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CASE REPORT

Primary small cell carcinoma with neuroendocrine properties of the mandible: A case report and literature review

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Abstract Small-cell carcinomas at extrapulmonary primary sites are rare but they have been documented to arise at various locations. We report a case of small-cell carcinoma arising in the mandible, which has so far not been reported in the literature. A 37-year-old male patient underwent partial resection of the left mandible and adjuvant chemotherapy. Immunohistochemistry confirmed the diagnosis of small-cell carcinoma with neuroendocrine properties. The patient has been free of disease for 18 months after receiving treatment and was alive at the time of writing. We recommend surgical resection followed by chemotherapy for managing small-cell carcinomas in the mandibular region.

Introduction

Small-cell carcinomas (SmCCs) are anaplastic, highly malignant carcinomas that are mainly bronchogenic. Extrapulmonary sites account for only 4% of all SmCCs.[1] SmCCs rarely originate in the head and neck region. Survival differs between locations. The median survival time is 8 months for SmCCs of the renal pelvis,[2] whereas the 5-year survival rate for primary submandibular gland SmCCs is 40%.[3] These malignant tumors are composed of undifferentiated small cells and exhibit neuroendocrine differentiation by immunohistochemistry. The purpose of this report was to present a single, rare case of SmCC that originated from the mandible with neuroendocrine properties, based on immunohistochemistry, and describe a viable treatment option.

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Case presentation

A 37-year-old man was admitted with a complaint of pain in the left lower posterior teeth for 2 months and numbness in the left lower lip for 20 days. The patient denied a history of tobacco smoking, and his medical history was noncontributory. His chest X-ray showed no signs of space-occupying lesions in the lungs and no findings suggestive of pulmonary neoplasia. Clinical examination showed a tender, unmovable swelling on the mandible, numbness of the skin of the left lower lip, and no palpable lymph nodes in the bilateral submandibular region. Intraoral examination showed a 2 cm $\times$ 2 cm tender mass on the buccal side of the left molar region. The teeth involved were firm with no dislocation, and the overlying mucosa was normal.

A panoramic radiograph revealed that an ill-defined radiolucency of about 1 cm $\times$ 1.5 cm in diameter was involved in the root apex of the left lower first molar, and the lesion exhibited invasive characteristics of root resorption and periodontal ligament space widening (Fig. 1A). A computed tomography scan showed bone resorption of the buccal mandible body from D3 to D6 (Fig. 1B).

Laboratory data were within normal ranges. Tumor markers including α-fetoprotein, carcinoembryonic antigen, total prostate-specific antigen, carbohydrate antigen 19-9, and carbohydrate antigen 125 were tested.

The examination results suggested a primary malignancy of the mandible (cT1cN0cM0). During the operation, a frozen biopsy was performed, which demonstrated multiple small round cells exhibiting atypia consistent with a malignant neoplasm. Then, partial resection of the left mandible was performed. The continuity defect of the mandible from the midline to the third molar was reconstructed using a vascularized free fibular flap. The healing process was uneventful. The patient received adjuvant chemotherapy of six courses using cisplatin (40 mg/m^2) and etoposide (130 mg/m^2). Regular and aggressive follow-up every 1–3 months was carried out. No evidence of local recurrence or metastasis was observed 18 months after treatment (Fig. 1C and D). After surgery, the chewing of food and speech intelligibility were partially affected, whereas tongue mobility, swallowing, and breathing were normal.

On gross observation, the excised biopsy sample measured 3 cm $\times$ 2.5 cm, was hard, and appeared grayish-white. The pathological characteristics of the surgical sample revealed many scattered small foci of tumor cells with a cicatrized fibrous stroma in the musculoadipose tissue (Fig. 2). Therefore, an undifferentiated small-cell malignant tumor of the mandible was diagnosed. Assessment of the surgical margins showed that all margins of the tumor were negative. No tumor was found in any of the resected regional lymph nodes. The pathological stage was pT2N0M0 (margin negative) according to the pathological examination.

Subsequent immunohistochemistry demonstrated positive staining for cytokeratins AE1/AE3, CD56, synaptophysin, chromogranin A, and glial fibrillary acidic protein (Fig. 3A–C), and negative staining for myogenic differentiation 1, epithelial membrane antigen, S-100 protein, CD3, CD20, CD43, CD79a, CD99, leukocyte common antigen (CD45), B-cell lymphoma 2 (BcL-2), human melanin black (HMB)-45, melan-A, myeloperoxidase, and thyroid transcription factor (TTF)-1 immunostaining (Fig. 4).

Discussion

SmCCs are anaplastic, highly malignant, and usually bronchogenic carcinomas that account for 14% of bronchogenic carcinomas.[4] Extrapulmonary sites account for only 4% of all SmCCs.[1] SmCCs rarely originate in the head and neck

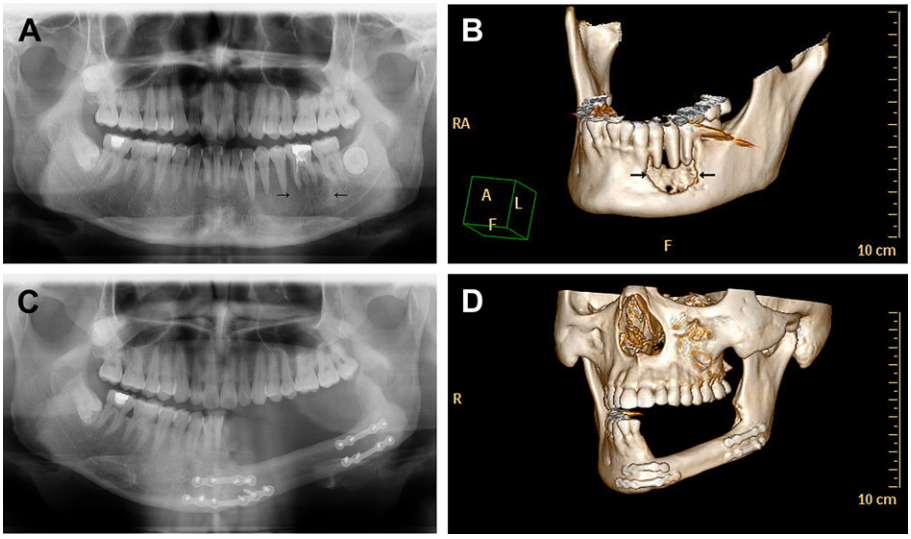

Figure 1 (A) Panoramic radiograph showing ill-defined radiolucency of the left mandible involving the apex of D6, root resorption, and periodontal ligament space widening. (B) Preoperative computed tomography scan showing a bone defect from D3 to D6. (C) Panoramic radiograph 18 months after surgery showing no absorption on either end of the fibular graft and good healing. (D) Computed tomography scan 18 months after the patient underwent surgery and adjuvant chemotherapy.

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