Case Report

Large Intradiploic Epidermoid Cyst of the Temporal Bone

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Intradiploic epidermoid cyst of the skull, despite being rare and representing less than 1% of all intracranial tumors, is a benign lesion that is derived from an ectodermal cell rest within the cranium. The frontal and parietal bones are the most common sites for the cyst; however, the temporal bone, although rarely, is also involved. Intradiploic epidermoid cysts have slow growth rates and often grow to enormous sizes without producing any measurable neurological symptoms. For treatment, a complete removal of the cyst with its capsule is important to avoid recurrence. Very rare malignant transformations of the cysts have been reported, generally in cases with repeated recurrences or infections. A 16-year-old female patient was diagnosed with a temporal bone intradiploic epidermoid cyst behind the left mastoid cavity, which was successfully removed through a transmastoid approach.

KEY WORDS: Epidermoid cyst, mastoid, temporal bone

INTRODUCTION

Intradiploic epidermoid cysts, unlike middle ear cholesteatoma, occur in the intradiploic space of the bones. Although the cranium is the most common site of the cysts, cases of intradiploic epidermoid cysts occurring in the temporal bone have also been reported [1]. The occurrence of intradiploic epidermoid cysts in the temporal bone is extremely low; there are currently only 20 global [2-6] and two Korean cases reported [7, 8]. The authors encountered a patient, who visited the tertiary referral hospital with conductive hearing loss, due to occlusion in the external auditory canal (EAC), with no other neurological symptoms. Based on the radiologic imaging results, an intradiploic epidermoid cyst was suspected, and the patient was given surgical treatment. This literature reports the histological findings of the detection and the surgical treatment of an intradiploic epidermoid cyst.

CASE PRESENTATION

A 16-year-old female outpatient with no history of head trauma and surgical vestibular intervention reported a 2-year presence of an EAC stenosis and progressive hearing impairment. Observations revealed a hard rubbery mass located postero-superiorly to the right EAC—despite the normally shaped right auricle—that was blocking the orifice and hindering the observation of the right tympanic membrane (Figure 1), along with an ipsi-lesional preauricular fistula. On the other hand, the left side auricle, EAC, and tympanic membrane were diagnosed as normal. The Weber test detected a lateralization to the right ear, and the neurologic examination results of the facial and vestibular nerves were normal. On the pure tone audiometric evaluation, air conduction was 42 dB and bone conduction was 7 dB on the right side, and the hearing level was normal on the left (Figure 1). In preoperative TBCT, the axial view, the lesion was located near the mastoid ad antrum from the level of malleus head, not involved with the labyrinthine, and the middle ear cleft was normal and destructing the cortical bone of mastoid and the superior bony portion of the EAC (Figure 2a). The CT coronal view revealed a 2 cm x 2 cm mass-like space-occupying lesion blocking the EAC and destroying the cortical bone of the mastoid and the superior bony portion of the EAC (Figure 2a). From the axial view, the lesion was located near the mastoid ad antrum from the level of the malleus head, not involved with the labyrinthine, and the middle ear cleft was normal (Figure 2b). The MRI examination revealed a heterogeneous cystic mass filling the EAC that was destroying the surrounding mastoid air cells with high signal intensity in the peripheral rim and irregular and low signal intensity in the center (Figure 2c, d). The transmastoid approach for the surgical intervention revealed a smooth, dark brown cystic mass located on the right superior EAC (Figure 3a, b). The cystic mass contained approximately 2 cc of a dark and serous fluid. The inner surface of the cystic mass was enveloped by a smooth mucosa, and a blind porch-partially granulated tissues that adhered to the temporal bone was formed. The inner table was raked out with a drill in order to completely remove the entire cyst, and a cortical bone of the mastoid was harvested and remodeled into...
1 cm x 1 cm sections to reconstruct the EAC wall (Figures 3c, d). The upper external auditory canal was reinforced with a Koerner flap and deep temporalis fascia. In the hematoxylin and eosin stain, the cyst-lined by hyperkeratotic squamous epithelium—was filled with keratinous material without skin appendages (Figure 4).

**DISCUSSION**

An epidermoid cyst is a benign tumor that originates from an ectodermal tissue. The most likely theory of its origin is that it is caused by the movement of epidermal tissue when a neural canal is closed at 3-5 weeks of fetal age [9]. However, it can also be caused by the insertion of epidermal tissue into the bone during a trauma; 24% of these cysts are reported to be associated with the trauma history [1]. Intradiploic epidermoid cyst is only 1% of all intracranial tumors and can be found in the cranial or spinal cavity. Its occurrence is about 25%, and it usually occurs in the cranial bones; according to Arana et al. [10], it occurs mostly in the parietal (35.1%), frontal (27%), and rarely temporal bones (8.1%). Moreover, among the 73 intradiploic epidermoid cyst examples from the old case studies that Miller et al. [2] analyzed, only 10 showed evidence of temporal bone involvement.

An intradiploic epidermoid cyst located near the mastoid bone, as reported by otolaryngologists, can sometimes be confused with congenital cholesteatoma. Both congenital cholesteatoma and intradiploic epidermoid cyst show keratinized squamous epithelia, including keratin, and are hard to differentially diagnose with MRI, because both have a low signal on T1-weighted images and a high signal on T2-weighted images [2-5, 9-11]. Our case is reported to be a back-side tumor that blocked the EAC inlet according to the computed tomography and is different from general cholesteatoma. The computed tomography and MRI examination revealed the destruction of the skull diploic space by a well-defined mass.
The most common symptom of an intradiploic epidermoid cyst is painless swelling of the scalp due to the slow growth of the tumor, caused by desquamation of the epithelium or proliferation of the epithelial cells [1-3]. They can grow to be gigantic without any neurological symptoms [3]. Occasionally, once they become sizeable, they can cause headaches and, more rarely, increased intracranial pressure, seizure, and on-going hemiparesis and other local neurological signs [10, 11]. Other problems that can occur include bone resorption near the intradiploic epidermoid cyst due to middle ear cavity or cholesteatoma infection, interleukin-1, fibroblast production, collagenase, and granulocyte-macrophage colony-stimulating factor secretion, etc [2]. Complications of intradiploic epidermoid cyst are abscess formation, hemorrhage, and malignant change, etc [1, 3, 6, 13]. There are more risks of malignant changes with recurrent operations or repetitive intra-cystic infections [11]. Malignant changes of intradiploic epidermoid cyst should be suspected when the enhancement media goes well by the injection of contrast media during the MRI or if the tumor is fast-growing [13]. Standard treatment of an intradiploic epidermoid cyst is surgical removal with complete resection, including the cystic capsule [2]. At this time, to reduce many complications from the operation, intradiploic epidermoid cyst has to be desquamated from the bone and dura mata carefully. The rate of recurrence after operation is reported as about 8.3% to 25% [11, 4]. This index case removed the cyst completely, making the connection with the aditus ad antrum and reconstructing the defective section with cortical bone to prevent EAC obstruction. For early detection in the future, MRI will be used for periodical follow-up.

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

Peer-review: Externally peer-reviewed.


Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: This case was supported by Basic Science Research Program through the National Research Foundation of Korea (NRF) funded by the Ministry of Education, Science and Technology (2013R1A1A2005018).

REFERENCES