Carcinosarcoma of Tongue with Predominant Osteosarcomatous Component

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Abstract

Carcinosarcoma is exceedingly rare in the oral cavity and shows presence of both carcinomatous and sarcomatous components. We report a case of carcinosarcoma of tongue in a 51-year-old male with predominant osteosarcomatous and squamous cell carcinoma components. There was no evidence of regional or systemic metastatic disease. This case represents the first reported example of this unusual neoplasm with a predominant osteosarcomatous component, arising in the tongue.

Keywords Carcinosarcoma · Tongue · Osteosarcoma · Squamous cell carcinoma

Introduction

Carcinosarcoma, also called true malignant mixed tumor, is characterized by dual malignant histologic differentiation of both epithelial and mesenchymal components [1]. It is an extremely rare and aggressive entity with early metastasis and high mortality rate. The tumor has been extensively reported in the lungs, urinary tract, breast, and uterus. In the head and neck region, pharynx and larynx are most frequently involved, followed by esophagus, the oral cavity, and the nasal area [2]. Its occurrence in the oral cavity is unusual, and only few cases have been

reported in the literature [3]. We report a case of carcinosarcoma of the tongue with predominant osteosarcomatous component, which to the best of our knowledge, has not been reported earlier.

Case Report

A 51-year-old male presented with a painful lesion over the right lateral border of the tongue and trismus since 2 months. History of smoking and alcohol consumption was present since past 25 years. Patient was a known diabetic and hypertensive on treatment. There was no history of any past surgical intervention. Hematological parameters were within normal limits. The C-reactive protein was elevated (96 mg/L). Viral markers (hepatitis B surface antigen/hepatitis C virus/human immunodeficiency virus) were negative. On examination, mouth opening was 4 cm. An ulcero proliferative lesion measuring 6 × 5 cm was present over the right lateral border of the tongue, crossing the midline, and reaching up to the base of the tongue (Fig. 1a). The cervical lymph nodes were not palpable. The clinical stage was assigned as cT4aN0. Whole body contrast-enhanced computed tomography (CECT) and positron emission tomography–computed tomography (PET–CT) scanning showed an area of sclerosis and heterogeneously enhancing soft tissue mass at the lateral border of the tongue, measuring 5 × 3.6 × 4 cm. The pharynx and larynx were normal. Few small mildly hyper metabolic

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Fig. 1 **a** Tongue lesion crossing the midline and reaching up to the base. **b** Whole body CECT and PET–CT showing no other evidence of metabolically active disease. **c** CT showing a heterogeneously enhancing lesion at anterolateral border of tongue (arrow). **d** PET–CT showing increased 18-FDG tracer uptake in the tongue lesion (arrow)

bilateral level Ib, II, and right level III lymph nodes, likely reactive, were present. There was no evidence of any other metabolically active disease in the whole body scan (Fig. 1b–d). Following a biopsy, subtotal glossectomy with right neck dissection (level I to IV) with pectoralis major myocutaneous flap reconstruction and tracheostomy was performed. On gross examination, an ulcero-indurated, firm to hard lesion measuring $8 \times 5 \times 3.5$ cm was noted. Cut section was grayish white with focal hard to gritty areas and hemorrhage (Fig. 2a). Multiple sections examined microscopically showed an infiltrative biphasic neoplasm with extensive areas of unmineralized and partly mineralized osteoid with intervening anaplastic, mitotically active tumor cells exhibiting increased nucleo-cytoplasmic ratio, pleomorphic vesicular to hyperchromatic nuclei, occasional prominent nucleoli, and scanty cytoplasm. Foci of cartilaginous differentiation were noted. Mitotic activity was brisk [3, 4]/hpf with presence of atypical mitotic forms. There were few interspersed nests of neoplastic epithelial cells. Focal areas revealed presence of spindled neoplastic cells. The overlying stratified squamous epithelium showed ulceration and focal full-thickness dysplasia (Fig. 2b–f). There was no lymphovascular embolus, perineural invasion, or necrosis. The salivary glands and all the resected margins were free of tumor. The right neck dissection revealed 23 lymph nodes, none showing evidence of metastatic tumor deposits. Immunohistochemistry was positive for

cytokeratin-5 in the carcinomatous component but negative in the sarcomatous component with 90% Ki67 labelling index. P63 was evident in the invasive epithelial islands (Fig. 3a, b). A final diagnosis of carcinosarcoma tongue with osteosarcoma (85%) and squamous cell carcinoma (SCC) (15%) components was given.

Patient was given adjuvant treatment with concurrent cisplatin-based chemotherapy at a dose of 40 mg/m^2 weekly for 5 weeks, and radiation (60 gray). On 5th-month follow-up, patient had no evidence of recurrence or treatment-related complications.

Discussion

Carcinosarcoma is a heterologous malignancy with uniphasic or mixed metastasis [4]. Saphir and Vass analyzed 153 cases of carcinosarcoma of various sites and observed that it needs to be differentiated from pleomorphic (spindle cell) carcinoma and carcinoma with pseudosarcomatous stroma [2]. Pleomorphic carcinoma is a biphasic epithelial malignancy with uniphasic or mixed metastasis. Carcinoma with pseudosarcomatous stroma is a uniphasic epithelial malignancy with a benign yet atypical stromal reaction or epithelial malignancy with postirradiation stromal and epithelial anaplasia [4]. The histogenesis is explained based on two antithetical hypotheses. One proposes a multiclonal origin

Fig. 2 **a** Cut section of the tumor with overlying mucosal ulceration (arrow). **b** Unmineralized osteoid with intervening neoplastic cells (H&E $\times$ 400). **c** Focus of cartilaginous differentiation (H&E $\times$ 400). **d** Interspersed nests of neoplastic squamous epithelial cells (arrow) [Inset, atypical mitotic figures] (H&E $\times$ 400). **e** Spindled neoplastic cells (H&E $\times$ 400). **f** Epithelial ulceration and focal full-thickness dysplasia (H&E $\times$ 400)

arising from two or more stem cells; the other proposes a monoclonal origin arising from a single totipotent stem cell that differentiates in separate epithelial and mesenchymal directions [2].

Carcinosarcoma composed of both carcinomatous and specific-differentiated sarcomatous elements with typical features of malignancy is extremely rare in the oral cavity [3]. It has been reported in the tongue, lower gingiva, and floor of the mouth [3]. In the oral cavity, it usually occurs in males during 6th and 7th decades of life (similar to our case) [5]. The usual clinical presentation of carcinosarcoma in the head and neck region is of a painful or painless mass that may undergo a sudden increase [6]. Our patient presented with a painful lesion and trismus. Predisposing factors are smoking, alcohol consumption, and previous radiation exposure [5]. The latter two were present in our patient.

To our knowledge, there have been only few previously reported cases of carcinosarcoma of the tongue. Of these, the sarcomatous component was identified as chondrosarcoma, spindle cell carcinoma, or rhabdomyosarcoma, but there was no case of osteosarcoma. Our case had a predominant osteosarcomatous and a minor SCC component. Hence, extensive sampling of the tumor and histopathological examination is required for a proper diagnosis. The diagnosis can be rendered preoperatively if multiple biopsy cores are obtained from different areas of the tumor.

It is suggested by immunohistochemistry and fluorescence in situ hybridization analysis that cyclin D1 gene amplification plays a role in its molecular pathogenesis [7].

Given the rarity of carcinosarcoma of the oral cavity, there is no definite consensus for treatment. Our case was treated by surgery followed by adjuvant chemotherapy and radiotherapy, similar to squamous cell carcinoma.

Fig. 3 **a** High Ki-67 labelling index. **b** P63 nuclear positivity in the invasive squamous component

The prognosis of carcinosarcoma of the head and neck is controversial. Levinton and Evans showed that its prognosis is closely related to the depth of invasion [7]. On the other hand, Ellis and Corio reported that no histomorphologic characteristic, including the depth of invasion, was a reliable prognostic indicator in carcinosarcoma of the oral cavity [7].

Conclusion

Carcinosarcoma of the oral cavity is unusual. Predominant osteosarcomatous component has not been reported earlier. It is important to diagnose this entity due to its aggressive behavior.

Abbreviations SCC, Squamous cell carcinoma; *HBsAg*, Hepatitis B surface antigen; *HCV*, Hepatitis C virus; *HIV*, Human immunodeficiency virus; *PMMC*, Pectoralis major myocutaneous; *CT*, Computed tomography; *PET-CT*, Positron emission tomography–computed tomography; *H&E*, Hematoxylin and Eosin

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