Human Pathology (2012) 43, 303–306

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Human PATHOLOGY

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Case study

Signet ring cell tumor of the minor salivary gland exhibiting benign behavior

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Received 16 March 2011; revised 22 April 2011; accepted 26 April 2011

Keywords:
Signet ring cell;
Minor salivary gland;
Lip;
Benign tumor

Summary Signet ring cell (SRC) carcinomas are usually aggressive malignancies, arising most frequently in the stomach and gastrointestinal tract, but also, although less often, in other organs such as the breast, bladder, and lungs. They are particularly unusual in the salivary glands, and the aim of the present study is to report a case of a tumor of the minor salivary glands of the lower lip composed largely of SRCs but which displayed benign clinical behavior.

1. Introduction

Signet ring cells (SRCs) are characteristically found in carcinomas of high malignant potential. The gastrointestinal tract, breast, bladder, and lungs are the most common primary sites, but tumors composed of these cells have also been described—although less frequently—in other organs.

Nevertheless, SRCs do not always signify malignancy. Nonmalignant mucinous SRCs have been described in the colon of patients affected by pseudomembranous colitis [1] and in gallbladders excised for gallstones [2]. In the salivary glands, a case of oncocytic cystadenoma of the parotid gland with prominent SRCs [3] and a further case of SRC proliferation in the minor glands of the lip [4] have been reported. The latter study describes SRCs in an enlarged minor salivary gland of the upper lip as a possible consequence of mechanical injury.

Otherwise, the presence of true SRCs in salivary gland neoplasms and tumorlike lesions is rare and their significance relatively poorly understood.

The purpose of the present study is to describe an unusual case of a tumor of the minor salivary glands composed largely of SRCs but showing benign histopathological features and behavior.

2. Case report

A 31-year-old woman presented with a small nodule of the lower lip that was surgically removed. The patient was otherwise in good health with no significant medical history, and in particular, she had had no previous tumors, either in the head and neck region or elsewhere. After the histological diagnosis, the patient underwent an extensive clinical workup (including a computed tomography scan and gastroscopy) to exclude the presence of additional neoplasms either locally or in other organs. As no other tumor was detected, the patient did not receive any additional treatment.

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0046-8177/$ – see front matter © 2012 Elsevier Inc. All rights reserved.
doi:10.1016/j.humpath.2011.04.025

More than 7 years after surgery, the patient remains alive and well, with no evidence of disease locally or systemically.

3. Pathologic findings

Macroscopically, the nodule had a smooth external surface, measuring 0.8 cm along the longitudinal axis; on cut section, it was focally cystic.

Histologically, the nodule was completely encapsulated (Fig. 1). Outside the capsule, a thin rim of normal minor salivary gland with a nonspecific inflammatory infiltrate was visible.

The lesion itself was composed mainly of cells each with a large intracytoplasmic vacuole containing mucin (positive with Alcian blue pH 2.5 but negative with periodic acid-Schiff [PAS] after diastase digestion) (Fig. 2), displacing the nucleus toward the periphery and distorting it, thus having the appearance of an SRC. These cells displayed bland cytological features lacking nuclear atypia, and no mitotic figures were present. Some ducts, which seemed to be in continuity with the neoplastic proliferation, contained SRCs, thus giving the appearance of an in situ lesion and consequently indicating that the tumor could have possibly originated in these ducts. The cystic part of the lesion contained papillary projections composed of fibrovascular cores lined by multiple layers of SRCs. No epidermoid or intermediate cells were seen. Tumor necrosis and evidence of invasion were absent, and no atypical mitotic figures were identified.

Immunohistochemically, the SRCs strongly stained for cytokeratin (CK) 7 and with an antimitochondrial antibody, but they lacked immunoreactivity for p63, c-kit, and salivary gland amylase. CK4 and smooth muscle actin were negative in the tumor cells, but they stained the nonneoplastic basal and myoepithelial cells of the surrounding salivary gland (Fig. 3).

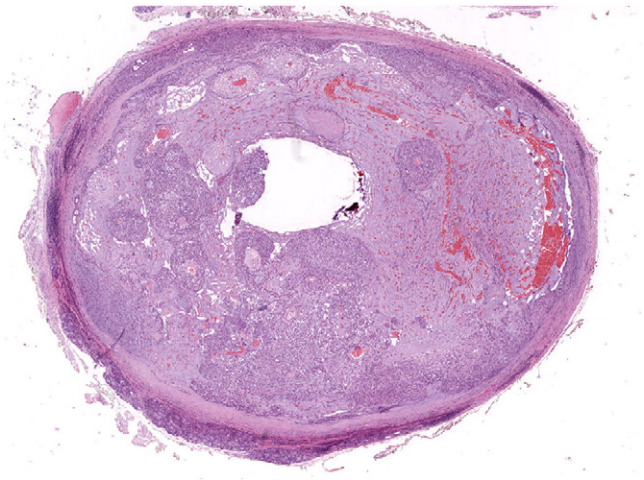

Fig. 1 At low magnification, the nodule is encapsulated and centrally cystic.

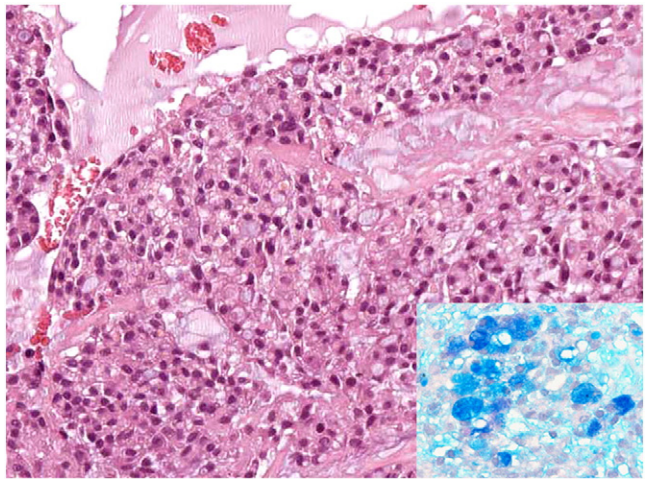

Fig. 2 At higher power, the lesion is entirely composed of cells with a large intracytoplasmic vacuole; the vacuole is filled with Alcian blue-positive mucin (inset).

The Ki-67 proliferative rate was low, not exceeding 2% of the total neoplastic population.

To rule out the possibility of a metastatic lesion, additional immunohistochemical staining was performed. TTF1, CK20, MUC2, and CDX2 antibodies gave negative results, which largely excluded a gastrointestinal or a lung carcinoma. Similarly, negativity for estrogen and progesterone receptors together with strong positivity for the E-cadherin cytoplasmic membrane rendered the diagnosis of metastatic lobular carcinoma of the breast extremely unlikely [5].

4. Discussion

The presence of a tumor in the salivary glands composed largely of SRCs raised the possibility of several differential

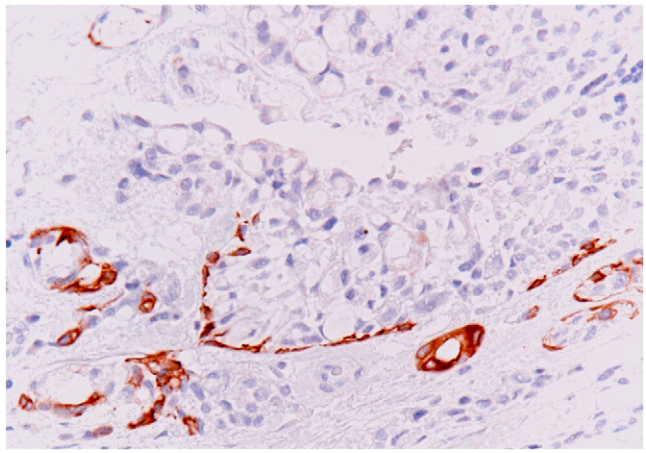

Fig. 3 CK14 stains the basal cells of some residual ducts located at the periphery of the lesion. The same ducts contain some SRCs similar to those observed in the lesion. This finding supports the salivary duct origin of the present lesion.