Journal of Medical & Surgical Pathology

Donato et al., J Med Surg Pathol 2016, 2:1
DOI: 10.4172/2472-4971.1000142

Case Report Open Access

Metastatic Salivary Duct Carcinoma of the Submandibular Gland Presenting as a Poorly Differentiated Carcinoma of Unknown Primary: A Case Report

Rosemarie Di Donato*, Samer Khader, Antonio Cajigas and Laleh Hakima

Albert Einstein College of Medicine, New York, USA

***Corresponding author:** Rosemarie Di Donato, Department of Pathology, Albert Einstein College of Medicine/Montefiore Medical Center, 111 East 210th Street, Bronx, NY 10467, USA, Tel: 7189204976; E-mail: rdidonat@montefiore.org

Received date: August 04, 2016; **Accepted date:** November 26, 2016; **Published date:** November 30, 2016

Introduction

Salivary duct carcinoma is an aggressive malignancy that most commonly arises in the parotid gland. Few case reports describe this entity occurring in the submandibular gland. This entity can metastasize locally early and therefore is an important differential diagnosis of metastatic disease in cervical lymph nodes.

Clinical Presentation

A 53 year old male presented to his primary care doctor with a palpable neck mass for the past four months. At that time he was given antibiotics with no change in the mass. The patient was a 20 pack-year, current smoker. He was sent for an ultrasound-guided fine needle aspirate (FNA) of the mass. The FNA of a right level I lymph node was performed and findings were consistent with a poorly differentiated carcinoma, possibly reflecting a primary salivary gland lesion or less likely a pulmonary lesion. Upon follow-up physical exam the patient was found to have mild asymmetry and firmness of the right base of tongue as well as a 1.0 x 1.0 cm, firm submandibular mass. The patient was referred to otorhinolaryngology. A PET scan was performed and showed multiple avid cervical lymph nodes in the right neck, focal activity in the supero-medial aspect of the right submandibular gland, as well as very small bilateral lung nodules. At this point, a primary site in the submandibular gland was favored; yet the aerodigestive tract was not excluded. Pleural fluid was sent for cytological analysis and subsequently the patient underwent a diagnostic laryngoscopy, biopsy, esophagoscopy, and right selective neck dissection.

Pathological Findings

The smears from the FNA showed many irregularly shaped, branching clusters and sheets of malignant cells with pleomorphism, large nuclei, prominent nucleoli and a moderate amount of cytoplasm in a background of necrotic debris and lymphoid cells, findings were consistent with a poorly differentiated carcinoma. Immunohistochemical staining was positive for cytokeratin 7 and Cam 5.2, and negative for p53, p63, p16, TTF-1, and NapsinA. Pleural fluid was also sent for cytological analysis and was found to be positive for malignant cells. The smears and cell block were consistent with metastatic adenocarcinoma. The patient underwent a diagnostic laryngoscopy, biopsy, esophagoscopy, and right selective neck dissection. Intraoperative consultation was performed on the right base and lateral tongue, right submandibular gland and right neck dissection level IB. The right base and lateral tongue was negative for tumor. The right submandibular gland, however was positive for carcinoma. The right neck level IB lymph node was found to have poorly differentiated carcinoma with areas suspicious for ductal carcinoma in situ suggestive of salivary ductal carcinoma. The final surgical pathology diagnosis was issued as follows: the right base and lateral tongue were found to have benign squamous mucosa with associated tonsillar tissue, no tumor seen. The submandibular gland and right level IB lymph node showed salivary duct carcinoma measuring 1.7 cm, poorly differentiated with tumor necrosis and infarct. Extensive perineural invasion and foci of lymphovascular invasion was seen. Additionally one of four lymph nodes was positive for metastatic carcinoma. The submandibular gland had an infiltration of crushed atypical cells compatible with salivary duct carcinoma. Androgen receptor immunohistochemical staining was performed on the specimen and was negative.

Approximately two months later the patient underwent a right upper lobe wedge resection of a pulmonary nodule which was positive for metastatic carcinoma, compatible with known salivary duct carcinoma. Immunohistochemical staining was positive for cytokeratin 7 and p16, and negative for TTF-1, Napsin A and p40, supporting the diagnosis.

Discussion

Salivary duct carcinoma is a very rare malignancy. It has been found that the vast majority, around 75% of cases affect the parotid gland. Only about 15% of cases have been found to arise in the submandibular gland. It is also a highly aggressive tumor, with approximately 65-73% of patients diagnosed with advanced disease (stage III or IV) at presentation. It has a male predominance with a mean age of diagnosis of 58.8 years [1]. Additionally it has been found that 50% of the patients have lymph node involvement at presentation [1]. Due to its rarity and highly aggressive nature, it is important to correctly diagnose upon initial presentation in order to provide appropriate therapy to the patient. The disease can metastasize widely and distant metastases have most commonly been found in the lungs, but brain and bone metastases were also noted. This case study highlights the aggressive nature of the tumor, with avid lung nodules discovered soon after presentation on PET scan, and positive lung metastasis documented at about two months after initial diagnosis.

Salivary duct carcinoma has been found to have negative prognostic indicators, such as perineural invasion, vascular invasion, and extracapsular spread. The case presented here was positive for perineural invasion. Lymph node involvement has also been found to be associated with a poor prognosis [2]. The patient presented here had lymph node involvement upon diagnosis, again highlighting the aggressive nature of the tumor. The patient presented here was also found to have positive pleural fluid cytology consistent with a salivary duct carcinoma primary.

Salivary duct carcinoma is therefore an important consideration in metastatic carcinoma to regional lymph nodes as well as to sites such

as the lung. Histologically this tumor resembles ductal carcinoma of the breast with both intraductal and invasive components. Cytologic distinction from other high grade malignancies is often difficult. Primary squamous cell carcinoma of the salivary gland is very rare and metastasis is far more common. Clinical history is necessary for exclusion. Squamous cell carcinoma is typically p63 positive and can also show positivity for p53. Metastatic adenocarcinoma of pulmonary origin can have similar cell clusters and cytologic features; however it is Napsin and TTF-1 positive on immunostudies [3]. The apocrine features of salivary duct carcinoma can be confused with oncocytic neoplasms such as acinic cell carcinoma. However, salivary duct carcinoma cells have a higher nuclear to cytoplasmic ratio, less granular cytoplasm and many three-dimensional clusters. Acinic cell carcinoma also lacks the nuclear atypia and comedo necrosis of salivary duct carcinoma. High grade mucoepidermoid carcinoma has a higher proportion of squamoid cells with cytologic atypia compared to cystic areas and can resemble salivary duct carcinoma with solid and infiltrative growth. However salivary duct carcinoma typically has extensive necrosis and can also have papillary architecture. It lacks the mucinous and intermediate cells of mucoepidermoid carcinoma.

This case is one of the few documented salivary duct carcinomas arising in the submandibular gland as opposed to the more common location of the parotid gland, and must be considered in the differential diagnosis of all major and minor salivary gland tumors.

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