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Salivary Duct Carcinoma: A Rare Entity

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Abstract

Salivary duct carcinoma (SDC) is a rare malignancy that most commonly affects males in the fifth and sixth decades of life. Patients commonly present with a fast-growing form mass, often around the facial nerve. There are no known etiologic factors. Their most peculiar feature is morphological and immunohistochemical similarity to ductal carcinoma of the breast. Therapeutic approach is non-consensual because of the limited data but an extensive surgical resection has been advocated, along with postoperative chemoradiation in selected cases. Antiandrogen therapy may be useful in patients with advanced SDC. The rate of locoregional recurrence is high and the prognosis for survival is poor.

We present the case of a 43 years-old patient with a right parotid lump of small dimension and rapid growth. Facial nerve deficits were not apparent and neck palpation identified no enlarged lymph nodes. MRI described a small lesion of oval shape and well defined contours. Fine-needle aspiration biopsy raised the possibility of a myoepithelial tumour and a superficial parotidectomy was performed. The patient experienced no complications during the postoperative healing. There were no facial nerve deficits. Microscopic examination concluded to a SDC. It was decided by a multidisciplinary team to apply radiation therapy to the surgical bed. At two years of follow up the patient remains with no clinical or imagological evidence of tumour recurrence.

Keywords: Salivary duct carcinoma; Chemoradiation; Metastasis; Immunohistochemical; Surgery

Introduction

Salivary duct carcinoma (SDC) is a primary malignancy of the salivary glands that was first described by Kleinsasser et al. [1]. This is a rare and aggressive adenocarcinoma that histologically resembles a mammary duct carcinoma [2]. An extensive treatment with total or radical parotidectomy plus neck dissection plus chemoradiation in selected cases has been advocated [3,4]. We present and discuss a case of SDC of the parotid gland diagnosed at an early stage and submitted solely to a superficial parotidectomy plus radiation therapy. The authors wish to stress that early diagnosis of small dimension SDC may allow a less extensive approach given that a tight control of local recurrence and metastasis is provided.

Case Report

A 43 years old male with no relevant past history presented with a small painless swelling in the right parotid gland region that had been apparent for two months. The patient had no history of fever or constitutional symptoms. Physical examination revealed a firm lump not fixed to adjacent structures.

Facial nerve deficits were not apparent and neck palpation identified no enlarged or pathological lymph nodes. Magnetic resonance imaging described a space occupying lesion in the inferior portion of the right parotid, with 9 × 14 × 10 mm, an oval shape and well defined contours, no relevant mass effect and no aggressiveness signs (Figure 1). A fine-needle aspiration biopsy identified cells with a myoepithelial pattern and the possibility of a myoepithelial tumour was raised.

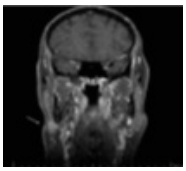

Figure 1: Gadolinium-enhanced T1-weighted MRI with fat saturation presenting a nodular lesion in the inferior pole of the right parotid gland (arrow).

A modified Blair incision was used to approach the tumour. A superficial parotidectomy was performed. Intraoperatively it was verified involvement of a small branch of the facial nerve by the tumour that had to be sectioned (Figure 2).

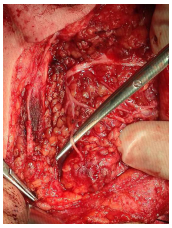

Figure 2: Intraoperative view with tumour involvement of a facial nerve branch.

Upon histopathological examination a definitive diagnosis of a SDC was presented (Figure 3). In order to further investigate tissue samples several immunohistochemical markers were analyzed. HER2 (Figure 4) and GCDFP15 (Figure 5) were positive.

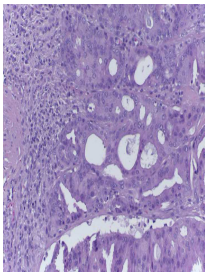

Figure 3: H&E stain, 20x magnification.

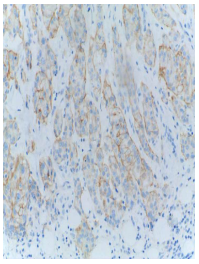

Figure 4: HER2+, 20x magnifications.

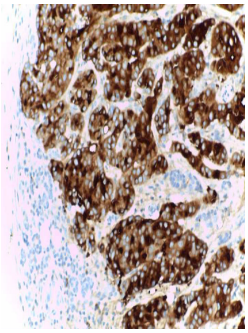

Figure 5: GCDFP15 stain, 10x magnifications.

The patient experienced no complications during the post-operative healing. There were no facial nerve deficits (Figure 6). The case was presented to a multidisciplinary head and neck cancer team and it was decided to start the patient on postoperative radiation therapy that was applied to the surgical bed sixty days after surgery.

Results

The final pathological stage was pT2NxMx. At two years of follow-up the patient has experienced no complications from surgery or radiation therapy and follow-up studies have revealed no local or regional recurrence of disease.

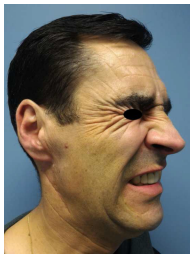

Figure 6: Post-operative photograph showing post-operative scar and no neurologic deficits.

Discussion

SDC is a malignant condition that accounts for up to 2% of all primary salivary epithelial neoplasms. Most patients are over 50 years old and there is approximately a 4:1 male to female ratio. It arises mainly in the parotid glands, but cases have been described in the submandibular glands and occasionally in the minor glands [5,6].

Patients commonly present with a fast-growing firm mass often around the facial nerve [5]. Clinical symptoms may not be present, while if they occur, mostly often pain, swelling and facial paresis are reported [7]. Cervical adenopathies are identified on palpation in 35% of patients [8].

There are no known etiologic factors. One case was reported in a patient with long standing chronic obstructive sialadenitis and another in a patient with IgG4 related sclerosing disease of the parotid [9,10]. In a percentage of cases, it develops from a pre-existing benign lesion. In a review of 75 cases, Gilbert et al. found that 41% of the cases were carcinoma ex pleomorphic adenomas [11,12].

Imaging findings, especially CT scan and MRI features, are nonspecific but they are helpful in the diagnosis of malignancy and in the management. They can indicate the malignant nature of the tumour by showing ill borders or an infiltration of the adjacent tissue. Positive diagnosis is based on histologic examination. The means of diagnosis consist of fine needle aspiration cytology which is useful but not always reliable, fine needle aspiration specimen, and surgical specimen [2].

Macroscopically, the aspect is of a yellowish or greyish-white tumour that may be nodular, multinodular, cystic or infiltrating, surrounded by fibrosis with hemorrhagic areas, necrosis and cystic degeneration. It is usually a firm ill-defined mass; a well-circumscribed nodule within the tumour may indicate a pre-existing pleomorphic adenoma. There may be intratumoral calcification [5,13].

Microscopically, the most peculiar feature is the similarity to ductal carcinoma of the breast. The tumour is composed of an intraductal and invasive components. Intraductal component (carcinoma in situ) is cribriform, papillary, solid with comedolike central necrosis. The infiltrative component is made of glands, cords of cells with desmoplastic reaction [2,5,14].

In addition to the usual type of SDC, a few less common morphological variants have been reported: papillary, micropapillary, mucin-rich, sarcomatoid and oncocytic, as well as pure in situ cases [2,5].

As SDC resembles breast carcinoma morphologically, not surprisingly there are immunohistochemical similarities, with HER2 protein overexpression reported in up to 90% of cases of SDC [15,16]. On the other hand, whilst in breast cancer, estrogen receptor α and progesterone receptor are found in >75% of cases, positivity for these markers is exceptional in SDC [**17**]. Androgen receptors and prostate specific antigen expression has been frequently reported [2,18].

The differential diagnosis of SDC includes primary oncocytic, mucoepidermoid and myoepithelial carcinomas, as well as metastatic melanoma, squamous, breast and prostate carcinomas [5]. The most relevant morphologic feature is the presence of an intraductal component which is specific of the diagnosis [2].

Therapeutic approach is non-consensual because of the limited data but a total parotidectomy is recommended even in T1 tumours because of local disease recurrence [4]. If facial paralysis is present, a radical parotidectomy is mandatory. In patients with involvement of the submandibular or minor salivary glands, tumour resection with wide margins of surrounding tissue is indicated to control local disease [2,19]. Neck dissection is necessary to allow ipsilateral lymph node excision [20]. Postoperative radiation therapy is indicated in case of extraparotid extension, pathological resection margins, cervical lymph node metastasis, lymphatic embolus, and/or neurologic invasion. Chemotherapy is generally reserved for metastatic forms of the disease [11,21].

The rate of locoregional recurrence is high and the prognosis for survival is poor. Over 60% of patients die of the disease within 5 years of the initial diagnosis, despite radical surgery and chemoradiation. About 33% develop local recurrence and >50% distant metastasis at sites including lungs, bones, liver, brain and skin [5] Jaehne et al. found that HER2 and p53 expression are statistically linked to early local disease recurrence, distant disease metastasis and survival rates [19].

In our case, a definitive diagnosis was not apparent until after tumour's surgical excision. The unsuspected nature of the lesion led to a more conservative approach with a superficial parotidectomy and no neck dissection. It was decided by a multidisciplinary team to do no further surgery, apply radiation therapy and provide the patient with a tight follow up that will allow early diagnosis of an eventual locoregional recurrence.

SDC has been described as an aggressive tumour with high local recurrence rate and development of distant metastasis. It has been advocated an extensive treatment with total (or radical) parotidectomy plus neck dissection in all cases plus chemoradiation in selected cases, but the best treatment approach has not yet been stablished and more studies with different treatment approaches need to be published to better understand the nature and best treatment for this malignant tumour.

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