Mucoepidermoid Carcinoma in a 33-Year-Old White Man

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ABSTRACT

Patient Demographics: A 33-year-old white man.

Chief Complaint: Longstanding bump on the roof of the mouth.

Treatment: The patient was referred to an oral maxillofacial surgeon for a pathology consultation. The patient had been experiencing pain on both sides of his jaw, but the oral mass was not painful (Image 1).

Medical History: The patient has no history of smoking, is slightly overweight with a body mass index of 26.5, and has no systemic or chronic medical conditions.

Examination Findings: On initial examination by the oral surgeon, the vital signs of the patient were normal. An indurated lesion was noted on the left hard palate of the patient. Suspecting a benign lesion or possible carcinoma/neoplasm, the oral surgeon scheduled and performed an excisional biopsy (Image 2). The mass was sent for pathological processing.

Postoperative Care: This included a diet of soft foods, with daily rinsing using 0.12% chlorhexidine gluconate, with a follow-up appointment made for 2 weeks later.

Principal Laboratory Findings: Figure 1; Image 3; Image 4

Keywords: mucoepidermoid carcinoma, MECT1-MAML2, salivary-gland neoplasm

Questions

1. What clinical and laboratory findings are most striking?
2. What is the differential and most likely diagnosis in this patient?
3. What are the major causes of this disease?
4. What tests are used to confirm this diagnosis?
5. What is the frequency of this disease in the general population?
6. What are the treatment options and management possibilities for this disease?

Possible Answers

1. When considering a diagnosis for the patient, noted clinical features included his age, sex, and medical history, particularly his longstanding abstinence from tobacco use. Location of the mass is key, while noting its presentation of induration and the lack of pain experienced by the patient. Important laboratory findings include the presence of mucous, epithelial, and epidermoid cells, as well as positive results via mucicarmine staining (Image 5).

2. Based on the clinical history and presentation of this specific patient, the differential diagnoses are listed in Figure 2. The most likely diagnosis in this patient is salivary gland cancer—specifically, mucoepidermoid
carcinoma (MEC). MEC treatment and prognosis are based on the TNM stage and grade of the cancer. Stage is determined based on the characteristics of the primary tumor (location, size, depth of growth at its origin, and growth into nearby tissue), its spread to nearby lymph nodes, and whether the cancer has metastasized.

Grading of MEC is determined microscopically by measuring the appearance of abnormal cancer cells. MECs can be found at different grades, ranging from low to high grade, with the latter having a worse prognosis. At each grade of MEC, this cancer can mimic other diseases and needs to be differentiated from other similar malignant conditions. For low-grade MECs, one must rule out Warthin tumors, benign salivary gland cysts, branchial-cleft cysts, sialolithiasis, and pleomorphic adenoma with excess mucoid stroma. High-grade MECs should be differentiated from squamous-cell carcinoma, salivary-duct carcinoma, and adenocarcinoma.

MECs were first identified in 1945 and can involve 3 types of cells: squamous, mucus secreting, and “intermediate” cells. MEC can develop with no signs or symptoms. The first indication may be determined after oral cancer screening by a dentist or during a physical examination by a physician. The tumor that develops is usually painless and grows slowly. Some tumors, however, grow quickly. The general symptoms of MECs include site tenderness, difficulty swallowing, ear discharge, and oral spasms. Tumors are generally less than 4 centimeters in diameter.

Although the exact cause of MEC is unknown, it is thought that major contributors for its development are age, ionizing radiation exposure, and possible exposure to certain chemicals. Other study results have reported a possible relationship with infection of cytomegalovirus (CMV) and human papillomavirus (HPV). However, a recent study provided no link between HPV and MEC with or without the MAML2 rearrangement. Although the presence of CMV and HPV has been shown in patients with MEC, there is no known indication of causation at this point.

The tests used to diagnose MEC include a physical examination with patient history, magnetic resonance imaging (MRI), computed tomography (CT) scan, positron emission tomography (PET) scan, endoscopy, and/or biopsy. The type of biopsy can include fine needle aspiration (FNA), excisional biopsy, or biopsy via surgical
The standard for initial diagnosis is through preoperative FNA. The FNA will aid in obtaining a specific diagnosis or will significantly reduce the possibilities in the differential diagnosis, which ultimately aids in the surgical approach. During pathological processing, mucicarmine staining by a pathologist can be used to help differentiate MEC from squamous-cell carcinoma. At time of diagnosis, it is also important to determine the cancer stage and whether the cancer has spread to other locations in the body. Determining the tumor grade is important because the overall likelihood of successful treatment and survival correlates with the severity of the neoplasm. Table 1 shows how the different MEC grades are determined and outlines the overall 5-year survival rate for each grade.²,³

### Table 1. Mucoepidermal Carcinoma (MEC) Grades

<table>
<thead>
<tr>
<th>Grade</th>
<th>Cellular Composition</th>
<th>Cell Differentiation</th>
<th>5-Year Survival Rate</th>
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<tbody>
<tr>
<td>Low</td>
<td>Mucus secreting and intermediate cells, possible goblet cells</td>
<td>Well-differentiated</td>
<td>92%-100%</td>
</tr>
<tr>
<td>Intermediate</td>
<td>Mixture</td>
<td>Intermediate</td>
<td>62%-92%</td>
</tr>
<tr>
<td>High</td>
<td>Squamous epithelial and intermediate cells</td>
<td>Poor differentiation</td>
<td>0-43%</td>
</tr>
</tbody>
</table>

It has been shown that patients with MEC possess a MECT1-MAML2 gene rearrangement [t(11;19)(q14-21;p12-13)]. This translocation has been shown² to occur in all grades of MECs and only in MECs. This translocation is identified using fluorescent in situ hybridization (FISH) or using reverse-transcription polymerase chain reaction (RT-PCR). This gene rearrangement is believed to disrupt the Notch signaling pathway, which is important in the normal development of cells and tissues. This disruption likely contributes to the lack of control and development of a neoplasm.²,³

5. MEC is the most common type of malignant neoplasm of the salivary glands in adults, with most cases occurring in the parotid gland. MEC is more prevalent in women than men; the mean onset is generally
approximately age 45 years. Although this disease is more common in middle-aged patients, MEC and acinic cell carcinoma are not uncommon in children and must be included in the differential diagnosis of salivary gland masses in children. One study group examined the frequency of MEC in its hospital facility and discovered that of 151 malignant salivary-gland tumors, 75 were MEC (49.6%). The average age of the patients with MEC was 42.6 years (range, 6 years to 67 years), with a distribution of 36 males and 39 females. Of the 75 cases, 29 were high grade, 10 were intermediate, and 36 were low grade.

6. Appropriate treatment and management depends on tumor grade, stage, clinical presentation, and location. The options for treatment include surgical procedures, radiation (fast neutron radiation or photon beam radiation therapy), and/or chemotherapy.

Patient Follow Up

After the initial excisional biopsy, the patient returned to the oral surgeon 2 weeks later, stating that he had felt pain in the biopsy area for a week, but that the pain had subsided. On examination, the biopsy area showed signs of a recurring lesion, so a second biopsy was performed. No recurrence of malignant neoplasms was found on the second biopsy. A few months later, the site is slowly healing, with no signs of recurrence at this time (Image 6).

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References


