Case Report

Sclerosing Mucoepidermoid Carcinoma with Eosinophilia of the Parotid Gland: A Case Report and Review of the Literature

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Sclerosing mucoepidermoid carcinoma with eosinophilia (SMECE) is an extremely rare malignant tumor in the salivary gland. In this case, the patient is a 41-year-old male with no significant history who presented a slowly growing right neck mass. The ultrasound revealed a 3.7 cm complex cystic mass in the right parotid. The surgery was pursued after a non-diagnostic fine needle aspiration. Grossly, the tumor was a 3 cm tan nodule with minimal attached soft tissue. Microscopically, the tumor is relatively well demarcated with anastomosing lobules of neoplastic cells with fibrotic stroma rich in lymphoplasmacytosis and eosinophils. Focal glandular differentiation was highlighted by mucicarmine stain. Immunohistochemically, the tumor cells are diffuse positive for pancytokeratin and p40, while negative for S100, CD45, CD3 and CD20. The tumor is negative for MAML2, EWSR1, and FUS rearrangements. The diagnosis of this case is challenging because the tumor is obscured by the marked fibroinflammatory background. Our findings indicated an exceptionally rare malignant tumor in the parotid that may be clinically and pathologically misdiagnosed as other conditions.

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Key Words: *sclerosing mucoepidermoid carcinoma with eosinophilia (SMECE), parotid gland carcinoma*

INTRODUCTION

Sclerosing mucoepidermoid carcinoma with eosinophilia (SMECE) is an exceptionally rare malignant tumor in the salivary gland.[1,2] It is defined by mucoepidermoid differentiation with a sclerotic stroma rich in lymphocytes and eosinophils.[3] It is considered a variant mucoepidermoid carcinoma (MEC) of the salivary gland. However, most SMECE cases lack MAML2 translocation in these tumors.[4,5] which are distinct from classical mucoepidermoid carcinomas (MEC) of the salivary glands.[6] SMECE has not been recognized by WHO classifications of head and neck tumors as of 2023.[7]

To date, only 28 cases of SMECE have been reported in the salivary gland.[3,4,8] The clinicopathological features of the tumor remain unclear. Here, we report a case of a 41-year-old man with SMECE in his parotid gland and present a review of the literature.

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CASE PRESENTATION

Clinical History

The patient is a 41-year-old male with a slowly growing right parotid mass for one year with some tenderness or pulling sensation. There is no associated discharge or foul taste in the mouth. There is no history of recent infections or trauma in the area. No history of smoking. The ultrasound revealed a 3.7 cm complex cystic mass in the right parotid. Fine needle aspiration was performed, and it was non-diagnostic. Therefore, surgery was recommended.

Pathological Findings

Macroscopically, the resected tumor of the right parotid was a 3 cm intact, tan-gray, rubbery nodule with minimal attached soft tissue. The cut surface showed solid, pale-tan, rubbery, nodular appearance with minimal focal peripheral cystic areas containing brown, gelatinous material.

Microscopic examination reveals a relatively well circumscribed lesion with mainly solid and focally cystic components (**Figure 1A**). The solid components consisted of

lobules, cords and trabeculae of squamoid cells (**Figure 1B**) with focal keratinization (**Figure 1B** inset). The neoplastic cells are embedded in the marked sclerotic stroma (**Figure 1C**). Prominent chronic inflammation rich in lymphoplasmacytic infiltrate, lymphoid follicle, and abundant eosinophils are also present in the background (**Figure 1D**). Cystic changes and primitive glandular differentiation with positive mucicarmine stain are focally identified (**Figures 1E** and **1F**). Mitosis is about 1-2/10 High power field. No necrosis is appreciated.

Immunohistochemically, the tumor cells are diffusely positive for pancytokeratin (**Figure 2A**) and p40 (**Figure 2B**), supporting the squamous differentiation. The tumor cells are negative for S100, CD45, CD3 and CD20. CD45, CD20 and CD3 showed polymorphic inflammatory background. S100 highlighted the dendritic cells. EBER is negative. BRAF VE1 immunostain is negative. IgG highlighted a subset plasma cells, but IgG4 is essentially negative rendering the IgG4: IgG ratio close to 0. Fluorescence in situ hybridization (FISH) studies were performed. The tumor is negative for MAML2, EWSR1, and FUS rearrangements.

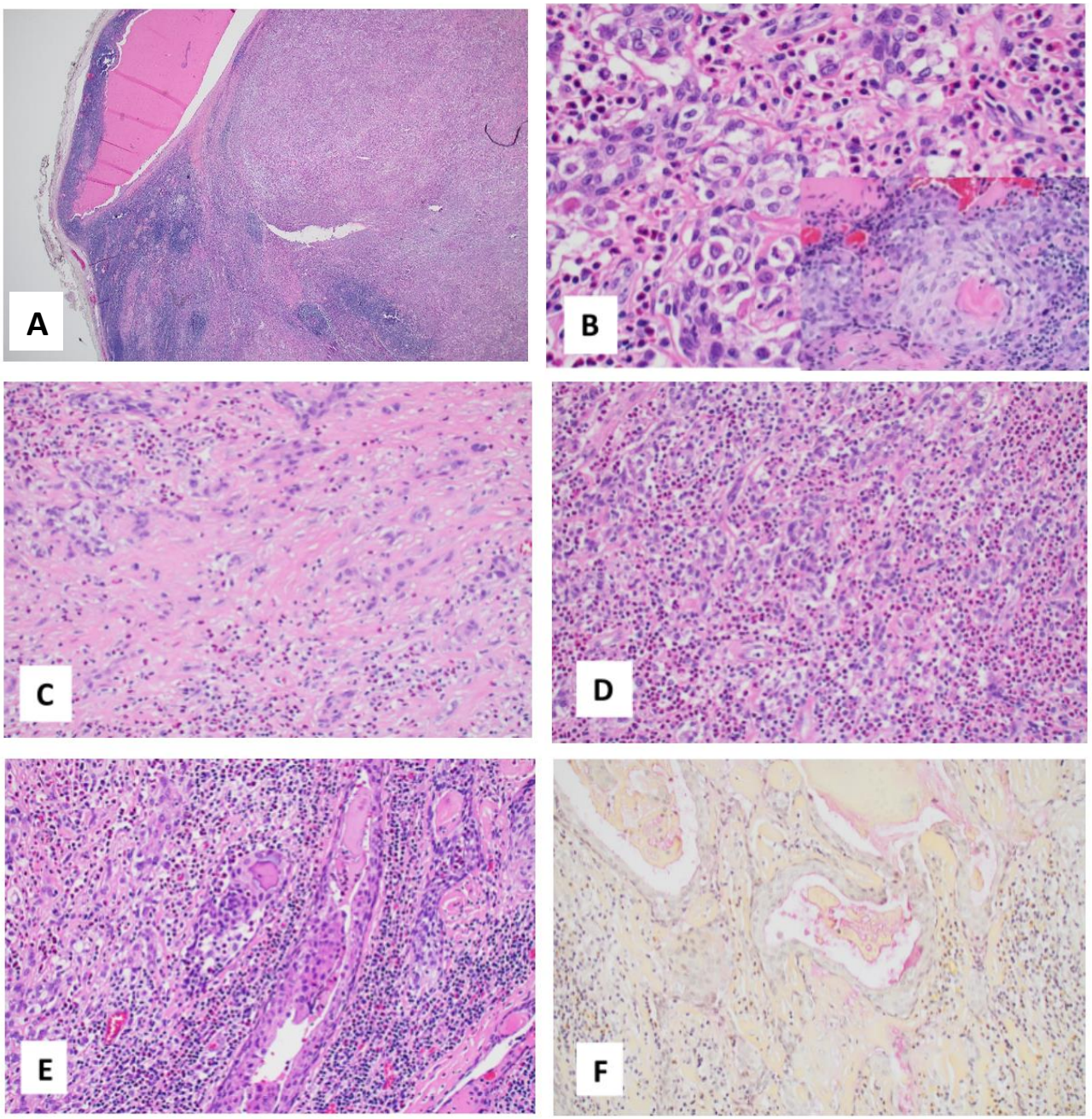

Figure 1. Histological and immunohistochemical features of the tumor. **A.** The tumor is a relatively well circumscribed lesion with rich lymphocytic infiltrate and germinal center. **B.** The squamoid neoplastic cells form anastomosing lobules, cords, and trabeculae with focal keratinization (inset). **C.** There is keloid-like fibrotic background. **D.** Tumor associated eosinophilia. **E.** Focal columnar mucinous cell. **F.** Focal primitive glandular differentiation is identified by a mucicarmine special stain.

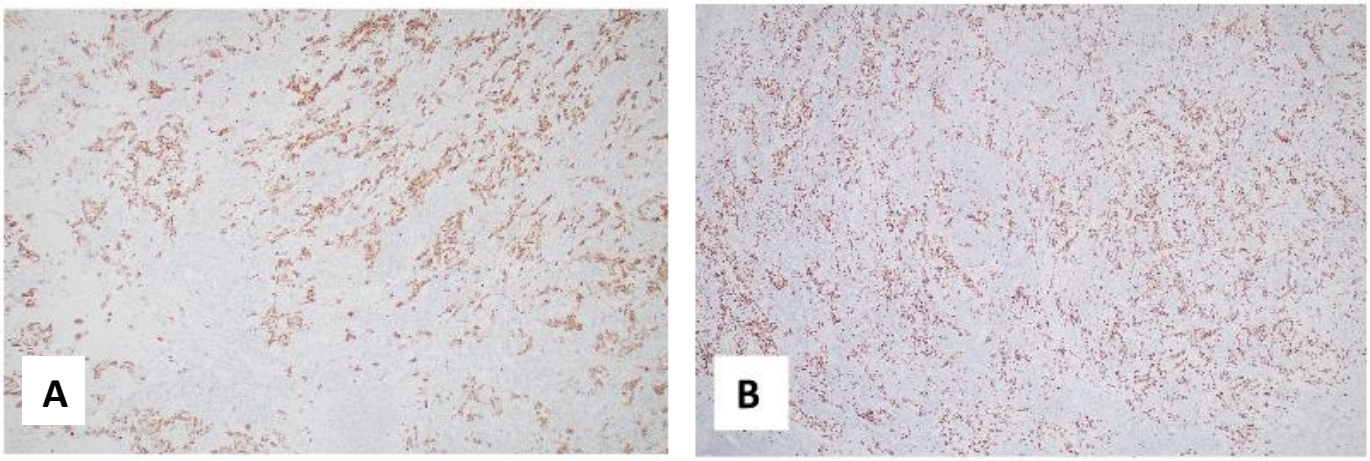

Figure 2. Squamous differentiation of the tumor. The tumor cells are diffusely and strongly positive for pancytokeratin (**A**) and p40 (**B**).

DISCUSSION

SMECE is an exceptionally rare tumor in the salivary gland. The keloid-like sclerotic background, dense chronic inflammatory infiltrate, abundant eosinophils and dendritic cells, prompt other considerations including non-neoplastic and neoplastic processes. Differential diagnosis includes chronic sialadenitis, IgG4 related disease, lymphoma, lymphoepithelial carcinoma and dendritic cell histiocytosis. SMECE is most likely to be low grade malignancy.[3] However, some tumors have intermediate to high grade malignancy.[4]

Chan[9] described a same-named tumor in the thyroid and the thyroid SMECE lacks common thyroid cancer mutations as well as MAML2 translocation according to studies by Shah et al.[10] SMECE of the salivary gland is extremely rare; only 28 cases have been reported in the literature and this entity was not listed in the 2022 WHO Blue Book. Morphologically, the combination of squamoid and glandular components argues that it is a sclerotic variant of classic MEC. The lack of the common MAML2 translocation seen in classic MEC argue against grouping SMECE under MEC. Most recently, CSF1 gene derangement was reported in SMECE of the parotid gland.[4] A lack of well-documented molecular marker except negative for MAML2 translocation makes categorizing SMECE as a distinct entity difficult.

CONFLICTS OF INTEREST
None

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