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ScienceDirect

British Journal of Oral and Maxillofacial Surgery 55 (2017) 77–80

BRITISH Journal of Oral and Maxillofacial Surgery

www.bjoms.com

Short communication

# Primary signet-ring cell adenocarcinoma of the head and neck: a case study and brief review

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Accepted 8 April 2016
Available online 13 May 2016

**Abstract**

We present a patient who had an aggressive primary signet-ring cell adenocarcinoma in the oral cavity that spread rapidly and led to his death. Most reports describe an indolent clinical course, but further reports are needed to better evaluate the particular clinical characteristics and course of this uncommon and biologically variable condition.


*Keywords:* Primary signet-ring cell adenocarcinoma; Signet ring lesion; Head and neck; Oral cavity; Metastasis; Review; Outcome

## Introduction

Primary signet-ring cell adenocarcinoma in the head and neck is extremely rare and over 96% of primary lesions arise from the gastric mucosa.[1,2] In the head and neck, they typically arise from minor salivary glands and have only recently been characterised as a separate entity.[1] Their incidence and biological behaviour are still uncertain, but a characteristic feature is the marked presence of mucin containing signet-ring cells. We found seven published papers that described 16 patients with this condition,[1–7] and here describe a further case of this uncommon lesion which was associated with an aggressive clinical course.

## Case study

A 56-year-old man presented with a 3 cm, fixed, non-painful, submucosal lump in the left retromolar trigone, the overlying mucosa was unremarkable. This lesion was found incidentally by his dental practitioner. Computed tomography (CT) showed a swelling of the soft tissue adjacent to the external margin of the left posterior mandible, with a lucent area of osteolysis (Fig. 1). There was evidence of enlarged lymph nodes at the left internal jugular chain and indeterminate lymph nodes at the left submandibular area.

He underwent an incisional biopsy, which had histomorphological features and alcian blue positivity in keeping with a high-grade signet-ring adenocarcinoma (Figs. 2a-2c) and it stained strongly for cytokeratin (CK) 7, AE1/AE3, and EMA. There was weak expression of CA19-9 and Ber-EP4, but no staining with CK5, CK14, CK19, CK20, CD10, TTF-1, or thyroglobulin. The working diagnosis was of a primary tumour of the salivary gland. The differential diagnoses included metastasis from a gastrointestinal primary, mucin producing salivary duct carcinoma, mucoepidermoid carcinoma, and metastatic primary signet-ring adenocarcinoma of the lung.

Given the rarity of this type of neoplasm, he had a full staging CT of his chest and abdomen, magnetic resonance imaging (MRI) of his abdomen and pelvis, in addition to upper and lower gastrointestinal endoscopic examinations. These additional investigations did not support a diagnosis of a lung or gastrointestinal primary tumour. In the absence

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http://dx.doi.org/10.1016/j.bjoms.2016.04.013

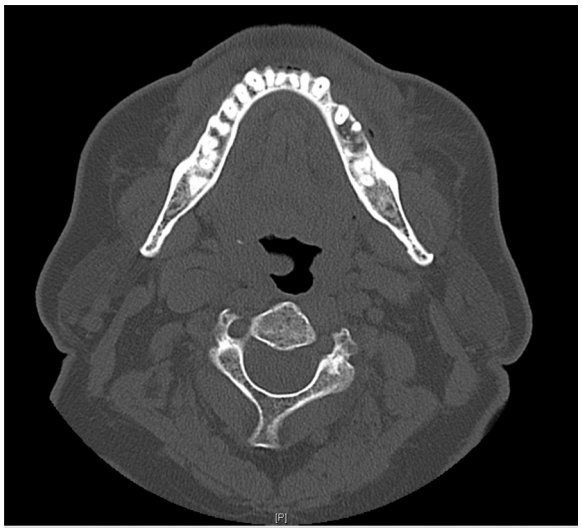

Fig. 1. Axial computed tomogram that shows some swelling of soft tissues adjacent to the external margin of the left mandible, with a lucent area of osteolysis.

of clinical or radiological findings to suggest metastases in the head and neck, we made a diagnosis of primary signet-ring cell adenocarcinoma. The patient went on to undergo radical surgery, which included wide local excision, with rim resection of the mandible, left selective neck dissection (levels I-IV), and reconstruction with a radial forearm free flap.

The results of the final histopathologcal examination were identical to the initial biopsy examination, with evidence of aggressive features that included an infiltrative pattern of invasion, with perineural and lymphovascular spread. The tumour was completely excised with clear margins. Of note, lymph nodes containing metastatic carcinoma were seen in all levels of the neck, with evidence of extracapsular spread in the lymph nodes at Level IIa, but the mandible was not involved. We concluded that the radiolucency in the left posterior mandible, seen on initial CT, was caused by a previous dental infection. His disease was finally staged pT3 N2b M0. The patient went on to receive adjuvant radiotherapy.

Six months after his initial operation, he reported to the follow-up clinic with new symptoms, including back and shoulder pain, which raised concerns about possible recurrence. He underwent a bone scan that confirmed widespread bony metastases (Fig. 3), that were most likely to be metastatic dissemination from the primary tumour. He was subsequently referred to specialist palliative care and eventually died from disseminated disease nine months after his initial presentation.

## Discussion

Several salivary gland tumours can show as a component signet-ring cell morphology, so the presence of "true" signet-ring cell adenocarcinoma in the salivary glands remains a rare and poorly understood entity.

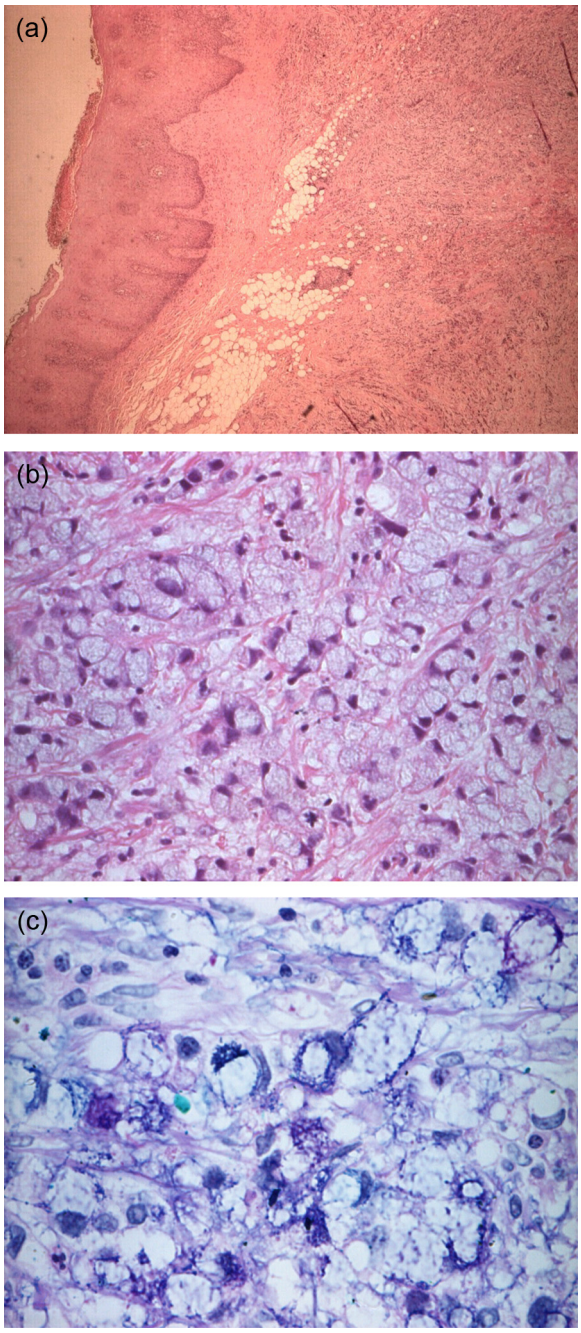


Fig. 2. Photomicrographs of biopsy examination that shows primary signet-ring cell adenocarcinoma of salivary origin.
Fig. 2a. Low power, with mucosa on the left.
Fig. 2b. High power, with typical signet-ring cell morphology.
Fig. 2c. Strong staining with periodic acid–Schiff-Alcian blue.

We evaluated the cases previously published to better understand this rare head and neck neoplasm (Table 1). The mean age was 57 years (range 18 to 82 years), with a slight preponderance of women (n = 10). The palate represented the most common site (n = 6), with 14 arising from minor salivary glands and 3 within the parotid gland. Most tumours were treated by excision alone, but two patients were found to have cervical metastases at the time of initial presentation,