INTRODUCTION

Heterotopic brain tissue is defined as a mass-like lesion composed of mature brain tissue isolated from the cranial cavity or spinal cord (1). It usually occurs in the non-midline structures, including middle ear, is rare. We described a 50-yr-old man with heterotopic glial tissue in the middle ear and mastoid bone. The patient presented with progressive hearing loss for 8 yr. There was no history of congenital anomalies, trauma, or ear surgery. Computed tomography revealed a mass-like lesion with soft tissue density occupying the middle ear cavity and mastoid antrum. At the operation, a graywhite fibrotic mass was detected in the epitympanic area. Mesotympanum and ossicles were intact. The patient underwent left simple mastoidectomy with type I tympanoplasty. During operation, definite cranial bone defect or cerebrospinal fluid leakage was not found. Histologically, the lesion was composed of exclusively mature, disorganized glial tissue with fibrovascular elements in a rather loose fibrillary background. Glial tissue showed diffuse positive reaction for glial fibrillar acidic protein and S100 protein on immunohistochemical study.

CASE REPORT

A 50-yr-old man presented with progressive hearing loss and otorrhea of the left ear. Previously, he had frequently complained of ear fullness and hearing disturbance for 8 yr. Intermittent otorrhea was developed about 4 months ago. There was no history of congenital anomalies, trauma, or ear surgery. Physical examination revealed otorrhea in the external auditory canal of the left ear. The right ear was normal. Pure tone audiometry showed conductive hearing loss on the left ear (Fig. 1). The right ear was normal. Otorrhea was improved after treatment of otitis externa. The patient had suffered from progressive hearing disturbance. Tympanic membrane showed attic retraction. Computed tomography revealed a mass-like lesion with soft tissue density in the middle ear cavity and mastoid antrum. Bony erosions were suspected in tegmen tympani and long process of incus of the left ear (Fig. 2). Pre-operative diagnosis was attic cholesteatoma. At operation, there was no evidence of cranial bone defect or leakage of cerebrospinal fluid. Microscopic examination of the hematoxylin-eosin stained section revealed a poorly circumscribed pale staining lesion with low cellularity. The lesion consisted of exclusively mature, disorganized glial tissue with fibrovascular elements in a rather loose fibrillary background. Glial tissue showed diffuse positive reaction for glial fibrillar acidic protein and S100 protein on immunohistochemical study.
could be identified. Additional sections were stained for glial fibrillar acidic protein (GFAP), S100 protein, cytokeratin (CK), and microtubule associated protein (MAP-2). The glial tissue revealed diffuse positive reaction for GFAP (Fig. 4A) and S100 protein (Fig. 4B), but negative reaction for MAP-2 and CK.

**DISCUSSION**

Since the initial description of heterotopic glial tissue over the dorsal surface of the cervical spinal cord in 1907 by Wolbach (11), heterotopic brain or glial tissue have been reported in various sites. These lesions may be classified based on the
location and possible pathogenetic mechanisms as follows (7, 12): 1) intraparenchymal central nervous system lesions, 2) dural and leptomeningeal lesions, 3) intracranial extracerebral lesions, 4) midline lesions, 5) distal lesions of the lung and uterus, and 6) extracranial non-midline lesions. Most reported cases have involved extracranial midline structures including the nose and nasopharynx (so-called nasal glioma) as well as the lips, oral cavity, oropharynx, palate (3), and tongue (4). These lesions are most often considered to represent displaced brain tissue during development and are therefore thought to be pathologically related to encephaloceles. Non-midline heterotopic brain tissue is rare. Cases have been reported to involve the scalp, eye and orbit, lung, peritoneum, and middle ear (8). A review of the literature reveals that the most frequently reported choristomas of the middle ear is salivary gland tissue (5, 6), but heterotopic brain or glial tissue is very rare in this region (1, 7-10).

Unlike their midline counterparts, most middle ear hetero-

Fig. 3. Histologic features of the lesion. (A) The lesion is composed of exclusively mature disorganized glial tissue and fibrovascular elements in a rather loose fibrillary background. (B) Distended glandular structures are noted at the periphery of the glial tissue (H&E, ×100).

Fig. 4. Immunohistochemical features. The lesion shows diffuse positive reaction for GFAP (A) and S100 protein (B) (×100).
topic brain tissues 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 (7, 8). These clinical findings support the concept that heterotopic brain tissue in the middle ear region represents acquired encephalocele (7). However, our case had neither apparent predisposing factors nor evidence of connection between the lesion and the central nervous system. Similar cases have been reported in the literature (7). In these cases, and also in our case, a possible explanation for pathogenesis is that the lesion might represent a true neuroglial heterotopia. Other possibilities, such as prior tiny congenital bone defect or remote trauma unrecognized at the time of presentation, can also be considered.

Histologically, heterotopic brain tissue is characterized by varying proportions of neurons and glia associated with chronic inflammation, choroid plexus, and ependymal or leptomeningeal components. Rarely, heterotopic brain tissue of the middle ear was associated with cholesteatoma (8). Heterotopic brain tissue must be distinguished from true neoplasms such as glioma, ganglioglioma, meningioma, neuroma, and schwannoma. The patient’s clinical presentation, the location of the lesion, its relation to surrounding structures as well as careful histologic examinations are helpful in differential diagnosis. In the present case, the heterotopic glial tissue was admixed with distended glands lined by bland-looking flattened to cuboidal epithelial components at the periphery of the lesion, most likely representing entrapped tympanic cavity or Eustachian tube epithelium. Absence of elements unrelated to normal anatomic structures is helpful to avoid the misdiagnosis as teratoma (13). The lesion of our case was exclusively composed of glial tissue lacking neuronal or other brain components. The relative lack of neuronal elements in some heterotopic brain tissue has been attributed to the poor blood supply, resulting in neuronal ischemia and gliosis (14). Genut et al. (15) have noticed the fact that neuronal precursors appeared in the developing brain at the 10th week and suggested that there would be no neuronal elements in the heterotopic brain, if the embryonic tissue has been separated prior to the 10th week.

It should be noted that there are no significant histologic differences between heterotopic brain tissue and encephalocele. It is important to distinguish heterotopic brain or glial tissue from more common middle ear encephalocele because of the risk of infection in the latter. The accurate diagnosis of heterotopia versus encephalocele requires the knowledge of radiologic and operative findings of the patient.

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