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Original research Articles

Neuroendocrine Parotid Mass: Atypical Salivary Gland Case

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Abstract:- Neuroendocrine tumors (NETs) are notably rare and typically located in the gastrointestinal tract and lungs. Neuroendocrine small cell carcinoma (SCC), a subtype of NETs, is frequently found in the nasal cavity, while primary neuroendocrine SCC of the parotid gland is exceedingly rare, constituting less than 1% of all salivary gland tumors. This report details the case of a 79-year-old Hispanic male with an unusual parotid neuroendocrine tumor, as confirmed by the final pathology report. To our knowledge, there are no existing data on neuroendocrine parotid tumors in the Latin-American population, and no previous cases have been documented. This report presents the first such case in this demographic, highlighting diagnostic approaches and recommended management strategies.

Keywords: parotid, neuroendocrine tumors, parotid neuroendocrine mass, parotid carcinoma

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Teaching Points:

1. Treatment involves surgical excision of the tumor followed by postoperative radiation and chemotherapy.
2. Differentiation of NETs from the more prevalent squamous cell carcinomas and metastases from other primary tumors is crucial due to the distinct therapeutic approaches and prognoses.

Introduction:

Carcinomas of the major salivary glands are uncommon, accounting for about 11% of all oropharyngeal neoplasms in the United States (1). These tumors are morphologically diverse, with poorly understood causes and risk factors. The parotid gland is the most frequently affected, followed by the submandibular glands. Common etiologies include mucoepidermoid carcinoma, squamous cell carcinoma, adenoid cystic carcinoma, and adenocarcinoma. Conversely, lymphoepithelial carcinoma and neuroendocrine tumors, including small cell and large cell carcinomas, are among the least common salivary gland tumors.

The grading criteria for primary neuroendocrine tumors of the salivary glands are not well established. The pulmonary World Health Classification is often used to classify extrapulmonary neuroendocrine carcinomas(2-3). There are four major types of neuroendocrine tumors of the lung: typical carcinoid, atypical carcinoid, small cell carcinoma, and large cell carcinoma. These tumors are rare in the head and neck region, including the salivary glands, but most reported cases refer to tumors of the parotid gland(4,5,6).Most neuroendocrine tumors in the salivary glands are undifferentiated carcinomas, either small cell or large cell carcinomas. Well-differentiated, poorly differentiated, and moderately differentiated tumors are even rarer and not frequently described in the literature.

Small cell carcinoma of the parotid gland represents a rare and aggressive malignancy among salivary gland tumors. Immunohistochemical analysis is essential for confirming the diagnosis. Positive staining for neuroendocrine markers such as synaptophysin and CD56 is typically observed, indicating neuroendocrine differentiation of the tumor cells. Additional markers, such as chromogranin, and areas of necrosis within the tumor mass further support the diagnosis.

This report presents the case of a 79-year-old male with a right preauricular mass diagnosed as a neuroendocrine neoplasm with features of small cell carcinoma. Additionally, relevant literature on salivary gland neuroendocrine malignancies, clinical presentation, diagnostic workup, and current management is reviewed.

Case Presentation

A 79-year-old male with a medical history of hypertension, deep vein thrombosis, and benign prostatic hyperplasia reported noticing a firm, non-tender preauricular mass while shaving. The mass remained stable in size. He denied symptoms such as dysphagia, facial numbness or weakness, flushing, headaches, and hypertension. Neck CT with IV contrast revealed a well-defined solid mass in the superficial right parotid gland with benign characteristics, measuring 1.5 cm x 1.2 cm x 1.2 cm. The patient underwent ultrasound-guided fine needle aspiration biopsy, which revealed malignant neoplastic processes with neuroendocrine differentiation. He was evaluated at the surgical oncology clinic and scheduled for a right superficial parotidectomy.

Surgical Procedure and Outcomes

The patient underwent a superficial parotidectomy with right neck dissection. The facial nerve was preserved. The specimen was sent to pathology for final diagnosis. The patient was hospitalized for two days, with the procedure being well tolerated. The tumor was successfully resected with no major complications, no neurological deficits, no wound infection, and no recurrence on follow-up visits. He was referred to an oncologist for further management.

Pathology

Gross examination of the mass included a right parotid gland specimen and right neck level 2, 3, and 4 excisional specimens. The final pathological diagnosis confirmed a neuroendocrine neoplasm with small cell carcinoma characteristics, displaying tightly packed small round cells with minimal cytoplasm and a high nuclear-cytoplasmic ratio. The nuclei exhibited granular chromatin and prominent nucleoli, contributing to the classic "salt-and-pepper" appearance. Numerous mitotic figures reflected the rapid cellular proliferation characteristic of small cell carcinomas. Regions of necrosis were present, emphasizing the aggressive nature of the malignancy. Immunostaining for neuroendocrine markers including synaptophysin, CD56, and chromogranin was positive.

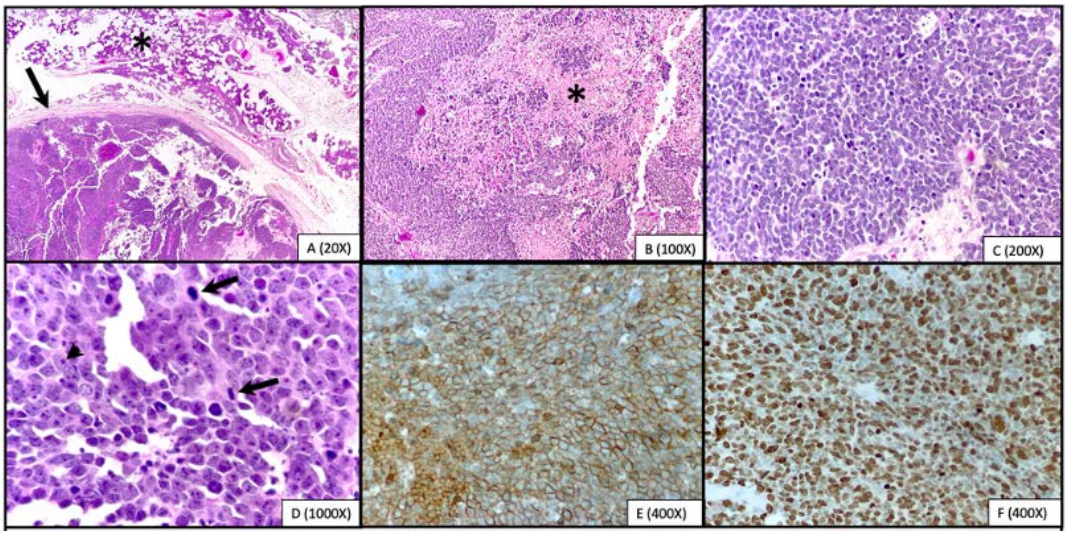

Figure 1- A: H&E stain showing an encapsulated (arrow) well-differentiated neuroendocrine tumor and surrounding salivary gland tissue (asterisk). **B:** Neuroendocrine neoplasm showing areas with necrosis (asterisk). **C and D:** Tumor showing neoplastic cells with granular pale acidophilic cytoplasm, pleomorphism, and nuclei with the typical "salt-and-pepper" appearance. Some nuclei also show vesicular nuclei with prominent nucleoli (arrowhead). Multiple mitotic figures are also seen (arrows). **E:** Tumoral cells showing strongly and diffuse membranous immunostaining for CD56. **F:** Ki67 showing a high proliferation index (more than 90%).

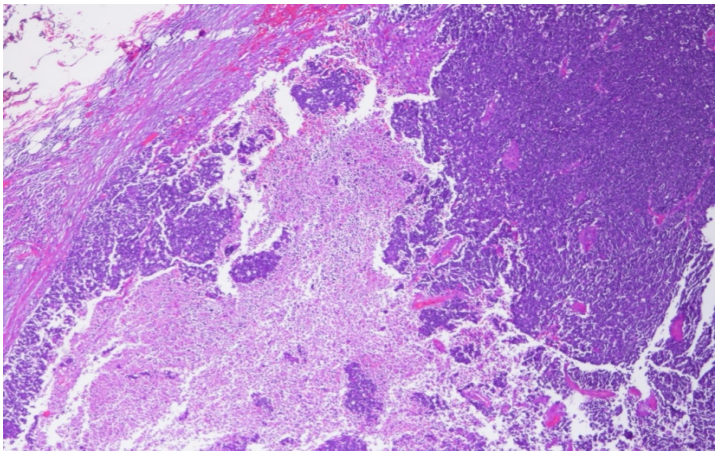

Figure 2: Neuroendocrine neoplasm with necrotic areas.

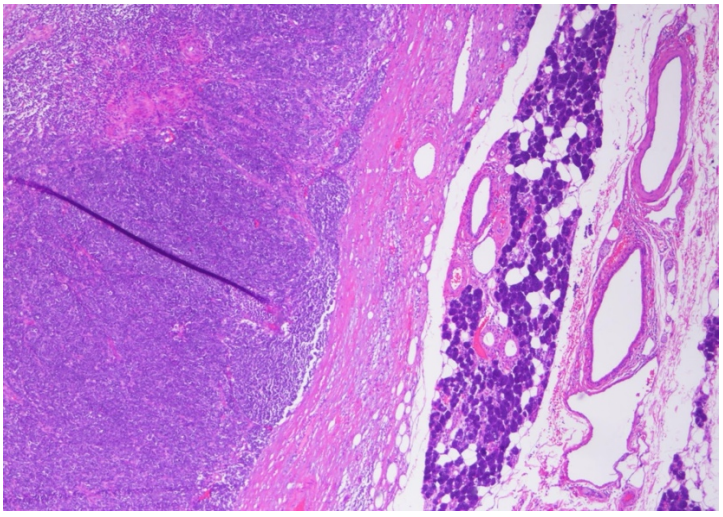

Figure 3: H&E stain showing encapsulated well-differentiated neuroendocrine tumor and surrounding salivary gland tissue.

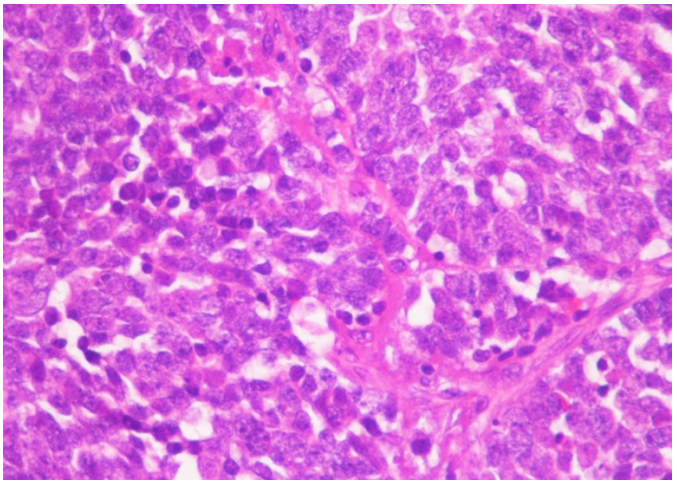

Figure 4: Tightly packed small, round to oval cells with minimal cytoplasm, exhibiting a high nuclear-cytoplasmic ratio.

Discussion

Neuroendocrine tumors typically arise from distal anatomical segments, composed of hormone-secreting cells located in mucosal membranes (1,2). Subtypes include carcinoid, atypical carcinoid, large cell neuroendocrine, and small cell carcinoma (SCC). Head and neck NETs are classified based on tumor proliferation grades. Small cell carcinomas were initially identified in the lung, with the first head and neck case reported in 1972 in the larynx (5,6). Although rare, salivary gland SCCs have been identified. Most salivary neuroendocrine tumors are SCCs, with some cases of large cell neuroendocrine tumors. Differential diagnosis for salivary gland malignancies should consider adenoid cystic carcinomas, basal adenomas, and metastatic lesions. A history of cigarette smoking raises concern for head and neck squamous cell carcinoma and pulmonary metastasis to salivary glands (7,8). Local recurrence and hematogenous metastasis should be considered in patient follow-up. Primary treatment for parotid SCC involves surgical resection with ipsilateral neck dissection, and sometimes radiation therapy (10, 11, 12).

Literature on salivary gland neuroendocrine malignancies is scarce. When diagnosing head and neck masses, consider neuroendocrine tumors of salivary gland origin. Metastatic workup and thorough follow-up are essential. This is the first case of this type at our institution, the primary oncologic referral center in Puerto Rico. Our patient tolerated treatments well and showed no recurrence. Neuroendocrine tumors can be asymptomatic, but their hematogenous spread potential means they are not benign. We recommend tumor resection and radiotherapy, with comprehensive clinical history and metastatic workup, as past literature suggests, when diagnosing parotid masses with primary SCC.

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