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Neuroendocrine Parotid Mass: Atypical Salivary Gland Case

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1. Abstract

Neuroendocrine tumors (NETs) are very rare and commonly found in the gastrointestinal tract and pulmonary system. Neuroendocrine small cell carcinoma (SCC), a NET variant, is often found in the nasal cavity; whereas primary neuroendocrine SCC of the parotid gland is extremely rare, accounting for a minimal percent of all tumors of the salivary gland. We present the case of a 79-year-old Hispanic male who presents with an atypical parotid neuroendocrine tumor, as confirmed by the final pathology report. To the best of our knowledge, no data is available regarding neuroendocrine parotid tumors in the Latin-American population and no case has been reported. We report the first case on this population, emphasizing on how to make the diagnosis and the recommended management.

2. Teaching Points

1)Treatment includes surgical excision of tumor with postoperative radiation and chemotherapy

2)Differentiating NETs tumors from the much more common squamous cell carcinomas and from metastasis from another primary tumor due to the completely different therapeutic approaches and prognosis

3. Introduction

Carcinomas of the major salivary glands are rare tumors that comprise only about 11% of all oropharyngeal neoplasms in the United States [1]. These represent a morphologically diverse group of tumors whose causes and risk factors have been scarcely studied and are not well understood. However, the parotid gland is the most commonly affected, followed by the submandibular glands. The most common etiologies include: mucoepidermoid carcinoma, squamous cell carcinoma, adenoid cystic carcinoma and adenocarcinoma. Nonetheless, lymphoepithelial carcinoma and neuroendocrine tumors are the least common salivary gland tumors; neuroendocrine tumors including both, small cell carcinomas and large cell carcinomas.

The grading criteria for salivary primary neuroendocrine tumors is not well established, although the pulmonary World Health Classification and terminology are generally used to classify extra pulmonary 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 [3]. The occurrence of these tumors in the head and neck region, including the salivary glands is extremely rare. However, most of the other cases reported about such tumors in the salivary gland specifically refer to tumors of the parotid gland [4-6]. The mostly reported cases of neuroendocrine tumors in the salivary glands involve undifferentiated carcinomas, either small cell or large cell carcinomas. Well differentiated, poorly differentiated and moderately differentiated tumors are even more odd and not frequently described in the literature. Small cell carcinoma arising in the parotid gland represents a rare and particularly aggressive malignancy within the spectrum of salivary gland tumors. Immunohistochemical analysis plays an important role in confirming the diagnosis. Positive staining for neuroendocrine markers such as synaptophysin and CD56 is commonly observed, providing evidence of the neuroendocrine differentiation of the tumor cells. Additional

markers, such as chromogranin, and areas of necrosis within the tumor mass are often encountered, which further contribute to the diagnosis. We present the case of a 79 years old male patient who presented with a right preauricular space mass which was diagnosed as a neuroendocrine neoplasm with features of small cell carcinoma. Furthermore we review relevant literature on salivary gland neuroendocrine malignancies, clinical presentation, diagnostic workup, and current management.

4. Case Presentation

Case of a 79-year-old male with a past medical history of hypertension, deep vein thrombosis, and benign prostatic hyperplasia who reported feeling 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 performed which found a well-defined solid mass in the superficial right parotid gland with benign characteristics. The mass measured 1.5cm x 1.2cm x 1.2cm. Patient underwent ultrasound guided fine needle aspiration biopsy. Pathology results reported malignant neoplastic processes with neuroendocrine differentiation. Patient was evaluated at surgical oncology clinics and scheduled for right superficial parotidectomy.

5. Surgical Procedure and Outcomes

The patient was taken to the operating room for a superficial parotidectomy with right neck dissection. Facial nerve was preserved. Specimen was sent to pathology for final diagnosis. The patient was hospitalized for 2 days and the procedure was well tolerated. The patient had no major complications. No neurological deficits. No wound infection and no recurrence of disease on follow up visits. He was sent to an oncologist for further management.

6. Pathology

A gross examination of the mass displayed a right parotid gland specimen in addition to right neck level 2,3,4 excisional specimens. The final pathologic diagnosis confirmed a neuroendocrine neoplasm with small cell carcinoma characteristics showing the unique tightly packed small round cells with minimal cytoplasm and a high nuclear-cytoplasmic ratio (Figure 4). The nuclei showed granular chromatin and prominent nucleoli, contributing to the classic “salt-and-pepper” appearance (Figure 1.C and Figure 1.D). Architecturally, this pathology lacks the typical glandular or ductal structures seen in more common salivary gland neoplasms. Furthermore, mitotic figures were numerous (Figure 1.F), reflecting the rapid and uncontrolled cellular proliferation inherent in small cell carcinomas. As previously stated, regions of necrosis frequently manifest in these tumors (Figure 2 & Figure 1.B), emphasizing the robustly aggressive character of this malignancy. Affirmative immunostaining for neuroendocrine markers including synaptophysin, CD56 (Figure 1.E), and chromogranin were also observed. 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%).

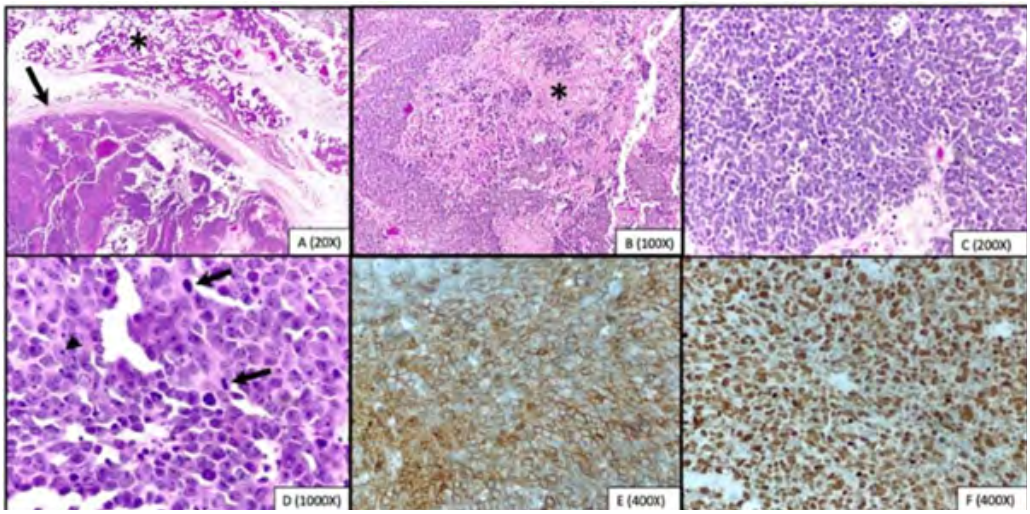

Figure 1: Histopathological and Immunohistochemical Features of a Well-Differentiated Neuroendocrine Tumor of the Salivary Gland.

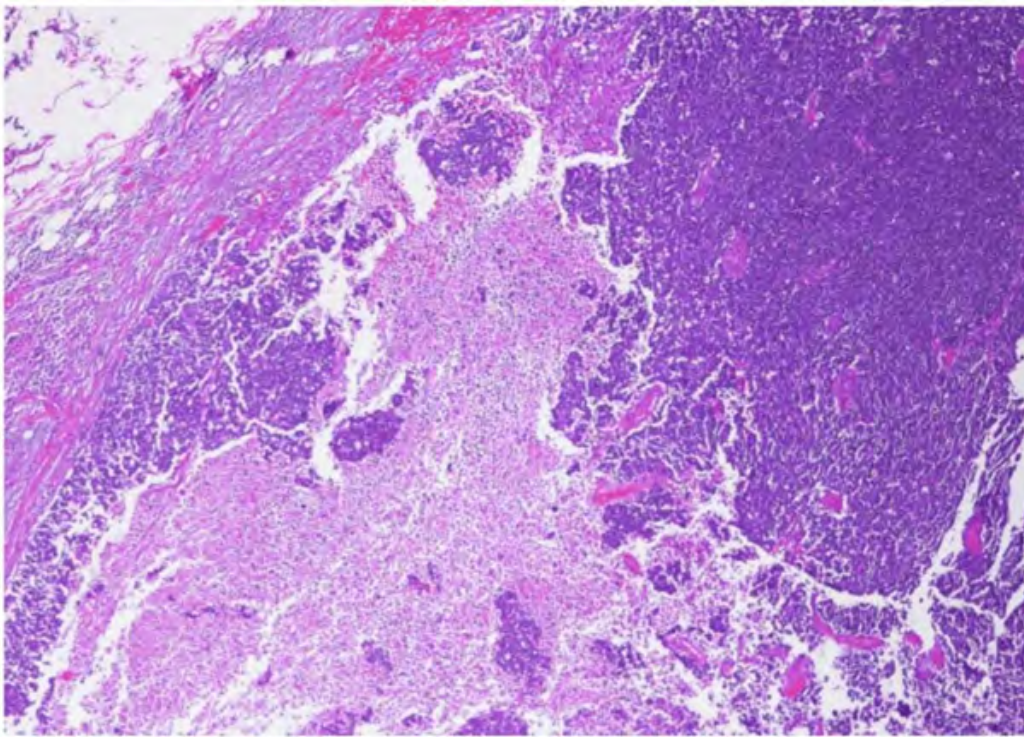

Figure 2: Neuroendocrine neoplasm showing areas with necrosis.

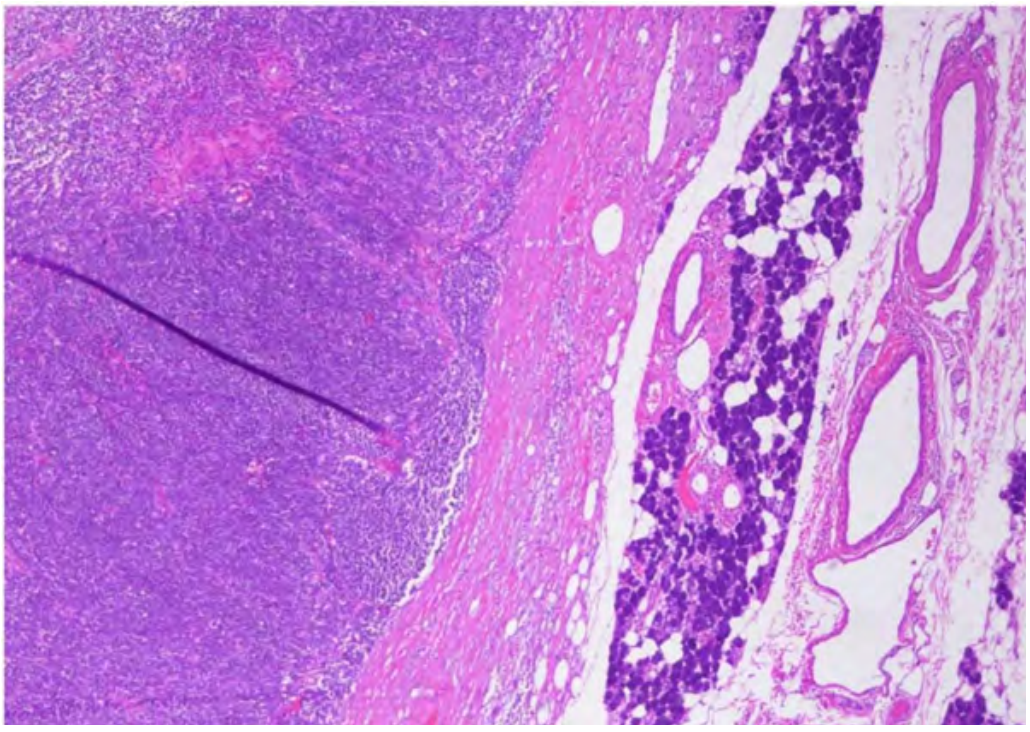

Figure 3: H&E stain showing an encapsulated well-differentiated neuroendocrine tumor and surrounding salivary gland tissue.

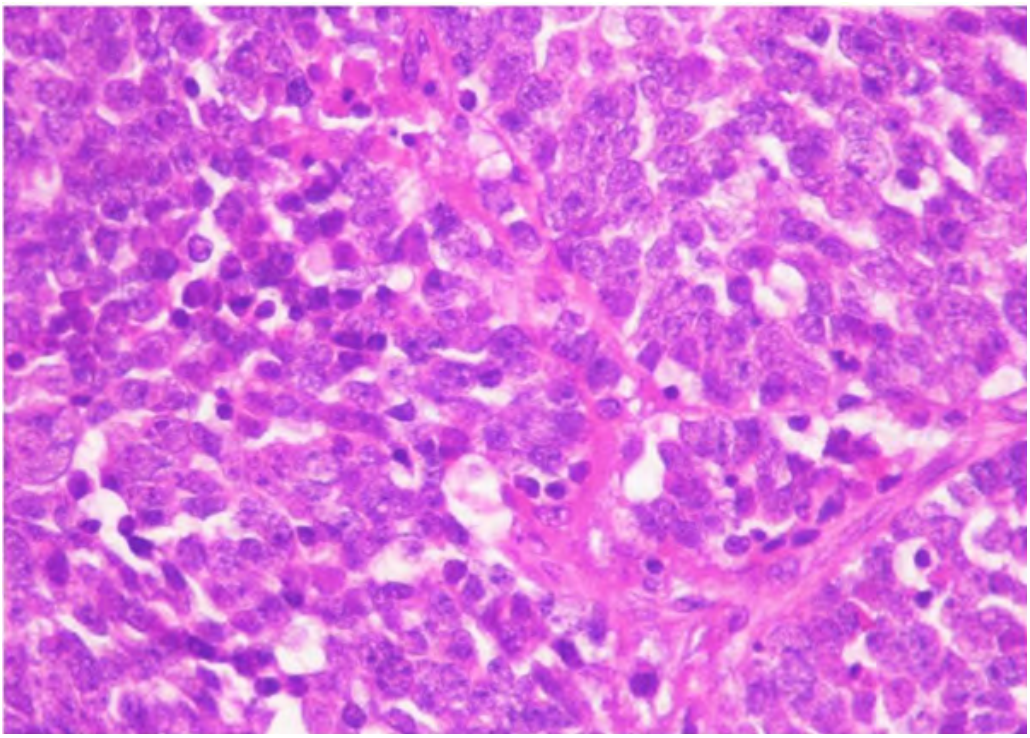

Figure 4: Tightly packed small, round to oval cells with minimal cytoplasm, exhibiting a high nuclear-cytoplasmic ratio.

7. Discussion

Neuroendocrine tumors generally arise from anatomical segments distal to the head and neck region [1]. They are generally composed of hormone secreting cells and located in mucosal membranes. Sub-types include carcinoid, atypical carcinoid, large cell neuroendocrine, and small cell carcinoma (SCC) [3]. Head and neck NET's have been classified based on grades of tumor proliferation.4 Small cell carcinomas were first identified primarily in the lung, and the first head and neck case was identified in 1972 with primary site in the larynx [5,6]. Nonetheless salivary gland SCC's have been identified. The majority of salivary neuroendocrine tumors are SCC, however cases of large cell neuroendocrine tumors have been identified [1,7]. When considering the differential diagnosis for salivary gland malignancies other tumors must be considered such as adenoid cystic carcinomas, basal adenomas, and metastatic lesions. A history of cigarette smoking should be concerning for head and neck squamous cell carcinoma and for pulmonary metastasis to salivary glands [8]. Local recurrence and hematogenous metastasis are to be considered upon working up and following up these patients. Parotid small cell carcinomas are primarily treated with surgical resection, which includes ipsilateral neck dissection, and in instances with radiation. A case of parotid SCC managed with radiotherapy has also been reported.

Literature regarding salivary gland neuroendocrine malignancies is scant. When considering a differential diagnosis for head and neck masses, neuroendocrine tumors of salivary gland origin must be considered. Metastatic workup must be included and thorough follow up is important in these cases. This is the first case of such type seen at our institution, which is the primary oncologic referral center in Puerto Rico. Our patient tolerated his treatments and until and has not displayed any recurrences. Neuroendocrine tumors can be predominantly asymptomatic, such as this one, but their hematogenous spread potential means they are not innocuous. We recommend tumor resection and radiotherapy as management, with thorough workup for clinical history and metastatic workup, as past literature has shown, when considering the differential diagnosis for parotid masses and a positive primary SCC of the parotid gland.

8. Conclusions

A rare case of a 79-year-old Hispanic man who had a neuroendocrine small cell carcinoma (SCC) in the parotid gland. Recognizing its pathological pattern and clinical characteristics guides diagnosis. This is the first case reported in a Latin-American patient, emphasizing the importance of differentiating these rare neoplasms from more common malignancies like squamous cell carcinomas. This report stresses the importance of making this a differential diagnosis from the beginning because they require distinct therapeutic approaches and have different prognostic outcomes. Surgical excision is critical, followed by radiation and chemotherapy in managing these aggressive tumors.

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