamous cell carcinoma [3, 7, 8] The variation in its nomenclature reflects the divergent interpretation of the sarcomatoid component as reactive or neoplastic, mesenchymal or epithelial [8]. It accounts for <1% of total tumors occurring in the jaw. Role of imaging in diagnosis of such lesions is irreplaceable considering the location and nature of the lesion.
In the present case of a 44 year old female patient with inability to open the mouth since over 6 months; the diagnosis was especially challenging due to location of the lesion and clinical symptoms. We aim to highlight significance of thorough clinical examination and appropriate imaging modalities for an unusual presentation of a rare neoplasm which can leave clinicians baffled.

Keywords: Sarcomatoid carcinoma, CECT, trismus, imaging, immunohistochemistry

Introduction
Spindle cell carcinoma is an exceedingly rare variant of OSCC with biphasic properties i.e., having properties of both epithelial as well as sarcomatous component. It is a true sarcoma in origin. WHO has classified it under highly malignant variant type of OSCC. Most common site of occurrence is upper aerodigestive tract. Sarcomatoid carcinoma is often referred to as a biphasic tumor with aggressive tendency to malignancy. It shows a poor prognosis with frequent recurrence and systemic metastasis [1]. One characteristic feature of sarcomatoid carcinoma is it can cause obstructive symptoms as seen in our case.

Case report
A 44 years old Female patient reported to the department of Oral Medicine and Radiology with the chief complaint of progressive reduction in mouth opening and pain since 2-3 months. She was asymptomatic three months back when she experienced dull, aching pain on the left side of the face along with gradual reduction in the mouth opening. The symptoms increased in severity over time disturbing her daily routine activities and causing complete inability to open her mouth. There was no associated history of trauma. This alarmed her and she visited an ENT surgeon 1 month later and was advised endoscopic examination of nasopharynx and oropharynx to evaluate presence of any pathology. The reports were normal, and she reported to our hospital for further evaluation.
On clinical examination, there was severe trismus (mouth opening - <6mm) with mild pain. (Fig 1, Fig 2) Her face was bilaterally symmetrical and examination of TMJ showed normal intra-auricular condylar movements without evidence of crepitus. Muscles of mastication were non tender on palpation.
Examination of area of chief complaint intraorally was performed under local anesthesia. The lower jaw was gently forced open which revealed presence of an ulcerative growth with indurated borders extending superiorly upto the maxillary tuberosity region. (Fig 3, Fig 4) On palpation, it was extremely tender and bled on probing.

Bilateral level Ib lymph nodes were tender, soft to firm in consistency, mobile and not attached to underlying tissues. A provisional diagnosis of trismus secondary to a chronic non-healing ulcer was given.

A screening OPG was taken which showed moderate horizontal bone loss irt 26, 27 and 28 and moderate occlusal caries with 27 (Fig 5). CECT revealed presence of a single, diffuse, uniformly hypodense mass on the left side of the jaw. The coronal section was taken with the patient in puffed cheek position. Coronal section was visualized at the level of maxillary sinus showed presence of a single, diffuse, hypodense mass filling the entire sinus on the left side, about 7 x 5 cms in size and extending inferiorly to involve the buccinator muscle. A discontinuity was noted in the floor of the maxillary sinus. The left masseter muscle appeared to be hypertrophied. The second section taken anterior to the maxillary sinus showed invasion of the lesion into the left lateral pterygoid muscle. (Fig 6, fig 7) The sagittal section at the level of premolars showed a single, diffuse, ill-defined hypodensity extending from the maxillary sinus posteriorly into the oropharyngeal space causing obliteration of oropharynx. (Fig 8, fig 9). The involvement of surrounding musculature could have manifested in extreme trismus in the present case. Bilaterally enlarged lymph nodes level Ib up to 1.1 cms was noted. A radiographic impression of soft tissue malignancy in the left maxillary and mandibular oropharyngeal region was delivered.

Punch biopsy of the lesion revealed poorly differentiated Sarcomatoid squamous carcinoma of left upper buccal mucosa. (Fig 10, 11) IHC of the specimen revealed expression of P40 tumor marker and CD68 granules in the cytoplasm indicative of malignancy. (Fig 12, 13)

Surgical excision of the lesion along with excision of involved lymph nodes bilaterally was done. Additional treatment protocols of chemotherapy and radiotherapy were not advised. The patient was followed up for 3 months post-operatively and reported with adequate mouth opening and uneventful wound healing. The excisional biopsy confirmed the lesion to be poorly differentiated Sarcomatoid squamous carcinoma of left upper buccal mucosa.

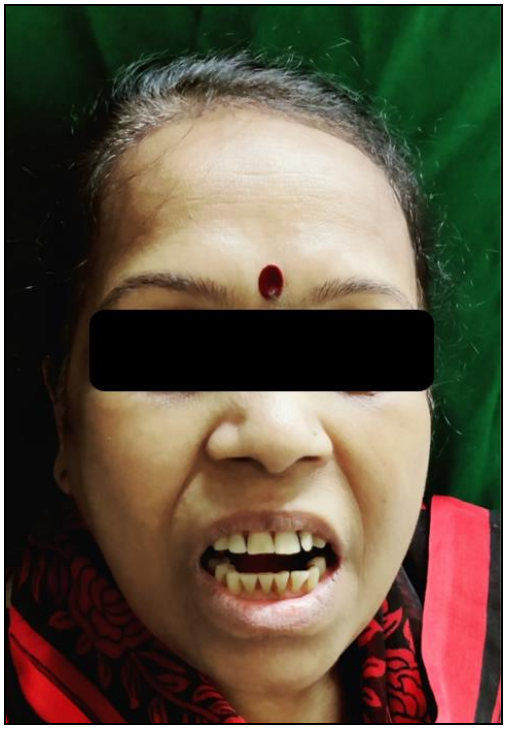

Fig 1: Extremely reduced mouth opening on clinical examination.

Fig 2: Mouth opening was <6mm as measured on Vernier caliper.

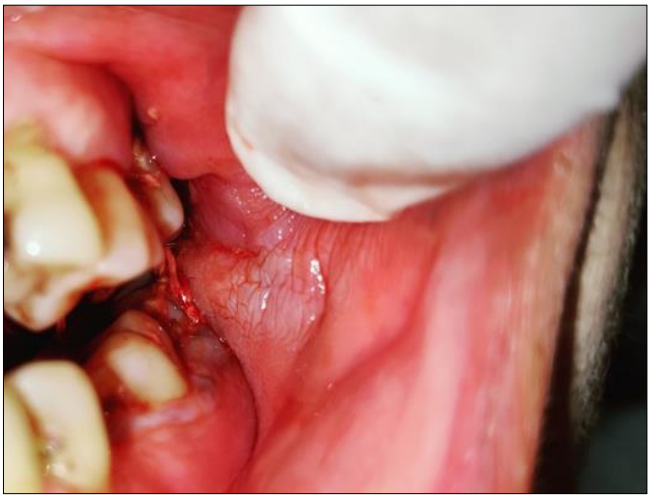

Fig 3: Forceful mouth opening shows presence of altered texture of mucosa on left side.

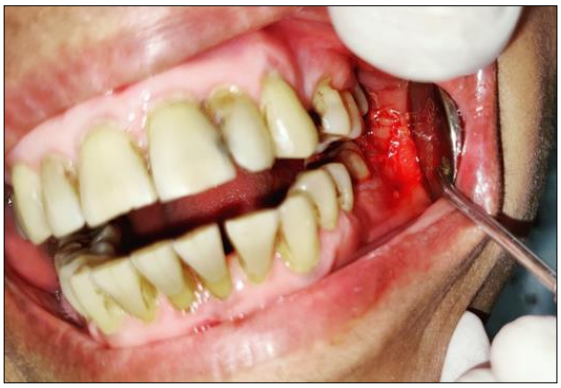

Fig 4: Forceful mouth opening shows presence of ulcerative growth in left buccal mucosa.

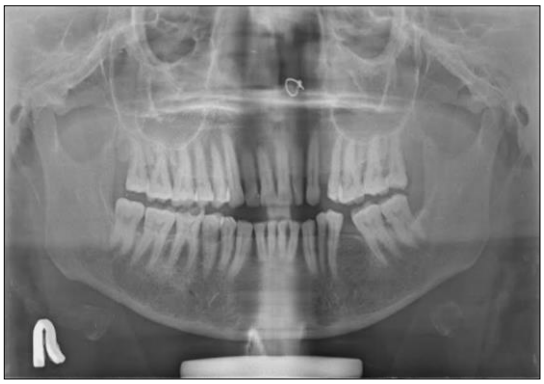

Fig 5: OPG shows presence of moderate horizontal bone loss irt 25, 26, 27.

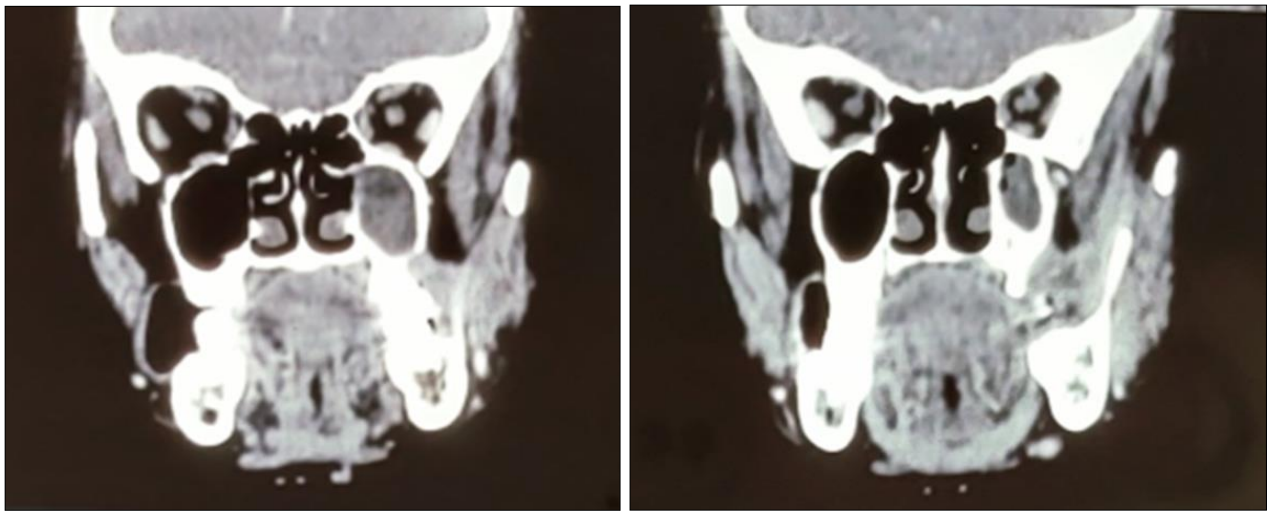

Fig 6, 7: Coronal CECT at the level of maxillary sinus shows presence of hyperdense mass in left maxillary sinus causing disruption in the maxillary bone with irregular borders as seen in malignancy. In Puffed cheek position of the patient, right side looks completely normal whereas the lesion seems to involve buccinators muscle anteriorly and lateral pterygoid muscle posteriorly. Severe hypertrophy of the masseter muscle on left side is evident.

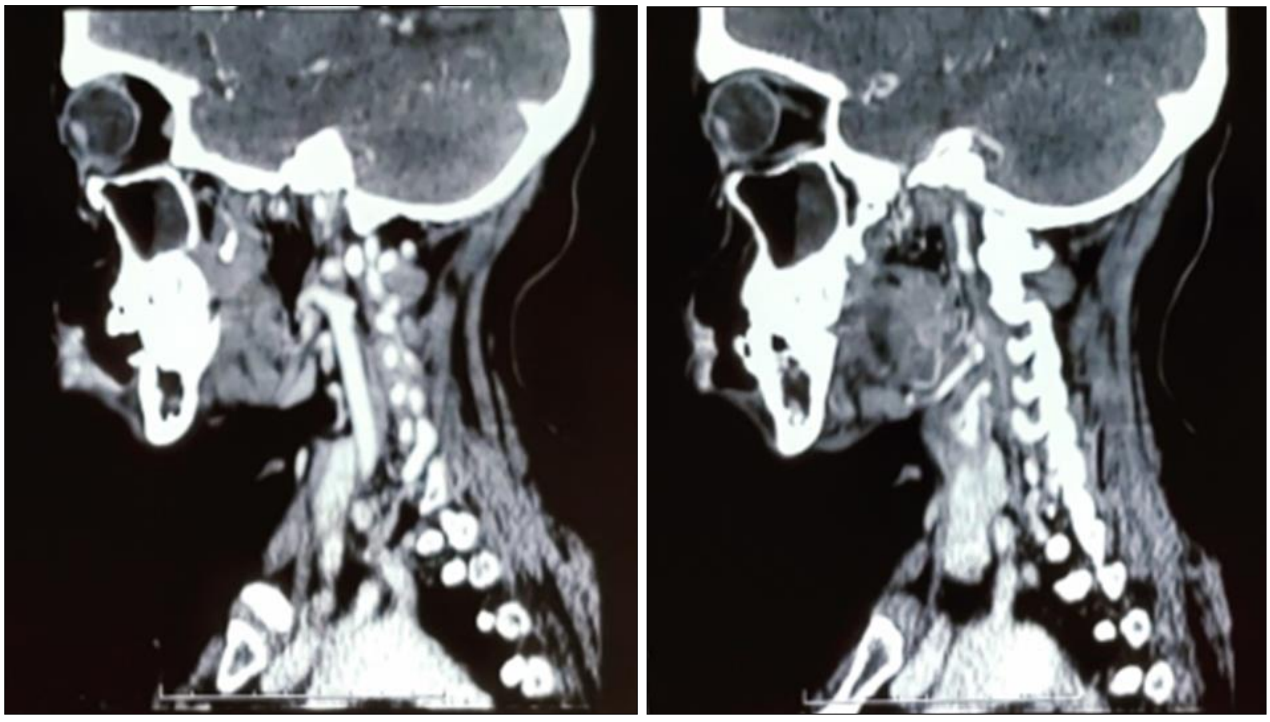

Fig 8, 9: Sagittal section of CT at the level of maxillary sinus shows presence of an ill-defined homogeneous hypodensity in pterygopalatine region extending into maxillary sinus.

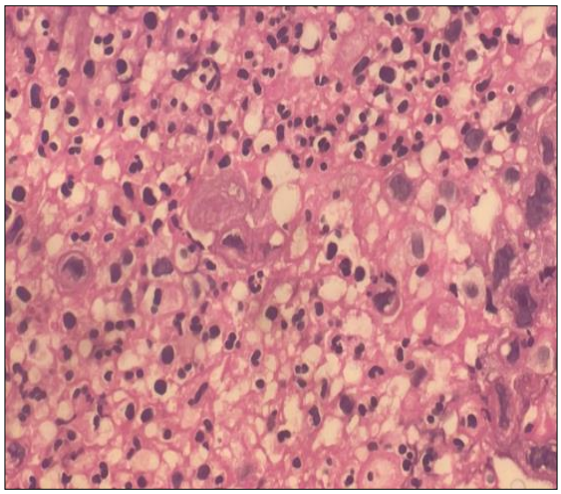

Fig 10: Spindle shaped cells interspersed with epithelial cells showing pleomorphism with vesicular nuclei and prominent nucleoli.

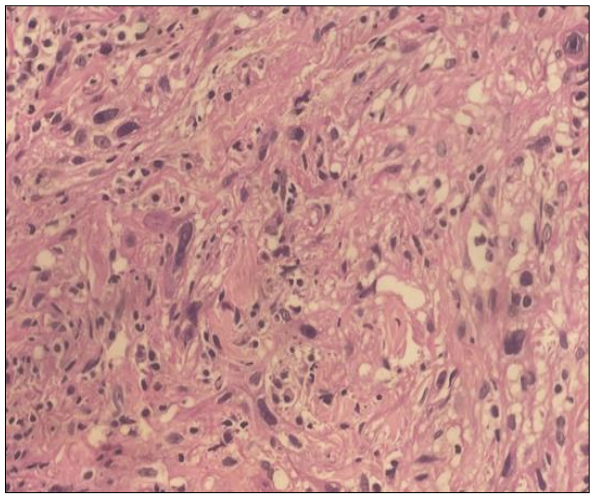

Fig 11: Moderate to severe cytological atypia with abnormal mitotic activity.

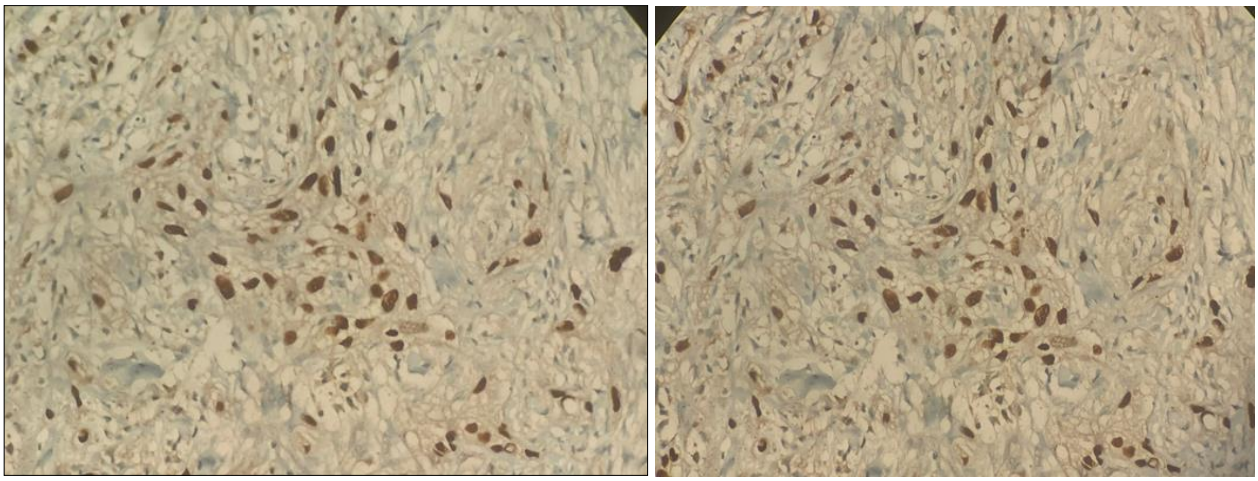

Fig 12, 13: IHC analysis shows presence of atypical cells having P40 marker and CD68 positive granules within cytoplasm. Cells show presence of enlarged nuclei.

Discussion

Sarcomatoid carcinoma is a rare variant of squamous cell carcinoma which is characterized by a dysplastic epithelial component and a stromal element with invasive fusiform or spindle shaped cells. The WHO recognizes SpCC as a variant of SCC, and has included it under highly malignant variant of SCC [2, 3]. The clinical and histopathological characteristics make it very difficult to distinguish SC from epithelial sarcoma. It is clinically a rare malignant epithelial neoplasm occurring primarily in upper aerodigestive tract. Other common sites include thyroid gland, breast, lungs etc. [4]. In the oral cavity, it accounts for less than 1% of all tumors. In the head and neck region, the oral cavity is the second most common site of occurrence. Specifically, the lower lip, tongue, and alveolar ridge are the commonly affected sub sites. The lesion was first reported by Virchow in 1864 and labeled as carcinosarcoma, suggesting that it may be a "collision tumor" between a carcinoma and sarcoma. Later, Krompecher in 1900 proposed the term "sarcomatoid carcinoma", proposing a dedifferentiation of the epithelium to spindle cell morphology [5] clinically, it may present as exophytic, nodular growth with extensive surface ulceration along with friable, shaggy exudate which can cause obstructive symptoms depending upon the site of occurrence. The clinical course is aggressive with a high rate of metastasis.

The present case posed a diagnostic challenge, in that, inability to open the mouth disallowed thorough clinical examination. Forced mouth opening using local anesthesia allowed visualization of an ulcerative lesion with tender indurated borders. Hence, it could be concluded that associated trismus in some cases may be caused by inward growth of the lesion. PET scan showed no secondary metastasis in this case.

Imaging

Radiographic features of this lesion are not distinctive, but adhere to the findings of neoplastic lesions, along with aggressive involvement of the surrounding soft tissues. Such ill-defined lesions may not be completely visualized using conventional radiographic imaging. Contrast imaging provides a complete assessment of the bony as well as the soft tissue components of the lesion. In the present case, CECT demonstrated a large, ill-defined widespread lesion, involving surrounding musculature posteriorly along with maxillary sinus and maxillary bone superiorly causing the trismus. The large size with ill-defined margins of the mass in a span of 3 months depicted the aggressive nature of the lesion.

Mahajan *et al.* [5] reported a case of SC with clinical presentation of an ulcerative growth involving the buccal mucosa with no reduction in the mouth opening. CECT did not reveal any bone involvement in their case. Ezulia *et al.* [3] have reported similar lesion in lower right alveolus which caused mild trismus similar to the present case. Yamazaki *et*

al. [6] presented a case report of pain and a polypoid mass on the lower left gingiva with associated trismus. Panoramic radiography showed bone resorption, exhibiting a moth-eaten appearance that involved the base of the mandible. Contrast-enhanced computed tomography revealed a soft-tissue mass lesion accompanied by extensive bone resorption of the mandible. Ezulia *et al*. [3] presented a case with a rapidly fungating mass over right lower molar region associated with pain, odynophagia and trismus. A pre-operative CT of the neck showed a lytic lesion in the alveolar margin of right angle of mandible. There is a soft tissue swelling medially and laterally and shows contrast enhancement. Shah *et al*. [7] based on the clinical and radiographic findings, gave a provisional diagnosis of malignant neoplasm with probability of SCC. Intraoral periapical radiograph of teeth associated with the lesion exhibiting mobility showed widening of lamina dura with mild displacement of 43 and horizontal bone loss prompting a diagnosis of SCC.

Histopathology

Histological presentation of spindle cell carcinoma is varied due to its biphasic nature of epithelial and sarcomatous components. Some of the more common patterns include, Presence of both epithelial and spindle shaped cells, cell in cell phenomenon, necrotic areas of connective tissue, abnormal or increased mitotic figures and inflammatory and polypoid stromal component. Mahajan *et al*. have reported that the epithelial component is usually found within the stalk or periphery of the lesion and forms a minor portion of the tumor mass, the sarcomatous component makes up the bulk of the tumor and consists of plump spindle cells along with scarcity of the carcinomatous component creating a diagnostic dilemma. The present case demonstrated Spindle cell proliferation in a myxoid background, associated with inflammatory cells in a polypoid stromal like component along with presence of pleomorphic epithelial cells under hematoxillin and eosin stains. Multiple spindle shaped cells interspersed with epithelial cells having cellular pleomorphism were seen, and, the nuclei were enlarged and showed moderate loss of basal polarity.

Immunohistochemistry

Owing to its biphasic nature, histopathology alone can cause a diagnostic dilemma. In such cases, using specific tumor marker, IHC can prove to be an important tool. IHC is able to show cellular atypia with enlarged nuclei, reactivity to tumor markers such as P63, P40, Ki-67 and positive expression of cytokeratin and vimentin which help to delineate the presence of mesenchymal cells in a carcinoma. In the present case, IHC using P40 marker revealed CD68 positive granules within cytoplasm indicative of malignancy and angiogenesis of tumor. (Fig 13).

Treatment

Management of SC is as tricky and controversial as its diagnosis. Some authors claim wide radical resection alone is sufficient while some prefers surgery with radiotherapy [5]. Surgical resection with adequate margins followed by radiotherapy is accepted as the best treatment of choice in the oral cavity; [3] Factors associated with good prognosis are smaller size, superficial location, and absence of previous irradiation.[8] Prognosis is dependent upon several factors such as depth and spread of the tumor but is usually not satisfactory and is considered potentially aggressive and with high tendency to recur and metastasize easily. The present case was treated surgically with excision of bilateral enlarged lymph nodes level Ib. Patient was followed up for 3 months and the course was uneventful. No radiotherapy was prescribed, however, to avoid recurrence of such highly anticipated recurrent tumors and to ensure comprehensive excision of the lesion, radiotherapy is best recommended adjuvant.

Conclusion

Sarcomatoid carcinoma of head and neck is a rare and unique subtype of squamous cell carcinoma. It is potentially aggressive than the classical Squamous Cell Carcinoma. It has very high tendency to recur and metastasize. It mimics other connective tissue sarcomas & spindle cell malignancies at light microscopic level [3]. 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 [8].

SC is unique in that, it presents with varied clinical, radiographic and histopathological presentations. Its biphasic nature warrants additional views of IHC in making the perfect diagnosis. The present case report highlights the diversity of the SC with respect to its clinical appearance, radiographic appearance and peculiar histopathologic presentation.

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Conflicts of interest

Authors declare no conflicts of interest.

References

1. Bo Young Kim. Kyoung Rai Cho. Jung Heob Sohn. Jung Yeon Kim. Sarcomatoid carcinoma after radiotherapy for early-stage oral squamous cell carcinoma Case report. Medicine. 2019;98:27
2. Palla Benjamin, Suhaym Omar, Majeed, Nasma, Garzon Steven, Callahan Nicholas. Spindle cell variant squamous cell carcinoma of the oral cavity: Case presentation and review of literature. Oral and Maxillofacial Surgery Cases. 2020;6(3):100174.
3. Ezulia T, Saim L, Sha PP, Kenali MS. Spindle Cell Carcinoma of the Oral Cavity: A Case Report. Clin Med Rev Case Rep. 2015;2:015.
4. Arshad HS. *et al*. A rare case of sarcomatoid carcinoma of the lung with spine metastasis. Am J Case Rep. 2017;18:760-765.
5. Mahajan A, Mohanty S, Ghosh S, Urs AB, Khurana N, Gupta S. Sarcomatoid Carcinoma of the Oral Cavity: A Diagnostic Dilemma. Case Rep Dent. 2017:7495695.
6. Yamazaki M, Maruyama S, Abé T, *et al*. Spindle cell squamous cell carcinoma exhibiting prominent neutrophil phagocytosis: a case report. J Med Case Reports. 2021;15:438.
7. Shah BJ, Tupkari JV, Joy T. Sarcomatoid squamous cell carcinoma of mandible: A report of two cases. J Oral Maxillofac Pathol. 2019;23:163.
8. Agrawal U, Kaur M, Narula V. Sarcomatoid Carcinoma of Tongue: A Case Report. Int J Head Neck Surg. 2020;11(4):75-77.