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Case Report

Metastatic Adenoid Cystic Carcinoma to the External Auditory Canal: Report of a Case and Brief Review of the Literature

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Abstract

Adenoid Cystic Carcinoma (ACC) is a slow growing neoplasm which often recurs within one to two decades after treatment. There are many subtypes of ACC, including cribriform, tubular, and solid forms, which have been found to correlate with overall prognosis as well as recurrence among patients with history of previously resected or treated ACC. Review of the existing literature shows that it is difficult to detect and control recurrence of ACC as most sites of metastasis do not show clinical signs or indications until the neoplasm has become increasingly advanced. Adenoid cystic carcinoma of the head and neck is usually found in the salivary glands, oral cavity, nasopharynx, and palate. There have been only a few reported cases of adenoid cystic carcinoma involving the External Auditory Canal (EAC). We present a case of ACC of the EAC along with a review and discussion of the literature to date.

Keywords: Adenoid Cystic; Recurrence; Metastasis; External Auditory Canal; Histomorphology

Abbreviations

ACC: Adenoid Cystic Carcinoma; EAC: External Auditory Canal; IHC: Immunohistochemistry

Introduction

Adenoid Cystic Carcinoma (ACC) is a rare malignant tumor of the major and minor salivary glands, accounting for 1% to 2% of all head and neck malignancies and approximately 10% of all salivary gland neoplasms [1]. It is a slow growing but highly invasive cancer with a high recurrence rate [2]. It is not uncommon for distant metastasis, mainly to the lungs, to occur over the course of many years [2]. Accurate and timely diagnosis of ACC can be especially difficult in situations when the tumor appears in an atypical localization, or presents with non-specific features. Specifically, ACC of the EAC is characterized by an indolent clinical course, which often leads to a delayed diagnosis of the condition and therefore delayed management. Optimal treatment of this tumor has not yet been fully established, but the mainstay of current treatment is surgical excision with the goal of achievement of clear surgical margins to reduce the risk of local recurrence [3].

Due to the rarity of the Adenoid Cystic Carcinoma of the EAC, there is limited literature regarding long-term outcomes and follow-up. We report a case of ACC of the parotid gland with metastasis to the EAC. The case illuminates the need for early detection of ACC, common presentations of ACC within the population, as well as the current guidelines and recommendations for detection and treatment of ACC of the External Auditory Canal (EAC).

Case Presentation

A 62-year old female presented with a right external ear scar mass extending into the external auditory canal (EAC) measuring 2.8 cm. A Computed Tomography (CT) exam revealed an oval hypodense, heterogeneous expansive process infiltrating the deep dermal tissue, the cartilage, and extending into the EAC. Radiographic images were suspicious for a malignant process. The mass was excised and histomorphology was diagnostic of adenoid cystic carcinoma (ACC). The diagnosis was made based on histomorphologic features classically seen in ACC and immunohistochemistry studies. The histomorphology was heterogeneous showing varying amounts of three distinct growth patterns, cribriform, tubular, and solid (Figure 1A). The cribriform subtype was the most frequent component displaying islands of basaloid cells surrounding variably sized cyst-like spaces containing mixed basophilic and eosinophilic material some of which positive for PAS. True glandular lumens composed of cuboidal cells showing ductal differentiation were also identified scattered throughout (Figure 1B). Immunohistochemistry studies showed smooth muscle myosin heavy chain, highlighting the cells with myoepithelial differentiation surrounding the pseudocysts. The lumens of the cystic spaces stained positively

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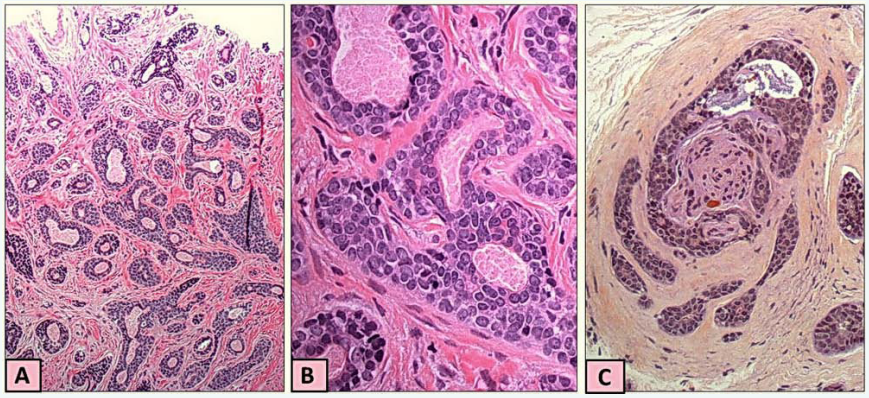

Figure 1 Histomorphology of the right external auditory canal mass

A- Heterogeneous mass showing varying amounts of three distinct growth patterns, cribriform, tubular, and solid (H&E stain X20)

B- True glandular lumens composed of cuboidal cells showing ductal differentiation were also identified scattered throughout (H&E stain X40)

C- Multiple foci of perineural invasion were identified (H&E stain X40).

for PAS. Most of the tumor cells were positive for c-KIT (CD117).

Prominent vascular invasion as well as perineural invasion were also noted (Figure 1C). All surgical margins were clear and negative for carcinoma. Initial diagnosis was Primary cutaneous adenoid cystic carcinoma, however at the time of planning post-operative therapy, it was discovered that the patient had a history of ACC of the parotid gland nine years prior to current presentation. This prior parotid carcinoma was treated by Total Parotidectomy with adjuvant chemotherapy and the patient was free of the tumor until current presentation. During metastatic work up to rule out a second primary adenoid cystic carcinoma versus metastasis from the prior carcinoma, three lung nodules were identified in both right and left lung, with the largest measuring 1.8 cm. Biopsy from one of lung nodules confirmed metastatic adenoid cystic carcinoma, thus, the external auditory tumor was diagnosed as metastatic adenoid cystic carcinoma. Surgical removal of the ear mass was followed by chemotherapy. Patient expired 13 months later from widespread metastasis.

Discussion

Adenoid Cystic Carcinoma (ACC) seldom arise in the EAC, however ACC is the most common malignancy of glandular origin [2]. ACC exhibits extensive local tissue infiltration and perineural spread, which results in a high rate of recurrence despite aggressive surgical resection in this case and many other published reports. Recurrence and metastasis are very common in ACC, and among those patients who had recurrence, 65% were women [1].

ACC is associated with a high mortality rate, and it often recurs after prolonged periods of time [4]. Although this tumor most often occurs in the salivary glands, it can also be found in many anatomic sites, including the breast, lacrimal gland, lung, brain, Bartholin gland, trachea, and the paranasal sinuses [9]. The most common site of metastatic disease is the lung. Other, more rarely reported areas of metastasis include lymph nodes, soft tissue, bone, brain, and kidneys. Incomplete removal of the primary tumor contributes to rare cases of local recurrence and metastasis [5].

The behavior of ACC has been shown to be unpredictable [4], and therefore identifying which subtype has become important when considering possible treatment options. When identifying ACC, it is important to note that there are 3 subtypes of ACC: tubular, cribriform, and solid [6]. It is believed that the three patterns reflect a progression of cellular proliferation and aggressiveness of biologic behavior [6]. In the case of this patient's biopsy of the skin in the EAC, it was demonstrated through immunohistochemical staining that there were various proportions of all three patterns present, with the cribriform subtype being the most prominent, as well as most of the tumor cells staining positive for c-KIT (CD117). Our patient's case showed strong positivity of c-Kit in all the three tumor components. The c-KIT proto-oncogene encodes a transmembrane receptor-tyrosine kinase; it is known to have a significant role in the normal migration and development of germ cells and melanocytes. The pattern of c-KIT expression in ACC differs with histologic subtype; the difference in the pattern of c-kit expression in tubular and cribriform ACC, as compared with solid variants of ACC, which suggests a loss of cellular heterogeneity [7].

Local lymph node involvement is rare because these tumors are rare and they often present at an advanced stage. Most sources agree that aggressive treatment of these tumors is necessary. Surgery is still the treatment of choice since radiotherapy has not been found to lead to a longer survival rate, but still has a role as a palliative measure when combined with Chemotherapy [8]. ACCs have a long clinical course and questionable prognosis with minor salivary gland ACCs having a worse prognosis than those of the major salivary glands [10]. Lymphatic spread occurs less commonly than with other malignant epithelial tumors. Blood spread to distant sites occurs late in the course of the disease and distant metastasis is common particularly in the lung, and usually whenever the primary tumor has been inadequately treated [11].

Today, immunohistochemistry and scrupulous surgical planning with the aid of high resolution CT scans and magnetic resonance imaging have enabled more accurate diagnosis to be made and, therefore, more radical treatment.8 Aggressive primary resection might be the only way to lower the mortality of patients with ACC in sites of prevalent local spread [4].

It is our hope that this report raises awareness of what remains an unmet need in metastatic adenoid cystic carcinoma management and that continued investigation drives further development of efficacious diagnosis and safe treatments for improving patient outcomes.

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