CASE REPORT

Late Neck Metastasis of Clear Cell Salivary Carcinoma of the Soft Palate

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ABSTRACT

Introduction: Clear cell carcinoma is a rare form of salivary gland tumor, most commonly affecting the palate and tongue. We present a patient with cervical lymph node metastasis 28 years after treatment for clear cell salivary carcinoma of the soft palate.

Case presentation: A new neck mass was found in a 69-year-old man with a remote history of clear cell salivary carcinoma of the soft palate. Fine-needle aspiration biopsy demonstrated no evidence of malignancy; however, excisional biopsy revealed carcinoma. Neck dissection was performed and final pathologic review demonstrated clear cell salivary carcinoma metastatic to a cervical lymph node.

Discussion: Clear cell salivary carcinoma may be less indolent than previously believed. Metastases to cervical lymph nodes and distant sites have been reported in one-fourth of patients and can occur many years after treatment. Immunohistochemical staining is a valuable adjunct for differentiating clear cell carcinoma from other neoplasms that demonstrate clear cell components.

Conclusion: This rare case of late neck metastasis after clear cell salivary carcinoma of the soft palate highlights the need for high clinical suspicion of recurrence many years after treatment and indicates the role of neck dissection for diagnosis and treatment in patients with possible nodal spread.

Keywords: Salivary gland tumor, Clear cell carcinoma, Late metastasis.

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INTRODUCTION

Clear cell carcinomas, a variant of adenocarcinomas, are one of the rarest forms of salivary gland tumors, representing less than 1%. Diagnosis is often difficult because the histologic features of these lesions overlap with those of several more common salivary malignancies. Recent evidence suggests the rate of metastasis and recurrence for clear cell carcinoma may be higher than previously thought. We report a case of metastatic clear cell carcinoma to a cervical lymph node 28 years after excision and radiation therapy for clear cell carcinoma of the soft palate. Appropriate Institutional Review Board (IRB) review was undertaken.

CASE REPORT

A 68-year-old man presented with a 1-month history of a new left neck mass. He had a history of clear cell carcinoma of the soft palate, which had been treated 28 years before with surgical excision and external beam radiation therapy (62.99 Gy in 30 fractions over 42 treatment days). Physical examination revealed a 2 cm firm, well-defined, mobile mass in the left jugulodigastric area. Fine-needle aspiration biopsy (FNAB) suggested reactive lymphadenopathy. Over the following month, the mass did not decrease in size. Given his history of malignancy, the decision was made to perform an excisional biopsy with possible neck dissection.

At the time of surgery, direct laryngoscopy revealed no evidence of residual carcinoma at the site of the previous excision or elsewhere in the upper aerodigestive tract. The mass and lymph node components of level IIA were mobilized and excised, with preservation of the accessory nerve and jugular vein. Frozen section analysis of the mass revealed a carcinoma of unlikely squamous cell origin. Consequently, a selective neck dissection was performed. The patient tolerated the procedure and did well post-operatively.

The final pathologic findings from the excised mass were indicative of metastatic clear cell carcinoma (Fig. 1). Tumor cells stained positive for keratin and negative for thyroid transcription factor (TTF-1), napsin and renal cell carcinoma (RCC) (Fig. 2).

Intracytoplasmic glycogen was diastase sensitive on periodic acid-schiff (PAS) stain. No evidence of malignancy was detected in the other lymph nodes (0/28).

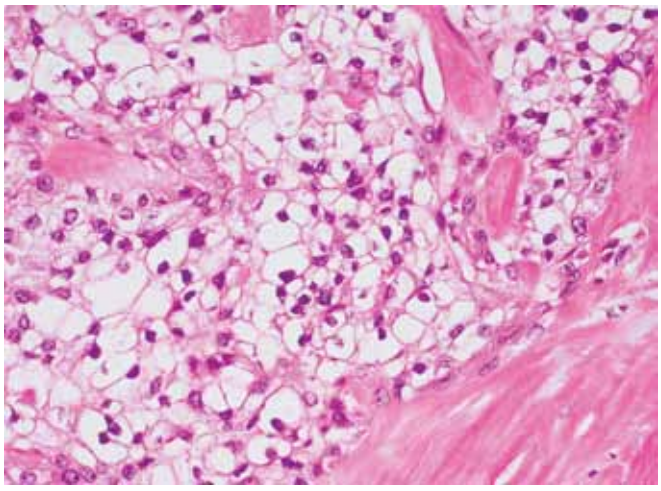

Fig. 1: Photomicrograph (×200) of the biopsy showing a tubular and focally trabecular growth pattern with prominent hyalinization

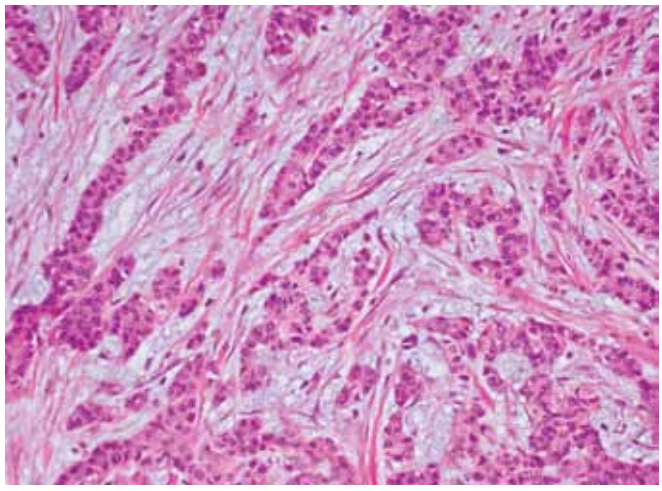

Fig. 2: Photomicrograph of an area of clear cells (×400). These were negative for RCC markers on IHC staining

REVIEW OF LITERATURE WITH DISCUSSION

With fewer than 100 cases reported in the literature, clear cell carcinoma is a rare form of salivary gland neoplasm that typically arises from minor salivary glands and most commonly affects the soft palate (27.5%) or tongue (25.5%).[1] Most cases present as a painless, slow-growing submucosal mass; however, ulceration of the mass as well as pain and speech alteration have been reported.[1,2] Diagnosis can be challenging, as clear cells constitute a minor component of the cellular content in these lesions and are also observed in many other benign and malignant salivary gland tumors, including mucoepidermoid carcinoma; myoepithelioma and myoepithelial carcinoma, oncocytoma and oncocytic carcinoma, acinic cell carcinoma and adenoid cystic carcinoma.[3,4]

The histomorphologic features of clear cell carcinoma may aid in establishing a diagnosis. Clear cells are typically seen in cords, trabeculae or clusters surrounded by a hyalinized stroma.[5] Immunohistochemical (IHC) staining is an important adjunct in diagnosis and should be strongly considered in patients with a history of prior malignancy. Clear cell tumors are diastase sensitive on PAS stain and typically positive for cytokeratin A1/A3, CAM5.2 and epithelial membrane antigen. They stain negative for mucicarmine, calponin and smooth muscle actin as well as CD10, a marker of RCC, and thyroglobulin and TTF-1, markers of clear cell thyroid carcinoma.[4-6]

Salivary gland clear cell carcinomas may be less indolent than previously believed.[7-9] Metastases to cervical lymph nodes or distant sites have been reported in 25% of patients at the time of presentation.[6] Moreover, salivary gland carcinomas may recur after extended disease-free intervals. Cumulative 10-year and 15-year recurrence rates of 13 and 18% respectively, have been reported for patients who were free of disease at 5 years.[10] Late recurrences have been described up to 12 years after treatment. The incidence of late recurrence may be higher than that reported in the literature, as many cases lack long-term follow-up data.[1]

To detect a recurrence of salivary gland malignancy, FNAB alone may not be sufficient. A negative FNAB finding has been reported to carry a low predictive value, with more than half of the negative FNAB specimens at a tertiary care cancer center found to represent neoplasms on final histological assessment.[11] As these findings suggest and as the present case illustrates, additional work-up may be warranted even in the context of a negative FNAB if the level of clinical suspicion is high.

CONCLUSION

This rare case of recurrent metastatic clear cell carcinoma 28 years after initial diagnosis and treatment carries several implications for the management of clear cell salivary gland carcinoma. First, the prevalence of cervical lymph node metastasis underscores the importance of thorough examination of the neck and suggests a role for neck dissection in addition to wide local excision for patients with suspected locoregional spread.[12] Second, given the potential for late recurrence, continued vigilant surveillance is warranted even after an extended disease-free interval. Finally, if the clinical presentation is concerning for possible recurrence, a negative finding on FNAB is not sufficient to rule out malignancy and should not deter additional work-up.

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