Case Report

Epithelial-myoepithelial carcinoma of floor of mouth: A case report with cytological, histological and immunohistochemical correlation

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ABSTRACT

A 61-year-old female presented with a 3-year-old swelling in the right floor of mouth. Clinical examination and fine needle aspiration cytology suggested a benign lesion. The mass was excised locally along with the involved sublingual and deep part of submandibular gland and duct. Post-operative histopathological examination revealed features of pleomorphic adenoma. However, on revision of histological sections, features were predominantly of a rare malignancy of the salivary glands, epithelial-myoepithelial carcinoma (EMC), along with focal areas of adenoid cystic carcinoma (Ad CC). The tumor was p-63, s-100 and smooth muscle actin positive but C-kit was negative, which ruled out Ad CC and the possibility of a hybrid carcinoma. The aim of this article is to describe a rare case of EMC in the floor of mouth and the confusing cytological picture that it created.

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Introduction

Epithelial-myoepithelial carcinoma (EMC) is a rare biphasic low grade malignancy accounting for only 0.5% of all salivary gland tumors.[1] There have been reports suggesting co-existence of another malignancy, like adenoid cystic carcinoma (Ad CC) with EMC, in the same tumor. Such tumors are named "hybrid carcinomas."[2] If EMC develops in a long standing case of pleomorphic adenoma (PA), it is called EMC-ex PA.[3] Ours is a case of a tumor in the submandibular and sublingual salivary glands diagnosed as EMC, having clinical and histological resemblance to cellular PA.

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Case Report

The present case report is about a 61-year-old woman who reported to us with a growth in the floor of mouth that gradually increased in size since last 3 years, without pain. On examination, a 3 cm × 2.5 cm ovoid swelling was palpated lingual to the edentulous alveolar ridge, extending from midline (lingual frenum) to the right premolars [Figure 1]. It was a firm, non-tender, non-compressible, non-reducible, did not move with deglutition and showed no change in size with meals. No pulsations were felt over the mass and no vascular channels seen. The swelling was not fixed to underlying tissues and salivary secretions were normal. Some cervical lymph nodes were palpable but not tender. There was no neural involvement or sudden increase in size. Complete hemogram, serum electrolyte levels, kidney function test and liver function test had all values within the normal range, but for a raised erythrocyte sedimentation rate (50 mm/h). Fine needle aspiration cytology (FNAC) of the lesion suggested PA. Node Ib was palpable, but non-reactive on FNAC. Magnetic

resonance imaging revealed a 20 mm × 18 mm × 15 mm well defined well-marginated lesion in right sublingual space in the region of anterior aspect of sublingual gland [Figure 2]. It indented the inferior surface of tongue and right genioglossus muscle and did not cross midline. Multiple mildly enlarged bilateral level Ib, II and III lymph nodes were noted. A provisional diagnosis of PA of sublingual gland was made and local excision of the lesion along with the gland was planned under L.A. Incision was placed from midline extending posteriorly near the alveolar ridge in order to protect the Wharton's duct. With careful dissection, lingual nerve was identified and protected. On surgical exploration, it was found that the mass involved the deep part of submandibular gland and duct with the sublingual gland being compressed by the mass. It was difficult to delineate the exact origin of the pathology (sublingual or submandibular gland); hence it was decided to sacrifice the sublingual gland, deep part of submandibular gland and its duct on the affected side [Figure 1]. Excision was done with a cuff of healthy overlying mucosa. Neck dissection was not indicated, hence not done. Initial histopathological examination, revealed a solid tumor tissue composed of myoepithelial and ductal cells presenting as double layered duct like structures with hyalinization of stroma. The epithelial component showed diversity in the form of sheets, nests and cords with different patterns of ductal arrangements and large areas of secretory material in the intervening areas, suggestive of cellular PA [Figure 3]. However since most of the tumors of submandibular and sublingual gland are malignant, a second examination of the specimen was undertaken. It showed that the entire tumor mass was covered by dense band of fibrous connective tissue. Tumor cells were arranged in a background of predominantly mucous acini, as small tubules lined by cuboidal to oval cells containing prominent nuclei and eosinophilic cytoplasm and surrounded by clear cells and increased hyalinization [Figure 4]. On the basis of this dual cell population, a provisional diagnosis of EMC was suggested. Some focal areas were also observed with cells arranged in a cribriform pattern with pseudocystic spaces that pointed toward Ad CC. Immunohistochemistry showed p-63 reactivity in outer clear cells, S-100 and smooth muscle actin were focally positive and C-kit for Ad CC was negative. Hence, a diagnosis of hybrid carcinoma was ruled out and a final diagnosis of epi-myoepithelial carcinoma was made. There is a chance that being a long-standing tumor; initial PA may have turned into EMC. Patient denied adjuvant Radiotherapy; is being followed-up since last 1 year and has no fresh complains or signs of recurrence until now [Figure 5].

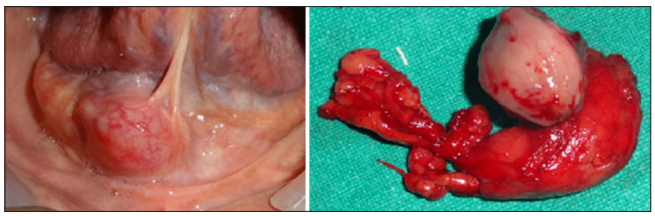

Figure 1: Clinical picture

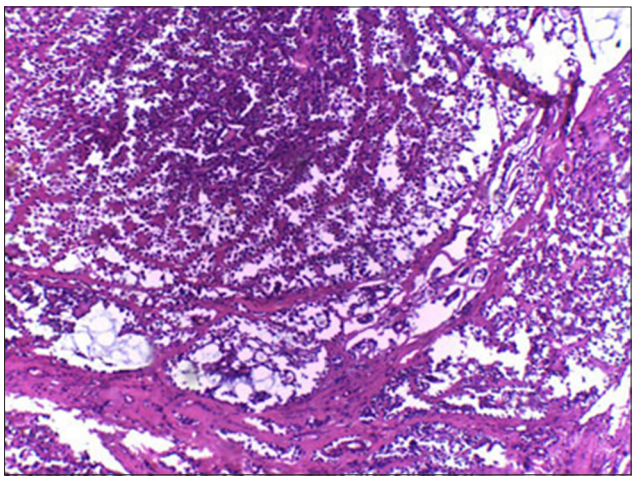

Figure 3: H and E section suggestive of cellular pleomorphic adenoma

Discussion

EMC was first described by Donath *et al*., in 1972 and officially defined as a salivary gland tumor by WHO in 1991. It commonly affects parotid gland (70%), sino-nasal glands, palatal and submandibular gland and rarer sites like lungs, breasts etc., (no reports on sublingual

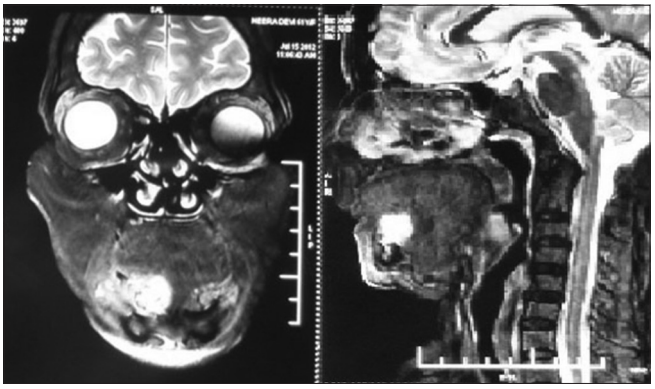

Figure 2: Magnetic resonance imaging of the lesion

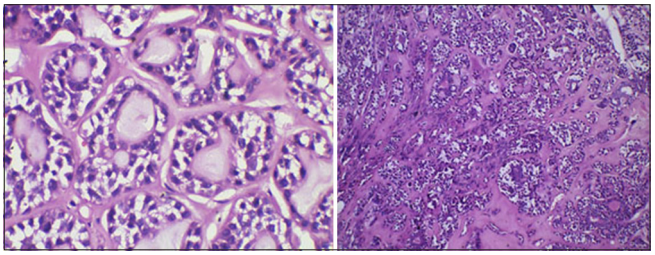

Figure 4: H and E section suggestive of epithelial-myoepithelial carcinoma

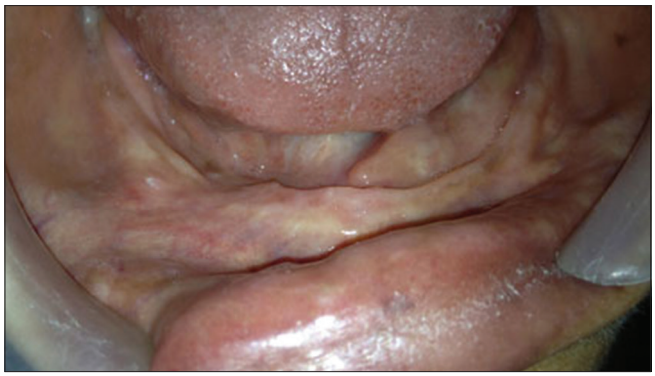

Figure 5: Post-operative clinical picture

gland origin were found). It is a low-grade carcinoma of ductal origin characterized by its dual cell population of ductal epithelial cells and clear myoepithelial cells. Apart from this characteristic picture, there are many histological variants such as oncocytic EMC, double clear EMC, EMC ex PA, high grade or dedifferentiated EMC, EMC with myoepithelial anaplasia etc.[1,4] The median age of occurrence is 60.4 years.[1,4] Many times, it is found that pre-operative FNAC points toward a benign pathology (PA).[5,6] A diagnosis of PA would be incorrect due to the excessive hyalinization seen in the tissue, in spite of the pleomorphic picture of cells. Differential diagnosis includes other clear cell tumors such as muco-epidermoid carcinoma, acinic cell carcinoma and sebaceous carcinoma, with immunohistochemical presence of myoepithelial markers, similar to those seen in EMC. However they lack the biphasic pattern. Furthermore, there have been reports describing EMC ex PA in parotid gland with the histological picture similar to ours.[2] Within the characteristic dual cell picture of EMC, arrangements of cells in cribriform pattern with pseudocystic spaces reminiscent of Ad CC were observed focally. Woo *et al*., have suggested such an overlap in previous literature. In the presenting case, the tumor could have been a primary onset EMC or an EMC-ex-PA. There have been cases of recurrence of tumor by simple excision alone,[1,3,7] hence we chose wider margins of healthy mucosa. The prognosis of EMC is fairly good as the median disease free survival rate is reported to be 11.34 years.[1] Long-term follow-up is needed for detection of early signs of recurrence and proper management.

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