Glial Heterotopia of the Middle Ear and Eustachian Tube in Children

Lei Wu, MM¹, Jing Sun, MM¹, and Feng Zhang, MM¹

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Extracranial glial heterotopia usually occurs along the midline structures. Nonmidline extracranial occurrences (eye, lung, skin) are rare. In the literature,¹ very rare cases of glial heterotopia of the middle ear have been reported, and none involved the parapharyngeal space or the Eustachian tube (ET). We report a case of a glial heterotopia in the left parapharyngeal space that extended into the middle ear cavity and external auditory canal via the ET. Approval for this report was granted by the Ethics Committee of the Children’s Hospital, Zhejiang University School of Medicine.

Case Report

A 3-year-old boy presented with a 10-day history of left-sided earache. There was no history of congenital anomalies, trauma, or ear surgery. Physical examination revealed a tympanic membrane that was intact but bulging because of a mass in the middle ear. Tonsil asymmetry was noted with the left side encroaching on the uvula. The right ear was normal. Auditory brainstem response (ABR) showed that the V wave was present at 90 dBHL and 15 dBHL on the left and right sides, respectively. High-resolution computed tomography (CT) showed an ovoid mass in the left parapharyngeal space that extended into the middle ear and external auditory canal via the ET (Figure 1). The right ET was clearly larger than the left ET. Contrast-enhanced CT showed a nonenhancing mass. Magnetic resonance imaging (MRI) showed a mass with equal T1 and long T2 signals with no enhancement after contrast injection (Supplemental Figure S1, available at otourjournal.org).

Biopsy was performed transcannal, through a myringotomy incision. Histopathology showed inflammatory cell infiltration and mature neuroglial tissue. A teratoma was initially considered, but a glial heterotopia could not be excluded. Although complete excision was not possible, the patient underwent a left tympanomastoidectomy to diagnose the mass. A gray-white fibrotic mass was detected in the epitympanic area. There was no evidence of cranial bone defect or leakage of cerebrospinal fluid. There were no postoperative complications.

Pathological examination of the specimen revealed mature neuroglial tissue representing heterotopic brain tissue in a fibrovascular stroma. There was no cellular atypia or mitotic activity. Immunohistochemistry found the cells to be intensely positive to S-100 and glial fibrillary acidic protein (Figure 2).

The mass recurred 12 months after surgery. High-resolution CT showed that the mass was similar to the clinical

Figure 1. High-resolution computerized tomography showed an ovoid mass in the left Eustachian tube.

¹The Children’s Hospital, Zhejiang University School of Medicine, Hangzhou, China

Corresponding Author:
Jing Sun, Department of Otolaryngology—Head and Neck Surgery, the Children’s Hospital of Zhejiang University School of Medicine, Hangzhou 310003, China
Email: zjuentes66@gmail.com
preoperative assessment. The patient’s parents did not want to repeat surgery, so there was no further treatment.

Discussion
Heterotopic neuroglial tissue is defined as a mass of mature brain tissue isolated from the cranium or spinal canal. Most reported examples involve midline structures, including the nose and nasopharynx (so-called nasal glioma) as well as the oropharynx, palate, lips, tongue, and tonsils. Nonmidline heterotopic brain tissue is rare. The most frequently reported choristoma of the middle ear is salivary gland tissue; heterotopic brain or glial tissue are very rare in this region. To our knowledge, this is the first report of a glial heterotopia involving the parapharyngeal space and the ET.

Unlike their midline counterparts, most middle ear region glial heterotopias are diagnosed in adult patients. Some authors have described chronic infection or inflammation, previous trauma, or surgical procedures as predisposing factors for the development of middle ear heterotopia or encephalocele. These findings lend support to the concept that middle ear region neuroglial masses are most often acquired encephaloceles. However, this patient had no predisposing factors and no evidence of a connection of the lesions to the central nervous system. This case has the possibility of being a true glial heterotopia.

In other case reports, lesions were confined to the middle ear cavity and mastoid antrum, and they were completely removed by tympanomastoidectomy. Tympanomastoidectomy was not the best surgical approach for a tumor involving the parapharyngeal space and the ET. Peroral or CT-guided fine-needle aspiration (FNA) are useful in establishing a preoperative diagnosis for a primary parapharyngeal space tumor.

Although glial heterotopias of the middle ear region are rare, they also are necessary to differentiate this lesion from gliomas, meningiomas, neuromas, and other neural neoplasms, although the latter are exceptional in this location. There are no significant histologic differences between heterotopic brain tissue and encephaloceles. The accurate diagnosis of heterotopia versus encephalocele requires the knowledge of radiologic and operative findings of the patient.

Author Contributions
Lei Wu, acquisition of data, drafting article, approval; Jing Sun, interpretation, revisions, approval; Feng Zhang, design and interpretation, revisions, approval.

Disclosures
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Supplemental Material
Additional supporting information may be found at http://oto.sagepub.com/content/by/supplemental-data

References