

Case Report

A Rare Case of Adenoid Cystic Carcinoma Isolated in the Mastoid

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Adenoid cystic carcinoma (ACC) isolated in the mastoid is very rare. Its diagnosis, especially in the early stage, is often challenging as during that stage, the signs and symptoms may be nonspecific. Our paper describes a case of a patient with an isolated mass in the left mastoid with persistent peripheral facial paralysis, and this was initially diagnosed as facial neuritis and granulation. However, histological examination later revealed an ACC exhibiting tubular and cribriform patterns. Our paper discusses the diagnostic basis, treatment, and outcomes for this case to improve the understanding of ACC isolated in the mastoid.

KEYWORDS: Adenoid cystic carcinoma, mastoid

INTRODUCTION

Adenoid cystic carcinoma (ACC) isolated in the mastoid is very rare. Because of nonspecific signs and symptoms at presentation, some patients may be misdiagnosed with benign conditions, which may delay initial treatment. We present a rare case of ACC isolated in the left mastoid that was initially diagnosed as facial neuritis and granulation; and we discuss the diagnostic basis, treatment, and outcomes.

CASE PRESENTATION

A 35-year-old woman presented to our clinic in 2016 with a history of persistent peripheral facial paralysis with facial numbness for one year. She had been receiving medical treatment by methylcobalamin (Mecobalamin; China Weicai Pharmaceutical Co. Ltd., Zhengzhou, China) for the presumptive diagnosis of facial neuritis. The patient's medical history included intermittent episodes of pus discharge from the left ear two years back that was successfully treated with antibiotics. The patient provided written informed consent.

On physical examination, the patient had left facial nerve (FN) palsy (House Brackman Grade V) with hypogeusia. Her ear canal, tympanic membrane, and audition were normal. The cranial magnetic resonance imaging (MRI) revealed a mass in the left mastoid (Figure 1a). Ultrasound of salivary glands was normal. The axial and the coronal computed tomography (CT) imaging of the mastoid revealed a mass beside the vertical FN in the left mastoid (Figure 1b, 1c), and no abnormalities were observed in the mid-ear, vestibular aqueduct (Figure 1d).

Left canal wall-up mastoidectomy (CWUM) and partial FN decompression was performed on October 24, 2016. The mastoid air cells were resected, preserving the posterior wall of the external auditory canal. An isolated tumor was found beside vertical FN without adhesion, which resembled a cholesterol granuloma (Figure 2a); and the FN was found intact after the tumor was resected (Figure 2b). In combination with the preoperative examination, we considered the mass in the mastoid to be benign, and that her FN palsy might be caused by compression of the FN by the mass. Therefore, we decompressed the whole vertical FN and a part of the horizontal FN by incision of the FN sheath.

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However, the final histological analysis showed that this tumor consisted of epithelial and myoepithelial cells appearing in tubular and cribriform patterns. Pseudoglandular lumens contained basophilic and eosinophilic material (Figure 3a). The myoepithelial cells expressed p63 (Figure 3b), smooth muscle actin, and calponin, while the epithelial cells showed strong positivity for cytokeratin (CK) (Figure 3c), epithelial membrane antigen, and CD117 (Figure 3d) by immunohistochemical staining. The Ki67 positive index was approx-

imately 15%. Only 5% of the cells showed low levels of p53 nuclear expression.

In light of the histological analysis, we revised the diagnosis to ACC, and we advised her to undergo resection of the FN and the parotid gland. However, the patient refused the operation because of the risk of complications; and she instead chose to undergo radiation therapy with a total dose of 60 Gy. As of her last follow-up visit in May

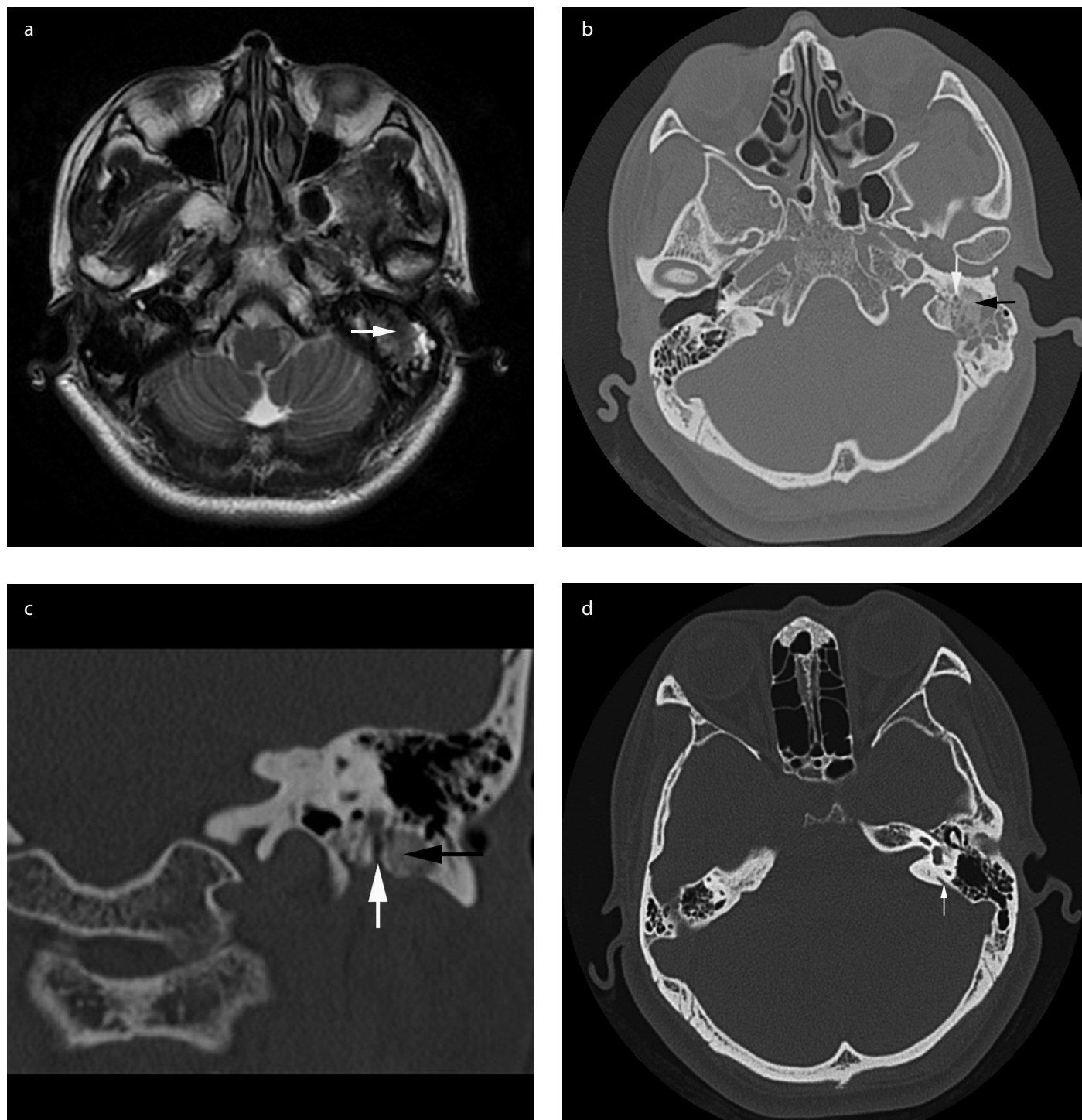


Figure 1. a-d. T2-weighted fast-spin echo magnetic resonance imaging (T2-FSE MRI) revealed a heterogeneous mass (black arrow) isolated in the mastoid (a). The axial and the coronal computed tomography (CT) of the mastoid showed the tumor (black arrow) was beside the vertical facial nerve (white arrow) (b, c). The CT section at the level of the vestibular aqueduct (white arrow) showed it was intact without tumor invasion (d).

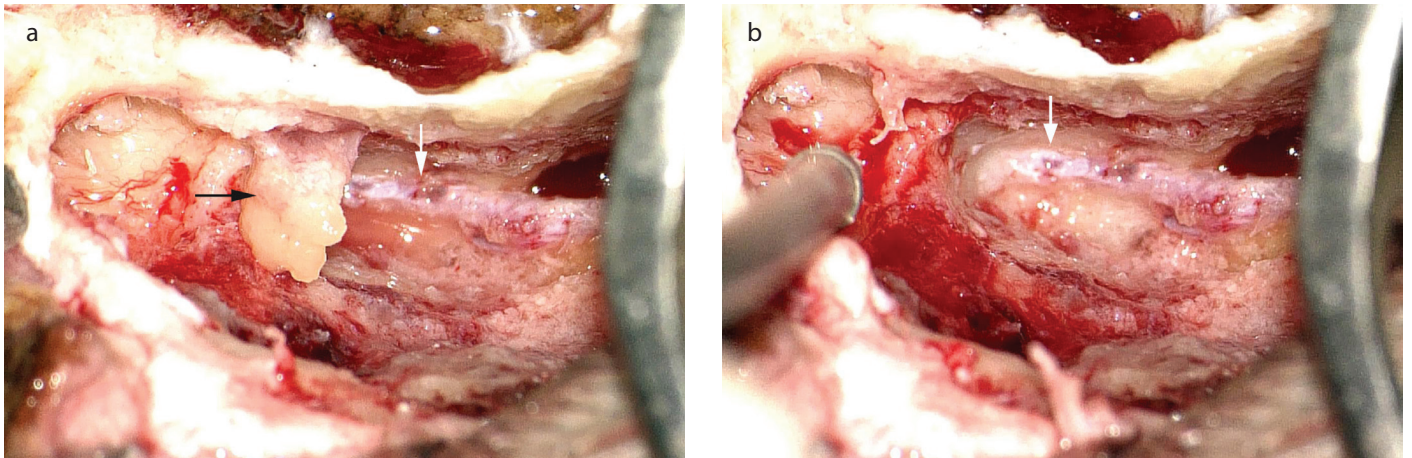


Figure 2. a, b. The intraoperation views before and after tumor resection showed that the tumor a) (black arrow) was beside the vertical FN b) (white arrow) that was found intact after the tumor was resected.

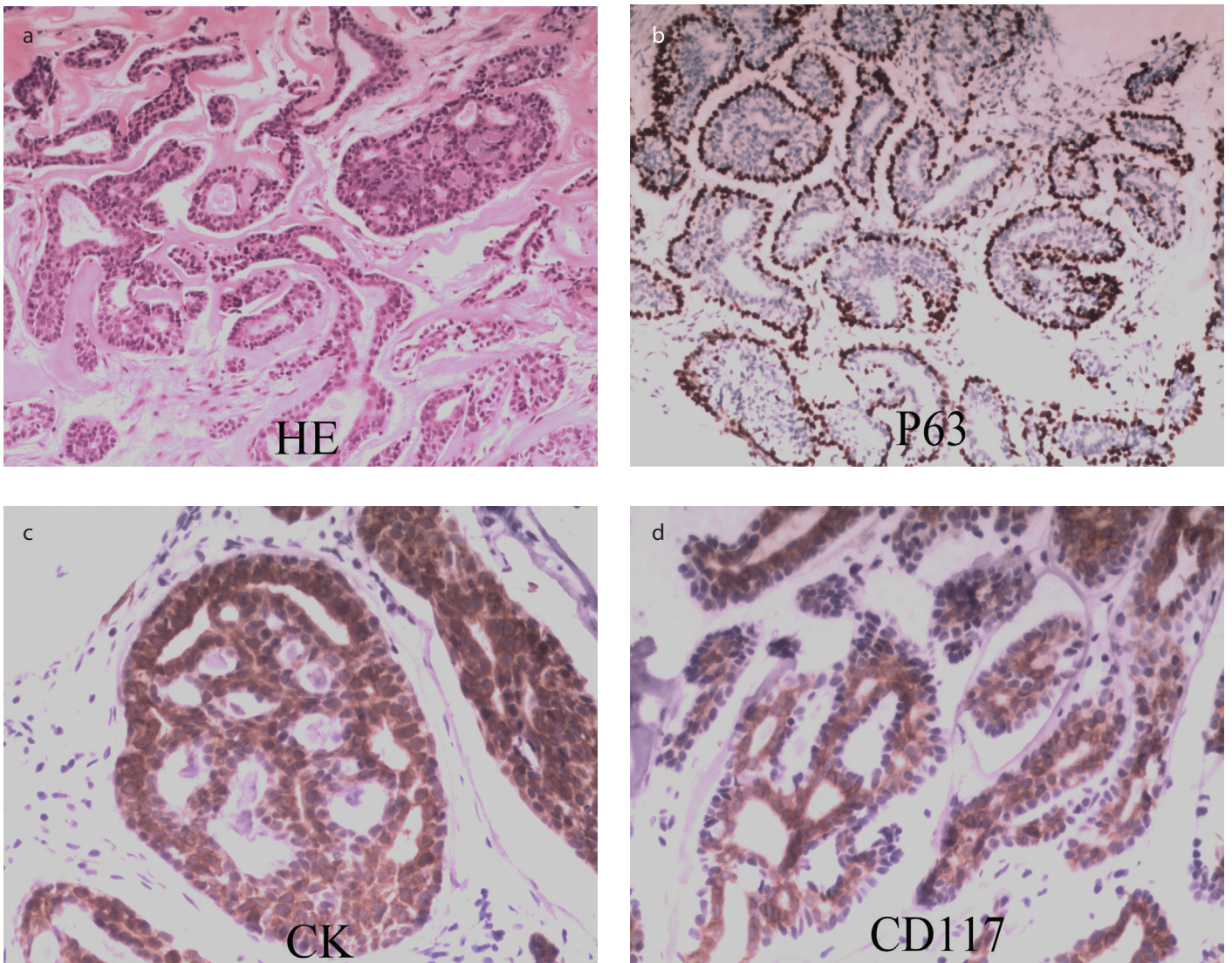


Figure 3. a-d. Hematoxylin and eosin staining ($\times 200$). Tumor was mainly composed of tubular and cribriform patterns. Pseudoglandular lumens contain basophilic and eosinophilic material (a). Immunohistochemical staining ($\times 200$). The myoepithelial cells express p63 (b). Immunohistochemical staining ($\times 400$). The epithelial cells express CK and CD117 (c, d).

2018, her hearing was normal, and the facial numbness was relieved, but the facial paralysis persisted. There were no signs of tumor recurrence on imaging examinations.

DISCUSSION

Adenoid cystic carcinoma is rare, and it accounts for only 1%~4% of all head and neck tumors ^[1]. ACC originates mainly in the small sali-

vary gland, but it may also originate in the lacrimal gland, bronchus tree, esophagus, and external auditory canal ^[2]. ACC isolated in the mastoid has not yet been reported. Magliulo ^[3] presented two cases of ACC within the middle ear and mastoid, and considered that the ACC could have arisen from ectopic salivary gland tissue located in the middle ear. Our case of ACC in the mastoid may have arisen from ectopic salivary gland tissue as well.

Initially, ACC isolated in the mastoid may not cause any obvious symptoms. However, with the growth of the tumor, especially after it invades the tympanic cavity, patients may suffer from hearing loss, tympanic cavity discomfort, otorrhea, and other symptoms. Because of its neurotropism ^[4], especially the invasion of FN, ACC can lead to facial numbness and facial paralysis. Imaging examinations are also helpful in diagnosing ACC. CT may reveal heterogeneous and irregular low-density shadows. MRI may also reveal a heterogeneously enhanced tumor after infusion of contrast material. Furthermore, MRI is more sensitive for the detection of perineural spread compared to CT. Although ultrasound is frequently used in the initial detection of ACC within the major salivary glands, it is difficult to distinguish ACC from other tumors ^[5]. Histological analysis is the most accurate method to diagnose ACC. Epithelial and myoepithelial cells appear as cribriform, tubular, and solid nests.

According to other research, the solid type of ACC is more prone to distant metastasis, and indicates a poor prognosis ^[6]. Radical surgery is recognized as the main treatment for ACC, but whether one also needs adjuvant radiotherapy or chemotherapy is still controversial. Most scholars have concluded that radiotherapy after surgery is more helpful than either surgery or radiotherapy alone ^[7]. However, Loyd S ^[8] analyzed the prognosis of 2286 cases of head and neck ACC, concluding that the overall survival and cause-specific survival was not improved by the use of adjuvant radiotherapy.

Our patient had a previous history of cured otorrhea with normal hearing. Thus, the doctors she visited in past ignored the lesion in the mid-ear and the mastoid. Even after she began experiencing facial numbness and facial paralysis, she was still diagnosed with facial neuritis for one year before she underwent an MRI examination. Further CT and ultrasound confirmed that the tumor was isolated in the mastoid, and that the salivary glands were normal. The clinical presentation, imaging examinations, and the intraoperative findings all mislead us to consider the mass as an inflammatory granulation or benign tumor. Therefore, we did not initially resect the FN or the usual suspicious primary foci such as the parotid glands.

Although we advised the patient to have a second operation after the diagnosis was confirmed, she refused and chose to undergo radiotherapy alone instead. Fortunately, the histological analysis showed mainly cribriform and tubular patterns without peripheral nerve invasion, which may lead to a better prognosis. A recent review reported that after initial treatment for ACC in the head and neck, the recurrence time was about 31-64 months, the metastasis time was about 47.7-60 months. Even if there is a relapse, survival rate was not affected as long as patients received timely treatment ^[9]. Our patient had negative results on chest CT and enhanced MRI of head and neck at her last follow-up visit in May 2018.

CONCLUSION

Otolaryngologists should be vigilant when counseling patients with persistent facial numbness and facial paralysis, especially when imaging examination reveals a mass in the mastoid, because this may indicate ACC. Enhanced CT or MRI and intraoperative frozen sections maybe helpful for detection. CWUM with complete tumor resection and FN decompression can preserve function, and radiotherapy after surgery may be effective for an isolated ACC in the mastoid. Furthermore, a very thorough follow-up with the timely recognition and diagnosis of ACC is necessary to improve outcomes and limit both regional recurrence and distant metastasis.

Informed Consent: Written informed consent was obtained from the patients' parents who participated in this study.

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Conflict of Interest: The authors have no conflict of interest to declare.

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