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Address:
 Seog-Kyun Mun
 Department of Otorhinolaryngology-Head
 and Neck Surgery
 Chung-Ang University
 Yongsan Hospital
 Hangangno 3-ga
 Yongsan-gu
 Seoul 140-757
 Republic of Korea
 Tel: +82 2 748 9847
 Fax: +82 2 792 6642.
 E-mail: entdoctor@cau.ac.kr

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Case Report

Head and Neck Oncology

Clear cell carcinoma of the major salivary glands in an HIV-infected patient

J. López-Quiles¹, E. Ferreira¹,
 J. A. Jiménez-Heffernan²,
 M. Del Canto¹

¹Department of Oral and Maxillofacial Surgery, Hospital La Zarzuela, Madrid, Spain;

²Department of Pathology, Hospital La Zarzuela, Madrid, Spain

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Abstract. Clear cell carcinoma is a rare type of salivary gland carcinoma. It has a low degree of malignancy and long-term prognosis is favourable after surgical removal. The authors describe the case of a human immunodeficiency virus (HIV) infected 43-year-old woman who presented with a tumour on the floor of the mouth. After biopsy, left suprahyoid lymph node dissection and removal of the submandibular and sublingual glands was performed, followed by radiotherapy. Histologically, the tumour presented the characteristic features of hyalinizing clear cell carcinoma, defined as a variant of clear cell carcinoma by the latest World Health Organization classification. Hyalinizing clear cell carcinoma has a characteristic histological pattern and, to date, there is insufficient information to determine whether both forms behave similarly or differently. The present case illustrates a highly uncommon tumour variant occurring in a HIV-infected patient. To date, this association has not been described in the medical literature. The low grade of malignancy reported for this tumour demands a precise diagnosis and complete tumoral excision.

Keywords: clear cell carcinoma; human immunodeficiency virus; HIV; major salivary glands.

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Human immunodeficiency virus (HIV) infected patients often experience an increase in the size of the salivary glands, due to a wide range of conditions, includ-

ing inflammatory processes, infections and neoplasms. In some patients, it is the first sign of disease. An understanding of the underlying salivary pathology is

necessary to guarantee correct treatment³. HIV patients have approximately a 40% chance of developing a malignant disease, a rate that is expected to rise due to their

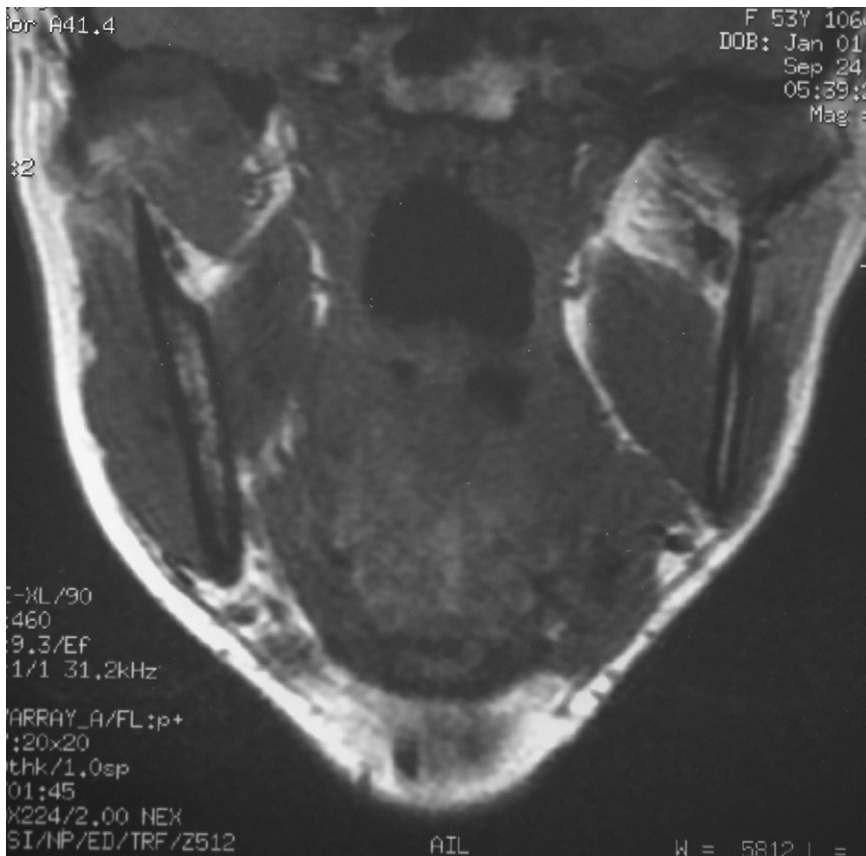


Fig. 1. MRI showing a tumour on the left side of the floor of the mouth.

increased survival⁴. The most frequent head and neck tumours in HIV-infected patients are Kaposi's sarcoma (over 15%) and non-Hodgkin's lymphoma (3–10%)^{4,7}. During the last few years, the risk of developing oral squamous cell carcinoma has increased⁴.

Clear cell carcinoma (CCC) is a rare salivary gland tumour that shows a low grade of malignancy^{1,2}. It has a certain potential for recurrence and metastasis, but long-term prognosis is favourable after removal, with or without radiotherapy¹. It most often affects minor salivary glands⁵ and occurs predominantly in women as a painless, submucosal mass^{1,9}. The latest WHO classification has defined it as a malignant epithelial neoplasm composed of a population of monomorphic cells having clear cytoplasm⁵. Many types of salivary gland neoplasms may contain a component of clear cells, but CCC is distinguished by the absence of the typical characteristics of other neoplasms and its monomorphic population of clear cells⁵.

Hyalinizing CCC (HCCC) is considered a variant of CCC, distinguished by a densely hyalinized stroma. Both tumours have similar clinical characteristics⁵.

The authors describe the case of a HIV-positive female patient who developed HCCC in a sublingual location. It is interesting not only because of the relation between HIV infection and the tumour

but also because it concerns a rare morphological variant and a major salivary gland.

Case report

A 43-year-old woman was referred by her dentist for the evaluation of a painless tumour on the floor of the mouth. In 1995, during a laboratory analysis she was diagnosed HIV-positive, and since then she has been receiving antiretroviral therapy (didanosine, lamivudine and efavirenz). She has been asymptomatic with no AIDS-related disorders. The tumour was detected 1 month before surgery. Laboratory analysis confirmed the HIV positivity and showed a normal CD4 count. Complementary studies included orthopantomography, which ruled out bone involvement, and MRI, which showed a tumour on the left side of the floor of the mouth (Fig. 1). No local or regional adenopathies were present. A diagnostic biopsy was carried out and a diagnosis of CCC of salivary gland was obtained. It was decided to perform a left suprahyoid lymph node dissection and to remove the submandibular and sublingual glands. The surgical sample had two nodular areas (of 3.5 and 3.9 cm) attached by a thin fibrous band (Fig. 2). The smallest had a firm, irregular, solid aspect with intermixed adipose tissue whilst the largest one corresponded to the submandibular salivary gland.

Microscopically, the tumoral nodule showed salivary gland parenchyma (sublingual) extensively infiltrated by a neo-



Fig. 2. The surgical sample had two nodular areas (3.5 cm × 3.4 cm × 1.5 cm and 3.9 cm × 3.1 cm × 2 cm) attached by a thin fibrous band.

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