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CASE REPORT
Reconstructive

Adenoid Cystic Carcinoma Developed from the Parotid Gland to the Ear Lobe of a Young Woman

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Summary: Adenoid cystic carcinoma (ACC) is a relatively rare malignant tumor. It is more common in women than in men and typically develops in the lacrimal, salivary, and breast glands. ACC of the external auditory canal (EAC) is exceedingly rare, and its invasion into the ear lobe is even more unusual. In this report, we present a case of ACC that presented as a mass on the surface of the ear lobe in a 28-year-old woman and was initially diagnosed as infected atheroma. For wide resection of the tumor, half of the entire auricula was resected and superficial parotidectomy was performed. After confirming no tumor cells on the surface of the facial nerve, the defect was reconstructed by the combination of platysma muscle flap to prevent Frey syndrome and free forearm flap for the ear lobe form. There was no recurrence or metastasis of the tumor, and Frey syndrome did not occur at 2 years and 8 months after surgery. The patient was satisfied with the result, oncologically and cosmetically. Even in young patients, comprehensive treatments (including diagnosis, resection, and reconstruction) are important in painful ear lobe masses.
(Plast Reconstr Surg Glob Open 2021;9:e3393; doi: 10.1097/GOX.0000000000003393; Published online 16 February 2021.)

Adenoid cystic carcinoma (ACC) is a relatively rare malignant tumor. It is moderately common, more so in women than in men, and the tumor usually develops in the fourth to sixth decade of life in the lacrimal, salivary, and breast glands.

ACC of the major salivary gland typically presents as a painful mass or swelling of the gland.[1] It generally progresses slowly; however, because of its wide perineural invasion, the tumor often extends far beyond the tumor margin, and adequate surgical resection is sometimes difficult.

In this study, we report our experience of an ACC that developed in a young woman and presented as a mass at the ear lobe, which was initially diagnosed as infected atheroma.

CASE REPORT

A 28-year-old woman noticed induration and a small mass at the posterior ear lobe from approximately 4.5 years before presentation with gradually increasing pain. Internal antibiotics had been administered several times previously; however, the mass remained. A diagnosis of atheroma or abscess was made at the dermatology clinic, and the patient was referred to our clinic for surgical intervention.

Upon presentation to our clinic, an 18 × 10 mm firm subcutaneous induration was noted in the upper ear lobe (Fig. 1). Doppler ultrasound showed heterogeneous low echoic area, and nutrient vessels were recognized inside the tumor (Fig. 1). The mass appeared to be solid, and magnetic resonance imaging (MRI) revealed a 21 × 18 mm irregularly formed mass extending from the ear lobe skin to the superficial parotid gland through the EAC (Fig. 1).

A biopsy was performed, and pathological findings showed alveolar invasion of small tumor cells full of chromatin into the dermis. The nest of the tumor presented with a solid to tubular structure and was diagnosed as ACC. Enhanced computed tomography scan showed no metastasis on the cervical lymph node or other organs. Therefore, we planned wide resection of the tumor and subsequent reconstruction by free forearm flap and pedicled platysma muscle flap. Wide resection secured a horizontal margin of 1 cm from the ear lobe induration, and half of the entire auricular area, including the caudal side of the EAC, was resected. MRI showed that the tumor remained at the superficial parotid gland; therefore, we performed superficial parotidectomy to preserve the facial nerve (Fig. 2). Resection of the nerve was decided by intraoperative rapid pathological diagnosis. Because

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Received for publication September 1, 2020; accepted December 2, 2020.

DOI: 10.1097/GOX.0000000000003393

Disclosure: *The authors have no financial interest to declare in relation to the content of this article.*

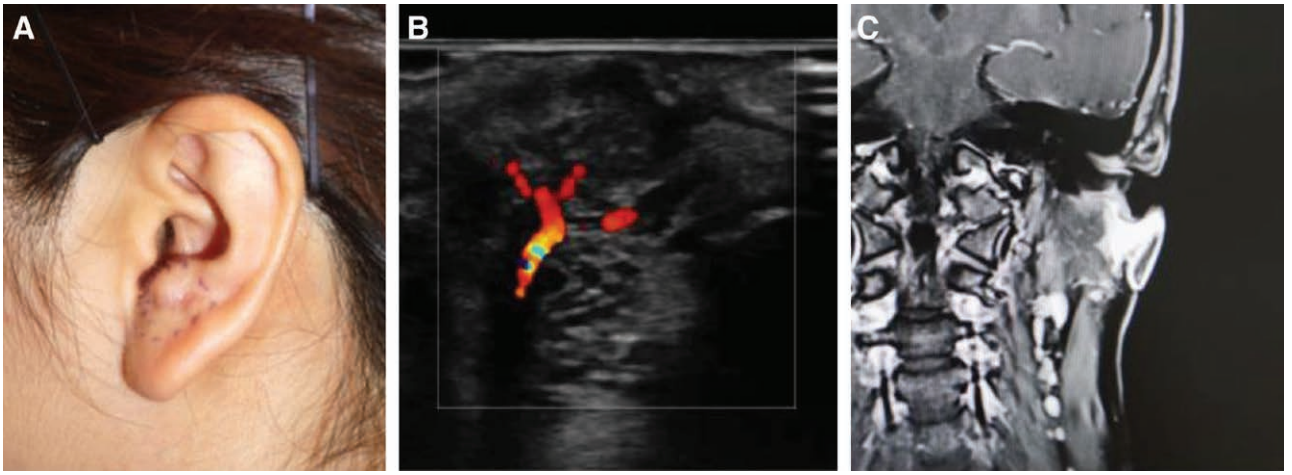

Fig. 1. Clinical appearance of the tumor at first visit to our clinic. A 28-year-old woman presented with a mass at the posterior ear lobe (A). Doppler ultrasound showed nutrient vessels inside the tumor (B). The entire mass was strongly enhanced by MRI (C).

no tumor cells were detected on the surface of the facial nerve, we could preserve the facial nerve.

After wide resection of the tumor, the remaining exposed parotid gland was covered by the 7 × 5 cm platysma muscle flap to prevent Frey syndrome and hollow at the cheek (Fig. 3).[2]

Cartilage defect from the edge of the ear canal resection was reconstructed using the conchal cartilage graft from the unaffected side, and the ear lobe defect was reconstructed by an 8 × 5 cm free forearm flap using the facial artery and vein as a recipient vessel (Figs. 2 and 3). The free forearm flap was folded into a U-shape, and the form of the flap was adjusted to the shape of the ear lobe (Fig. 3).

Pathological findings of the resected specimen also showed ACC originated from the superficial parotid gland. The tumor cells were extended to the dermis of

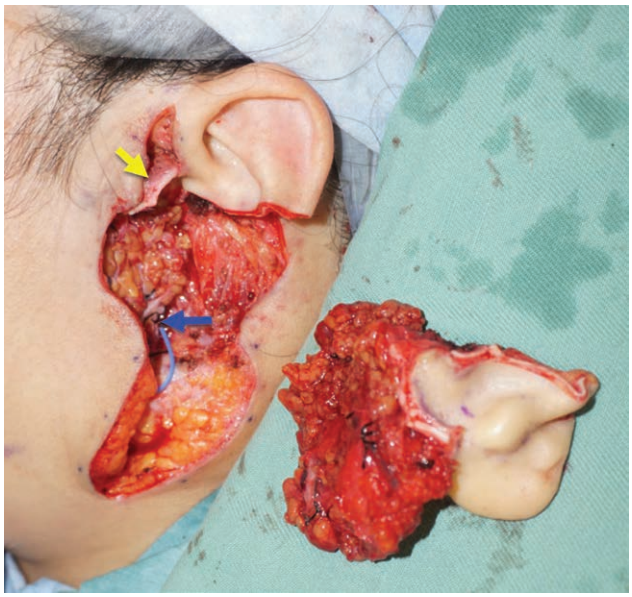

Fig. 2. Intraoperative appearance after resection of ACC through the external auditory canal (EAC) to the surface of the ear lobe. We identified all facial nerve branches (marginal mandibular branch: blue arrow), and kept those branches after wide resection with superficial parotidectomy. To reconstruct the EAC, we grafted conchal cartilage from the unaffected side to the edge of remaining EAC (yellow arrow).

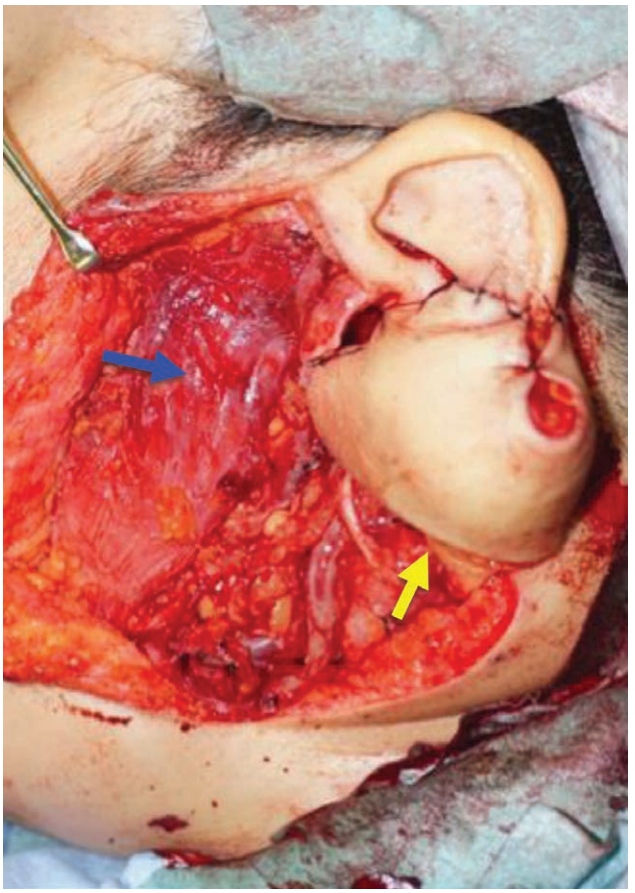

Fig. 3. Intraoperative appearance of reconstruction. The superior based platysma muscle flap was transferred to cover the exposed parotid gland (blue arrow). The radial forearm flap was folded into a U-shape, and the form of the flap was adjusted to the shape of the ear lobe (yellow arrow).

the ear lobe and main localization of the tumor was subcutaneous tissue (TNM classification: T4aN0M0, Stage IV A).

Given the patient's age and to preserve a treatment option in case of future recurrence, we did not perform postoperative radiation therapy.

Three years and 2 months after surgery, there was no recurrence or metastasis of the tumor, and gustatory sweating (Frey syndrome) did not occur (Fig. 4). Although future defatting of the flap is necessary, the patient was satisfied with both the oncologic and cosmetic outcome.

DISCUSSION

The diagnosis and treatment of ACC is challenging, particularly because of its biological behavior, high rate of local recurrence, and local perineural spread.[1,3] ACC of the EAC is exceedingly rare,[4] and invasion into the ear lobe is even more uncommon. We were unable to find any study describing ACC invasion into the ear lobe; however, Green et al reported a cohort study of ACC of EAC, and the rate of distant metastasis in the ACC of the external ear (7.9%) was much lower than that of the head and neck lesion in previous literature (25%–62%).[4] Lymph node metastasis had a negative impact on survival rate, which was not improved with postoperative radiation.[4] Thus, primary surgery is even more important in ACC of the EAC and external ear. Liu et al reported that the surgical procedure for an ACC of the EAC is more extensive.[5] Because of the EAC's intimate anatomic relationship with the parotid gland, an ACC in this location tends to invade the parotid gland even in the early stage. Perineural invasion is a significant pathological feature of ACC of the EAC, with an estimated incidence of 55%–88%, which is much higher than regular ACC.[5–7] Even without obvious parotid gland invasion in the enhanced MRI, superficial parotidectomy is recommended,[5,6] and even parotidectomy is suggested because of the false-negative signal of parotid gland invasion on preoperative MRI.[8]

In our case, parotid gland invasion was obvious on preoperative MRI; however, the tumor remained in the superficial gland. From pathological findings, the tumor was thought to be originated from the parotid gland, not from EAC, and there was no perineural invasion on the intraoperative examination. Thus, we decided to preserve facial nerve and widely excised the tumor with superficial parotidectomy.

Our patient was much younger than usual cases, and the initial diagnosis was infected atheroma. Even in cases with young patients, we should check minimal imaging study such as Doppler ultrasound at our first look and the tumor characteristics or stage should be evaluated precisely with additional imaging and pathological studies. As we secure 1-cm horizontal margin from the mass induration at the ear lobe and resect widely with caudal half of the EAC and superficial parotidectomy, we could obtain good prognosis without any adjuvant therapies including radiation.

By reconstructing the defect using a combination of platysma muscle flap and free forearm flap, we avoided gustatory sweating (Frey syndrome) and a hollow area at the cheek,[2] and we maintained the form of the external ear to the patient's satisfaction. Careful observation for recurrence and metastasis is still required; however, comprehensive treatments (including diagnosis, resection, and reconstruction) are even more important in painful ear lobe masses.

Fig. 4. Appearance at 3 years postoperative. The scar was matured and there was no hollow at the cheek. There was no recurrence or metastasis of the tumor.

CONCLUSIONS

We experienced a case of ACC that presented as a painful mass at the ear lobe in a young woman. Although there have been few reports of ear lobe malignancy, careful medical examination is required, even in young patients.

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