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CLINICAL RECORD

Management of sarcomatoid salivary duct carcinoma of the submandibular gland duct with coexisting seropositive human immunodeficiency virus

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Abstract

Background: Sarcomatoid salivary duct carcinoma of the submandibular gland is extremely rare. This paper highlights the impact of surgery and adjuvant radiation therapy on the outcome of this disease.

Methods: A 59-year-old man with human immunodeficiency virus presented with a painless, rapidly growing left neck mass. Biopsy followed by surgical excision of the left submandibular gland revealed sarcomatoid salivary duct carcinoma of the submandibular gland duct with perineural invasion and close margins, for which he underwent adjuvant radiotherapy. Post-operative positron emission tomography and computed tomography revealed no residual or metastatic disease. Pathological analysis of tumour–node–metastasis staging revealed a T_2 N_0 M_0 (stage II) tumour.

Results: The patient tolerated his treatment without serious acute or long-term side effects. There was no evidence of disease on comprehensive examination or on positron emission tomography or computed tomography scans at the 4.6-year follow up.

Conclusion: Surgery followed by adjuvant radiotherapy provided practical locoregional control with acceptable toxicity. Further detailed case reports are warranted to optimise the management of this rare malignancy.

Key words: Submandibular Gland; Salivary Ducts; Carcinoma; Pathology; Human Immunodeficiency Virus; Radiotherapy

Introduction

Salivary duct malignancies are an extremely rare entity.[1] Submandibular gland sarcomatoid salivary duct carcinoma of Wharton's duct has been reported in a few cases.[2–6] However, it has never been reported in a patient with coexisting human immunodeficiency virus (HIV). Given the rarity of these malignancies, treatments are non-evidence based and are extrapolated from experiences of other head and neck malignancies of similar histology and/or location. We report the first documented case in the English literature of sarcomatoid salivary duct carcinoma of the submandibular gland in a patient with HIV. The paper highlights the impact of surgery and adjuvant radiotherapy on the outcome of that disease. This is particularly important because it is unlikely that a prospective trial will be performed in this patient population.

Case report

In early 2007, a 59-year-old Caucasian man presented with a painless, rapidly growing left-sided upper neck mass, which had first become apparent two months earlier during routine examination. The man, who was diagnosed with HIV in 1993 and was receiving highly active antiretroviral therapy, had an undetectable viral load and cluster of differentiation 4 counts of 1200 cells/ul. He denied previous tobacco use and alcohol or drug abuse. There was no family history of malignancy. The remainder of the physical examination was unremarkable. A computed tomography (CT) scan of the neck carried out in late December 2006 had shown an enlarged left submandibular gland with a dilated duct (Figure 1). Fine needle aspiration was performed and showed atypical epithelial cells without a clear diagnosis.

Management

The patient was discussed at our institutional multidisciplinary head and neck tumour conference, and the consensus was to proceed with surgery.

Surgery

In January 2007, the patient underwent gross total excision of the tumour and upper selective neck dissection of levels I and II.

Pathology

Pathology revealed a sarcomatoid variant of salivary duct carcinoma, with extensive necrosis. The tumour measured $2.5 \times 2.0 \times 1.5$ cm with perineural invasion. The tumour abutted the resection margin. No definite evidence of angiolymphatic invasion was noted. There was one lymph node in the specimen, which was histologically negative.

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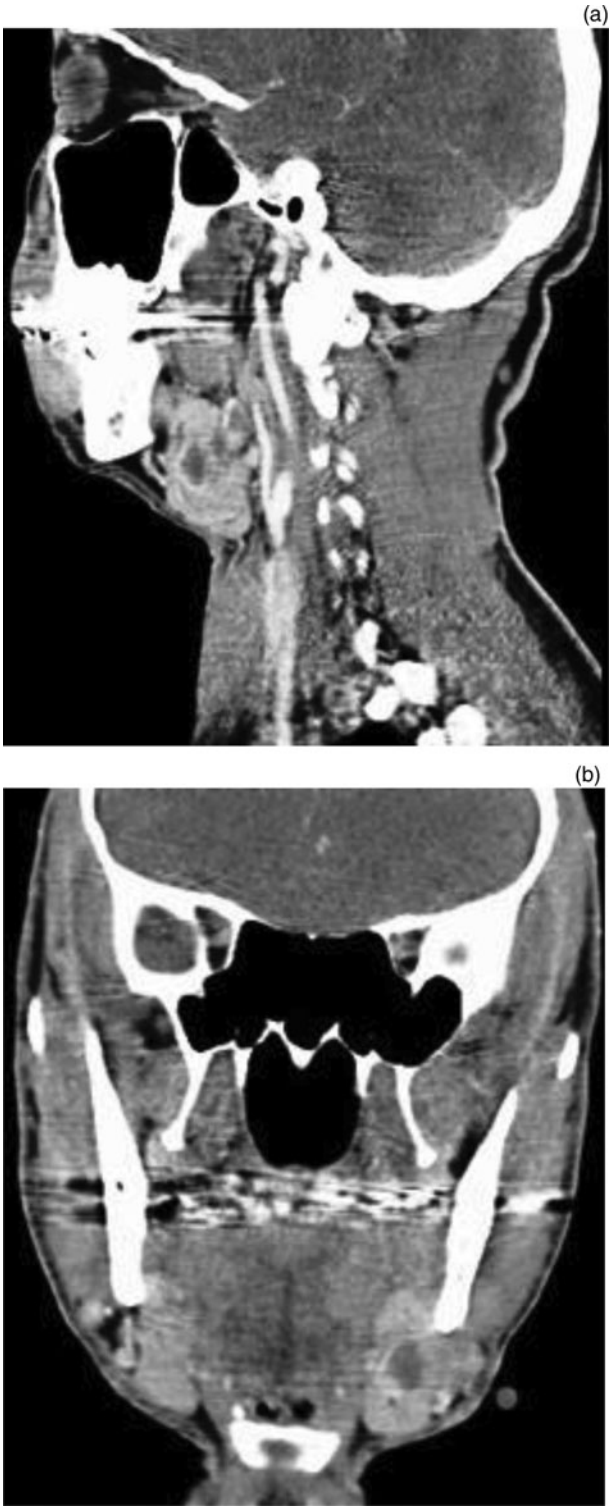

FIG. 1
Pre-operative sagittal (a) and coronal (b) computed tomography images showing an enlarged left submandibular gland with dilated duct.

Pathological analysis of tumour–node–metastasis staging revealed a T_2 N_0 M_x tumour.

Radiation therapy

Due to the rarity of the tumour, positron emission tomography (PET) and CT imaging were carried out; no obvious gross residual disease or evidence of metastatic disease was revealed. The patient was therefore diagnosed with a T_2 N_0 M_0 stage II submandibular ductal carcinoma. Given the perineural invasion and close margins on the surgical resection, the patient was further discussed at our institutional multidisciplinary head and neck tumour conference, and the consensus was to proceed with adjuvant external beam radiotherapy.

Three weeks post-surgery, the patient underwent CT simulation and intensity-modulated radiotherapy based treatment. Three planning target volumes were defined and dose painting plans were designed. The high dose region included the tumour bed and ipsilateral submandibular gland region, which received 66 Gy in 33 fractions. The intermediate dose region included the sublingual gland, the facial and lingual nerve routes to the base of skull, the lymphatics in transit, and the upper ipsilateral neck, which received 59.4 Gy in 33 fractions. The lower ipsilateral neck received a prophylactic dose of 50 Gy in 25 fractions with a matched lower neck field. Seven-beam intensity-modulated radiotherapy was chosen to preferentially spare the larynx, constrictors, right parotid and right submandibular gland, and minimise the doses to the oral cavity and tongue (Figures 2 and 3). The treatment was delivered via 6 MV photons.

The treatment was completed in April 2007. The patient was able to tolerate the treatment without a break, although he did experience the expected acute radiotherapy-related toxicity in the form of grades I–II dysgeusia, xerostomia, mucositis and dermatitis. Throughout the course of his treatment, the patient lost 1.8 kg without the need for aggressive nutritional support.

Follow up

After a follow up of 4.6 years from the date of radiotherapy completion, a comprehensive physical examination and PET and CT scanning confirmed the absence of any locoregional recurrence or distant metastasis (Figure 4). The patient reported no pain, skin change, altered taste, voice change, trismus, dysphagia, odynophagia or numbness, and his weight was stable.

Discussion

Salivary duct malignancies are an extremely rare entity. Only 25 cases of Stenson's duct squamous cell cancer have been reported.[1] In that series, which reported cases from 1927 to 1999, the most common histological diagnosis was squamous cell carcinoma.

The submandibular duct (i.e. Wharton's duct) is about 5 cm long, and its wall is much thinner than that of the parotid duct. It begins as numerous branches from the deep surface of the gland and opens by a narrow orifice on the summit of a small papilla at the side of the frenulum linguae. The duct drains saliva from the submandibular and sublingual glands to the sublingual caruncle at the base of the tongue. It was initially described by the English anatomist Thomas Wharton.[2]

Submandibular gland sarcomatoid salivary duct carcinoma of Wharton's duct has been reported in only a few cases.[3–6] Given the rarity of these malignancies, treatments are non-evidence based and are extrapolated from experiences of other head and neck malignancies of similar histology and/or location. The mainstay for treatment is primary resection when feasible, with or without nodal resection. Radiotherapy is used as adjuvant therapy, with or without chemotherapy, as dictated by the surgical pathology.

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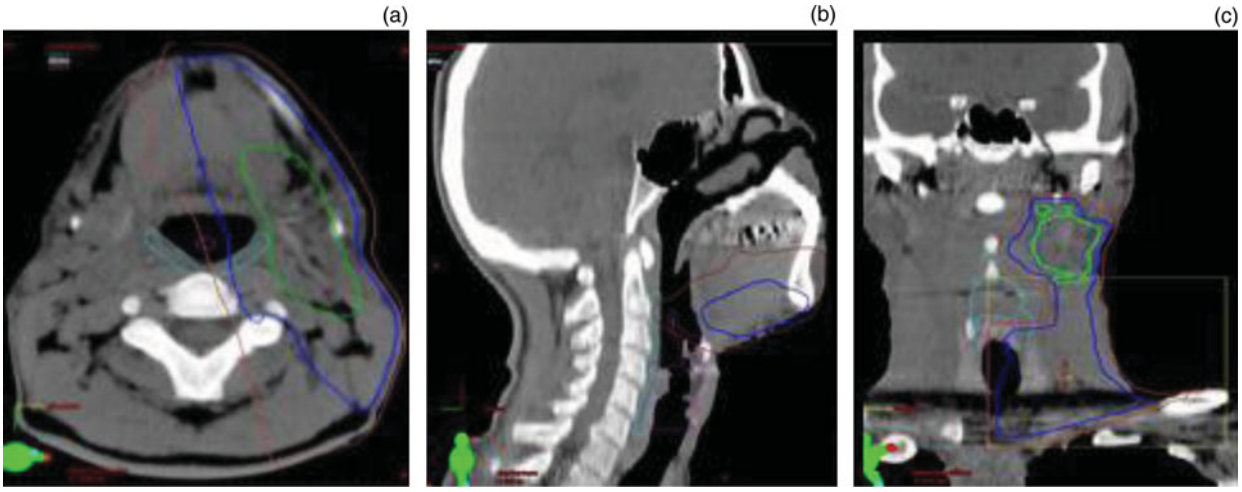

FIG. 2

Axial (a), sagittal (b) and coronal (c) computed tomography images of the constrictors and larynx showing the radiation doses (red = 66 Gy, green = 59.4 Gy, blue = 50 Gy) delivered to each target volume.

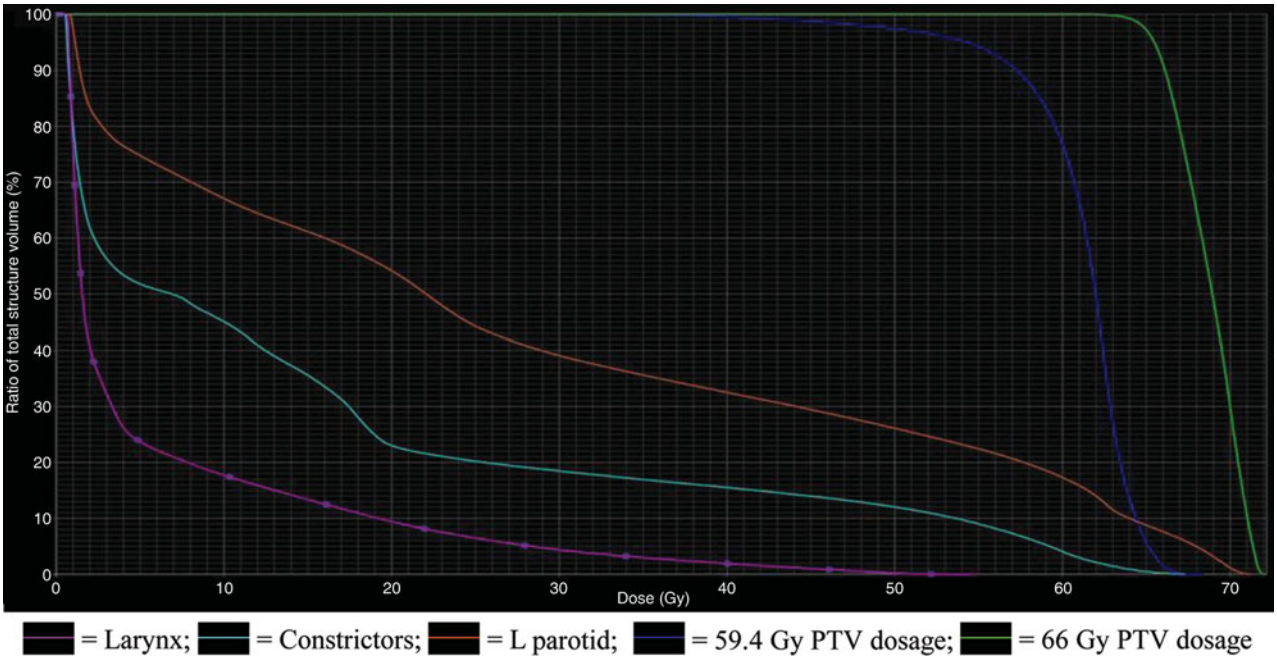

FIG. 3

Dose–volume histogram of planning target volumes for the left parotid, constrictors and larynx. L = left; PTV = planning target volume.

We report the first documented case in the English literature of sarcomatoid salivary duct carcinoma of the submandibular gland in a patient with HIV, and highlight the impact of surgery and adjuvant radiotherapy on the outcome of that disease. Due to the rarity of this disease, there is no universal consensus on management; however, surgery was performed in all previously reported cases.[3–6] There are no data to support the routine use of radiotherapy. We think that post-operative radiotherapy would be wise in cases with adverse prognostic features (such as locally advanced disease, close or positive margins, multiple lymph nodes, extra capsular extension, perineural invasion, and lymphovascular space invasion). In our patient, surgery and adjuvant radiotherapy appeared to offer long-standing locoregional control, with negligible long-term comorbidities.

- **Sarcomatoid salivary duct carcinoma of the submandibular gland is extremely rare**
- **This paper reports a case of submandibular gland sarcomatoid salivary duct carcinoma in a man with human immunodeficiency virus**
- **The patient underwent surgical excision of the left submandibular gland with left upper neck dissection**
- **He received adjuvant radiotherapy due to perineural invasion and close margins**
- **There was no evidence of disease at 4.6 years' follow up**
- **Surgery followed by adjuvant radiotherapy achieved locoregional control with tolerable toxicity**

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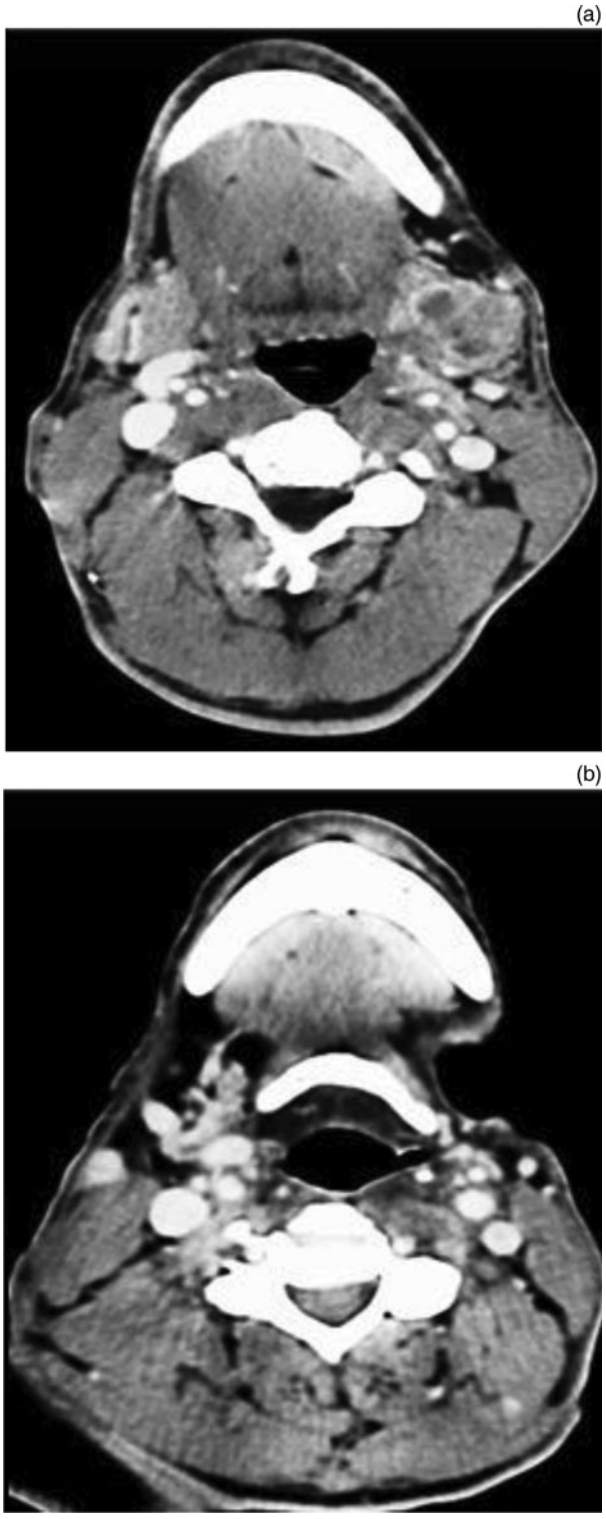

FIG. 4
Pre-operative (a) and 4.6 years post radiotherapy (b) axial computed tomography images, with no evidence of disease in (b).

This case report is particularly important because it is unlikely that a prospective trial will be performed on this patient population. There is little information in the literature to guide treatment. Often patients with this form of cancer are treated using regimens that have been employed at other sites with a similar histology or for a different histology but a similar location. We treated this patient's disease in a similar fashion to squamous cell carcinomas of the head and neck, with complete success thus far. The real risks of local or regional nodal relapse or metastatic potential are unknown. It is therefore unclear what the most appropriate areas (including nodal levels) to receive higher or lower doses are, and decisions have been guided by treatment of other head and neck cancers.

Conclusion

Submandibular gland sarcomatoid salivary duct carcinoma with coexisting HIV has not been reported previously. Surgery followed by adjuvant radiotherapy (due to close margins and perineural invasion) provided sustainable locoregional control. The patient suffered tolerable side effects, and there was no evidence of disease at a 4.6-year follow up. Further detailed case reports are warranted to optimise the management of this rare malignancy.

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