EMERGENCY ROUTE DIAGNOSIS

CASE HISTORY REPORT

ABSTRACT
Salivary gland malignancy (SGM) can affect both major and minor glands and manifests clinically with various presentations. The most common type of SGM is mucoepidermoid carcinoma (MEC), which has been previously reported to be associated with symptomatology associated with temporomandibular disorders (TMD). This case report describes a patient with an aggressive form of MEC of the parotid gland that was initially diagnosed as TMD. In addition, the patient’s MEC was diagnosed emergently based on development of acute clinical symptomatology. To the best of our knowledge, emergency route diagnosis of MEC affecting the parotid gland has not been previously reported in the literature.

KEY WORDS: emergency route diagnosis, mucoepidermoid carcinoma, temporomandibular disorder

Emergency route diagnosis of mucoepidermoid carcinoma initially diagnosed as a temporomandibular disorder

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Mucoepidermoid carcinoma (MEC) is the most common malignancy of the salivary glands with mean age at diagnosis of 45 years without marked gender differences.1 It most often affects the parotid gland;2 however, MEC may be associated with the submandibular, sublingual, or minor salivary glands. Clinically, MEC typically presents as a nonpainful swelling or mass, however, pain and facial nerve palsy may develop depending on the anatomical location of the tumor.3 All MECs have metastatic potential and the lungs, bones, and liver have all been reported as sites of metastasis.4

MEC is composed of squamoid cells, mucus-producing cells, and intermediate cells which can have a varied histological presentation.5 Grading of MEC tumors is dependent on a variety of characteristics including necrosis, mitosis, atypical nuclei, and size of cystic component of the tumor.6 Histology and grading of the neoplasm are important in predicting the rate of growth and metastatic potential of the disease.6 Low-grade tumors tend to have a higher mucinous component and have been observed to behave in a more benign fashion, while high-grade tumors consist of poorly differentiated cells and tend to be aggressive with the potential for metastasis.3

The route to diagnosis is an important factor in determining patient outcomes.7 Emergency route diagnosis of cancer may cause increased anxiety for the patient due to development of acute symptoms and increase demands on the health-care system by requiring urgent management and the need for out-of-hours personnel.7,8 Biological factors, such as later stage of malignancy at diagnosis and advanced age, contribute to poorer survival of Emergency Department (ED)-presenting patients.9 Delayed definitive diagnosis due to complex symptom presentation can also increase the chances of ED presentation.10

Salivary gland tumors, including MEC, have been previously reported to present with any combination of complex symptoms of facial pain, trismus, otalgia, and tinnitus mimicking temporomandibular disorders (TMD).11,12 However, it is essential to note that salivary gland tumors with symptoms...
mimicking TMD may also present with additional signs and symptoms of swelling, numbness, or nerve paralysis which should increase suspicion for a malignant process. Patients who do not respond to initial therapy or whose symptoms progress disproportionately to the typical course of condition should be further evaluated to rule out other pathologies.

This report presents a case of MEC, initially diagnosed as a TMD that was subsequently diagnosed through referral to an ED as a result of acute symptom onset. To the best of our knowledge, emergency route diagnosis of MEC affecting the parotid gland has not been previously reported in the literature.

**Case report**

A fifty-six-year-old Caucasian female was referred to the Oral Medicine service at the University of Pennsylvania Health System for evaluation of intermittent left facial swelling and pain of at least 5 months’ duration. She described persistent ache with occasional sharp pain of the left temporomandibular joint (TMJ) that was not affected by jaw function. The patient denied previous trauma to her face, jaw, and TMJ. Additionally, there was no history of parafunctional habits reported such as bruxism or clenching of the teeth. She was previously evaluated by an otolaryngologist and diagnosed with TMD. The patient was treated with a custom occlusal appliance fabricated by her dentist without benefit. Her past medical history was noncontributory and current medications included clindamycin 150 mg and oxycodone–acetaminophen 7.5–325 mg for her current condition. The patient reported no known drug or food allergies, and her family history was positive for cancer (mother—ovarian cancer, father—neck tumor of unknown origin). Her social history was negative for tobacco, alcohol, and recreational or intravenous drug use. Review of systems was positive for left ear tinnitus and numbness of the left preauricular area. Detailed cranial nerve exam II–XII was grossly intact. Extraoral exam did not reveal thyromegaly; however, generalized left facial swelling in the parotid gland region (Figure 1) and a firm 1.5-cm left submandibular lymph node was detected. TMJ examination revealed maximal inter-incisal opening of 35 mm, nontender, bilateral crepitus of the TMJs, and nontender muscles of mastication bilaterally on palpation. Intraoral exam revealed free-flowing saliva from the parotid glands bilaterally when milked. Masses, lesions, or ulcers of the oral mucosa and gingiva were not detected and dentition appeared stable without gross caries. Differential diagnosis consisted of sialadenitis versus salivary gland neoplasm of the left parotid gland. With the exception of nontender, bilateral crepitus of the TMJs, the patient’s symptoms did not correlate with a diagnosis of TMD. The patient was prescribed cephalexin 500 mg three times daily for 10 days, etodolac 400 mg every 6 hours as needed for pain management, and a magnetic resonance image (MRI) with and without intravenous contrast was ordered to rule out a neoplasm of the left parotid gland. Prior to obtaining the scheduled MRI, the patient developed acute left facial numbness, increased swelling of the left parotid region accompanied by left eye nerve palsy. She was advised to report to her local hospital for emergency evaluation.

The MRI was completed emergently and revealed a 2-cm fluid-filled, necrotic left submandibular lymph node and a 4.1×2.4×4.9 cm enhancing mass of the left parotid gland (Figure 2). The patient was urgently referred to the otolaryngology service and underwent cytologic analysis of the left parotid gland mass via fine-needle aspiration, which was consistent with poorly differentiated MEC. Subsequently, plain film imaging revealed metastases to the mediastinal and hilar lymph nodes, lungs and positron emission tomography (PET-CT) demonstrated metastasis to base of skull (Figure 3).

As a consequence of stage T4aN1M1 parotid gland MEC with symptomatic progressive parenchymal and pleural lung disease, the patient was considered unsuitable for surgical management. Treatment consisted of radiation to decrease the rate of primary tumor growth in addition to palliative care. The patient received a total dose of 5,000 cGy in 250 cGy fractions. Radiation was followed by intermittent chemotherapy; however, the patient experienced rapid disease progression and succumbed to the disease 18 months after MEC diagnosis.

**Discussion**

Acute symptoms prompting emergency route diagnosis of head and neck cancer is uncommon. Variable patterns have been observed in emergency route diagnosis of cancer based on age, gender, income, and type of cancer, which suggests that factors beyond biological qualities of a neoplasm may predict the odds of an emergency route diagnosis. Advanced age and low-income status has been associated with emergency head and neck cancer diagnosis, which contributes to poor survival prognosis. Patients with emergency route diagnosis of cancer often have seen primary care...
providers prior to their ED presentation, however, complex symptom presentation and lengthy diagnostic testing may delay diagnosis and contribute toward patients seeking emergency care. Improving recognition and understanding of the complex clinical presentations of malignancies may help prevent emergency route diagnosis. A thorough medical history, review of systems, physical examination, and appropriate diagnostic testing are critical for accurate and timely diagnosis of neoplasm. Clinicians should recognize common features that increase suspicion of a neoplasm such as cranial nerve deficits, including numbness or palsy, and palpable lymph nodes. The diagnostic work up for neoplasm of the salivary glands may include imaging, such as MRI, CT, and ultrasonography, to determine the location and extent of a tumor. Further tests include fine-needle aspiration cytology, for histologic interpretation, and PET with or without CT to localize metastases.

An increased understanding of prognostic factors may help patients and clinicians choose treatment to maximize both patient outcomes and quality of life. Prognostic factors that have been found to be associated with decreased survival in cases of MEC include higher histologic grade, older patient age, larger tumor size, extraparenchymal extension, presence of positive lymph nodes, and distant metastasis. Advanced techniques can measure levels of membrane-bound mucins, particularly MUC1 and MUC4, whose expression provides insight into disease recurrence and prognosis. Increased expression and distribution of MUC1 across the membrane of MEC tumor cells may promote tumor growth and survival, however exact mechanisms are not yet known. MUC4 expression in MEC tumor cells has been associated with decreased recurrence and increased survival, while the exact mechanism of action are still under investigation some speculate that MUC4 may play a role in MEC tumor cell differentiation.

The National Comprehensive Cancer Network provides guidelines for surgical management of low and high-stage tumors but there are no current...
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guidelines for the surgical management of intermediate-grade disease.\textsuperscript{18} Treatment is frequently limited in cases of emergency diagnosis, due to the late stage at presentation, and often is limited to palliative care.\textsuperscript{14} In the case of MEC treatment, there is no established chemotherapy protocol and attempts to use chemotherapy have had poor results.\textsuperscript{19} This case highlights acute symptom onset of a high-grade parotid gland MEC diagnosed via the emergency route. Due to the poorly differentiated nature of the tumor and advanced stage of disease at the time of diagnosis, the patient’s survival rate was poor. Earlier detection of the MEC might have led to a more favorable prognosis due to decreased tumor size, risk of metastases, and possible additional therapeutic options. In addition, this patient was initially diagnosed with TMD and was treated for this condition without benefit. It is important for clinicians to understand these disease processes and appreciate clinical signs and symptoms of these disorders to ensure appropriate patient evaluation and management in a timely manner for optimal treatment outcomes.

References