

# Small Cell Carcinoma of the Oral Cavity: Report of a Rare Case

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**ABSTRACT** Small cell carcinoma is primarily a lung malignancy occurring rarely in extra pulmonary sites such as the larynx, nasal cavity, paranasal sinuses, and oral cavity. The authors report a rare case of primary small cell carcinoma of the maxillary sinus presenting as a growth of the alveolus extending into the hard palate and the buccal vestibule.

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Small cell carcinoma is considered to be a distinct form of a malignant tumor that is most commonly of bronchogenic origin and well-recognized for aggressive clinical behavior. First described as an “oat cell carcinoma” by Barnard in 1926, this neoplasm can arise in extra pulmonary sites, including multiple areas in the head and neck.<sup>1</sup> In such cases, comprised of 2.5-5 percent of all SCC cases, however, a primary lesion in the oral cavity is very rare.<sup>2,3</sup>

This report describes a case of SCC of the maxillary antrum extending to involve the maxillary alveolus, buccal vestibule, and the hard palate. The clinical, radiographic, and histological features of this rare entity are presented.

## Case Report

A 72-year-old Indian Dravidian male patient with no premorbid condition reported to the authors’ department with a two-month history of tender swelling on the right side of face with associated symptoms of nasal congestion, epiphora, and rhinorrhoea. The patient was a chronic pan chewer and alcoholic for about 35 years but had no history of smoking. His past medical and dental history were noncontributory to his present oral condition.

Extraoral examination (**FIGURE 1**) revealed a large swelling in the right side of the face, which was approximately 8 x 8 cm in size, extending superior–inferiorly from the infraorbital margin to the inferior border of the mandible,



**FIGURE 1.** Extraoral view of the patient revealing a large swelling in the right side of face.



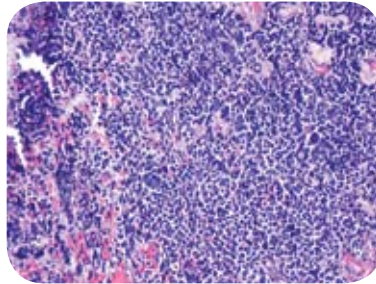
**FIGURE 2.** Intraoral view showing proliferative growth in the right alveolus in the posterior region.



**FIGURE 3.** A panoramic radiograph showing haziness and bony destruction of the floor and lateral wall of maxillary sinus.



**FIGURE 4.** Paranasal sinus view showing haziness and bony destruction of the floor, lateral wall of maxillary sinus, infraorbital margin, and lateral wall of the nose.



**FIGURE 5.** High-power photomicrograph showing large groups and sheets of diffusely infiltrating tumor with sheets of malignant cells with small round hyperchromatic nuclei, high nuclear cytoplasmic ratio, and nuclear moulding (H/E, X100).

and mediolaterally from the ala of the nose to 3 cms medial to the tragus of the ear. It had a woody consistency and was tender on palpation. However, submandibular lymph nodes were palpable on the same side, firm, mobile, and tender.

Intraorally (**FIGURE 2**), a proliferative growth was seen in the right alveolus. This was in the posterior region extending mediolaterally to the palate and the buccal sulcus causing the obliteration of latter. The teeth in the involved area had grade II mobility. A panoramic radiograph (**FIGURE 3**) revealed haziness and bony destruction of the maxillary sinus floor and lateral wall, the infraorbital margin, and lateral wall of the nose. A paranasal sinus, PNS, view (**FIGURE 4**) also showed similar radiographic changes.

The patient declined a computed tomography scan of the head and neck region due to financial constraints but did agree to have a surgical biopsy of

the lesion. Clinical laboratory reports were also within normal limits.

An incisional biopsy revealed large groups and sheets of diffusely infiltrating tumor with sheets of malignant cells containing small round hyperchromatic nuclei, high nuclear cytoplasmic ratio and nuclear moulding (**FIGURE 5**). A diagnosis of small cell carcinoma was made.

The chest radiograph, bone marrow aspiration, ultrasonography-abdomen, USG-ABD, were negative for any primary metastatic deposits. The patient was then referred to a specialized hospital, where four cycles of cisplatin-ethoposide chemotherapy was administered. Unfortunately, the patient expired within few weeks.

### Discussion

Small cell lung cancer is one of the most aggressive and lethal human cancers, and is the most common malignancy of the lung. The seminal case of SCC of

the lung was reported by Barnard in 1926.<sup>3</sup> One-third of patients present with limited stage disease confined to the chest. The remaining two-thirds of patients have disseminated disease involving the non-pulmonary organs, including in the head and neck: larynx, salivary glands, oral cavity, tongue, nasal cavity, and paranasal sinuses. These are collectively called “extra pulmonary small cell carcinoma.”

EPSCC is defined as biopsy proven small cell carcinoma in a nonpulmonary site. Additionally, chest radiograph, bone marrow biopsy, and USG-ABD should be negative for neoplastic cells.<sup>4</sup> It is postulated these tumors originate from totipotent stem cells that are native to all tissues and can differentiate into various cell types.<sup>5</sup>

Clinically, the presentation of EPSCC is difficult to characterize due to rarity of the lesion. The age at presentation has ranged from 26 to 77 years, with a median of 51.64. There is no clear sex predilection.<sup>4,5</sup> The lesions may appear as a smooth, polypoid mass or granular mucosal thickening. The most common presenting symptoms are epistaxis, nasal obstruction, and local pain. Proptosis and visual disturbances may occur when there is orbital involvement.<sup>6</sup>

SCC has a propensity to metastasize to other tissues. Hence, it is important to diagnose any extra pulmonary SCC as a primary site and not a metastases from another primary SCC. Two hypotheses have been proposed to explain the inability to detect the primary lesion.

These include involution of a primary tumor via spontaneous regression and the primary tumor possessing a phenotype and genotype favoring early metastasis.<sup>7</sup> In the authors' patient, since chest radiograph, bone marrow aspiration, USG-ABD were all negative, the oral lesion may be considered as a primary neoplasm.

Review of cases of HNSCC revealed that few cases have been described in the literature, with most occurring in the parotid area, one in the mandibular molar socket, and one in the nasal cavity.<sup>8-10</sup> Primary SCC of maxillary sinus is extremely uncommon.

The development of SCC is irrefutably correlated to cigarette smoking. In nonsmokers, SCC is extremely rare. In the authors' case, the patient was a nonsmoker. Even though it is reported that conventional microscopy is generally insufficient for diagnosis of SCC, an immunohistochemical analysis was not done in this case.<sup>11</sup>

With the paucity of reported cases of HNSCC, there is a definitive lack of standard protocol in the management of this tumor.<sup>7</sup> Surgery can be a treatment option in localized disease, however, this is usually not the case. Chemotherapy, RT, and combination therapy are the most preferred modalities.

Combination chemotherapy is considered more effective than the use of a single agent because of additive and synergistic effects. Cisplatin-Ethoposide (PE)-based regimens are most often used. Many chemotherapy regimens are not affective across the so-called "blood-brain barrier" thus the brain is left as a potential "sanctuary site" for metastatic disease. By giving prophylactic cranial irradiation, there is a decreased incidence of this complication. PE has been compared with other combination chemotherapy and has shown better results in terms of

survival and prognosis for the patient.<sup>12</sup> Carboplatin has also been shown to be highly active and less toxic than cisplatin.

Despite high response rates, relapse and progression occur in the majority of SCC patients, and the median survival is one year. The prognosis of such patients is poor and five-year survival is only 13 percent, with a median survival from diagnosis of only 14.5 months.

## Conclusion

SCC infrequently occurs as a primary tumor of the head and neck, and is considered to occur in nearly all structures associated with the aerodigestive tract. The prognosis for these patients is very poor. Defining the new combinations of agents for the survival of these patients is just a beginning, but the future appears to be promising. ■■■■

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