

# B-Cell Malignant Lymphoma Presenting as Otitis Media and Mastoiditis Associated with Sinus Thrombosis

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## Key Words

Malignant lymphoma · Otitis media · Sinus thrombosis

## Abstract

Cerebral venous thrombosis as a manifestation of paraneoplastic angitis and otitis media, revealing non-Hodgkin B-cell lymphoma (NHBL), is extremely rare. A 57-year-old woman presented with headache, auditory disturbance and recalcitrant otitis media. Magnetic resonance imaging showed brain edema in the temporal lobe and transverse sinus thrombosis. External drainage under antibiotic treatment was repeated based on a diagnosis of invasive otitis media and mastoiditis associated with infectious sinus thrombosis, but the condition deteriorated progressively. Open surgery for otitis media was performed 6 years after the initial symptoms and after a tumorous lesion had been detected in the middle ear. Pathological findings revealed NHBL. We report a rare case of NHBL presenting as otitis media and mastoiditis associated with sinus thrombosis, and a literature review.

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## Introduction

Direct invasion or metastasis of malignant lymphoma in the middle ear is rare and the clinical features remain unknown [1, 2]. We treated a case of non-Hodgkin B-cell lymphoma (NHBL) in the middle ear presenting as recalcitrant otitis media and mastoiditis associated with transverse sinus thrombosis. The patient was diagnosed 6 years after the initial

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symptoms and was treated successfully with surgery and chemotherapy. We report the clinical characteristics of a rare case of NHBL in the middle ear and review the literature.

### Case Report

A 57-year-old woman presented with chronic temporal headache and right tinnitus, and the symptoms gradually aggravated. Conservative treatment continued at the local Otolaryngology Clinic based on a diagnosis of otitis media. Slight consciousness disturbance, slight fever and auditory disturbance were noted 1 year after the initial symptoms, and the patient was referred to our department. Neurological findings showed slight consciousness disturbance, and an audiometric hearing test demonstrated 30 dB in the right ear, diagnosed as a mild hearing impairment. Magnetic resonance imaging (MRI) showed edematous brain swelling in the right temporal lobe and right tentorium and dural enhancement around the transverse sinus (fig. 1a). Digital subtraction angiography demonstrated occlusion of the right transverse sinus (fig. 1b). No marked venous cortical reflux was shown. MRI showed a heterogeneous mass lesion in the right middle ear and mastoid air cells with dural enhancement around the lesion (fig. 1d). She was diagnosed with recalcitrant otitis media invading the mastoid associated with transverse sinus thrombosis and was treated with antibiotics, hyperosmotic fluids, steroids and repeated external drainage of the middle ear. Consciousness disturbance gradually improved. Symptoms of temporal headache, auditory disturbance, slight fever and radiological findings of brain swelling did not disappear, and remission and exacerbation of these symptoms occurred repeatedly.

Consciousness disturbance occurred again, and chronic temporal headache gradually aggravated 5 years after the first examination in our department. MRI revealed enlargement of the edematous swelling around the right transverse sinus, and the enhanced lesion was also enlarged (fig. 1c). Open drainage of the middle ear with mastoidectomy was performed 6 years after the initial symptoms. Granulation had invaded the eardrum and stapes, and the auditory tube was also filled with granulation (fig. 1e). Infectious granulation was removed and a tumorous lesion was detected. Pathological examination demonstrated diffuse proliferation, predominantly consisting of small to medium-sized lymphoid cells with slightly irregular nuclei (fig. 1f), which were positive for CD20, CD79 and bcl-2, suggesting malignant lymphoma, B-cell type, classified as extranodal marginal zone B-cell lymphoma. Computed tomography and blood and bone marrow evaluation did not show other lesions of malignant lymphoma, suggesting that the otitis media lesion was the primary lesion. She was treated with RCHOP (rituximab combined with cyclophosphamide, doxorubicin, vincristine and prednisolone) chemotherapy. Symptoms improved markedly and there has been no recurrence for 4 years.

### Discussion

Malignant lymphoma can spread to all areas of the body, including the head and neck. Involvement of the temporal bone as part of generalized lymphoma has been reported; however, clinical evidence of temporal bone or middle ear involvement is unusual [3–6]. Presentation with otoneurological signs prior to systemic involvement of a lymphoproliferative disease is also unusual [5]. To our knowledge, only 10 cases of malignant lymphoma originating around the middle ear have been reported (table 1) [1, 5–14]. In the previous 10 reports, the lesion was located in the middle ear in 3, internal in 3, and external in 4. B-cell

type was dominant over T-cell type. Facial nerve palsy was the most common initial symptom and mastoiditis was associated in about 50% of the cases. There was no direct invasion into the intradural region and no association with sinus thrombosis in 10 cases.

Our present case included the following marked characteristics: first, it was difficult to diagnose malignant lymphoma under the suspicion of intractable otitis media. Second, transverse sinus thrombosis caused by dural invasion of a tumor or spread of otogenic infection manifested neurological symptoms. Third, tentorial or dural enhancement adjacent to the transverse sinus did not decrease, and sinus occlusion continued even after remission. Fourth, RCHOP chemotherapy was effective, and the lesion has not recurred for 4 years. The precise mechanism of transverse sinus thrombosis was unknown. Infectious symptoms and dural enhancement showed repeated remission and abrogation under treatment for otitis media with antibiotic administration and drainage. Otogenic infection progression might be dominant in sinus thrombosis formation. Transverse sinus obstruction continued even after symptomatic remission. There was a possibility that sinus occlusion induced by infection progression might abrogate venous circulation, resulting in brain edema in the early stage, and collateral venous flow might gradually develop during the progression of sinus occlusion.

We treated a rare case of B-cell malignant lymphoma presenting with otitis media and mastoiditis associated with transverse sinus thrombosis. Our report may provide relevant information for the management of this disease.

### Disclosure Statement

The authors report no conflicts of interest concerning the case report presented herein or the findings specified in this paper.

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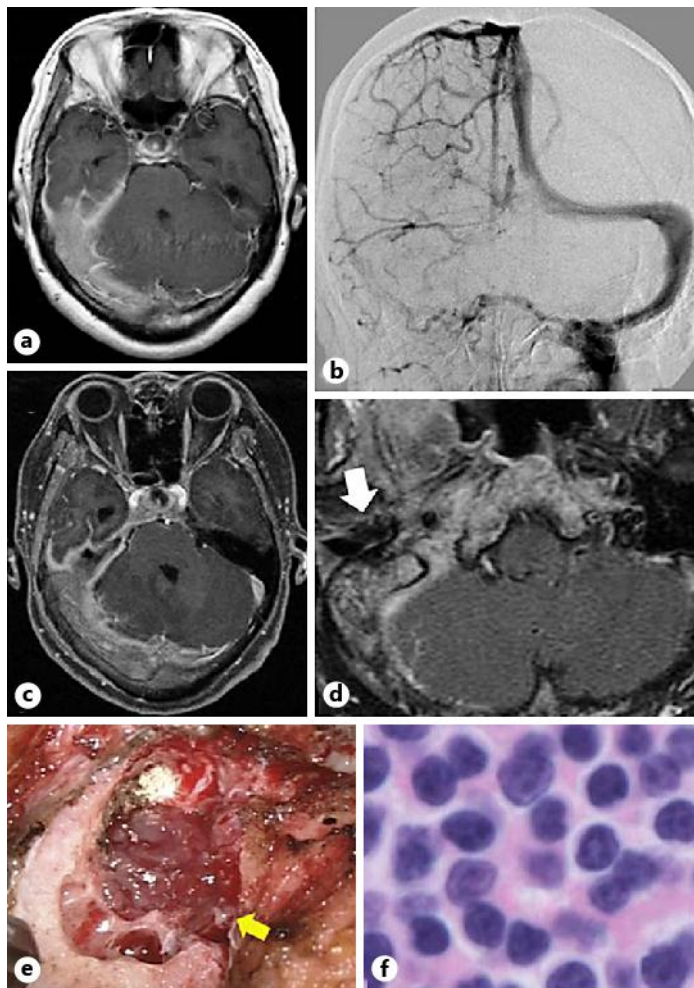
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**Table 1.** Literature review of malignant lymphoma around the middle ear

First author, year	Age, years	Sex	Initial symptom	Location	Cell type	Association
Ide, 1993	55	M	Facial nerve palsy	Mid	T cell	Mastoiditis
Kieserman, 1995	–	–	Cranial nerve palsy	Ext	–	HIV
Danino, 1997	–	–	–	Mid	T cell	Mastoiditis
Angeli, 1998	–	–	–	Int	B cell	None
Fish, 2002	53	F	Otalgia	Ext	B cell	None
Shuto, 2002	49	M	Mass effect	Ext	B cell	Bilateral
Ho, 2004	1	M	Otorrhea, otalgia, facial nerve palsy	Int	T cell	Mastoiditis
Hersh, 2006	–	F	–	Ext	B cell	None
Knapp, 2008	–	–	Facial nerve palsy	Int	B cell	None
Kanzaki, 2011	13	M	Otorrhea, facial nerve palsy	Mid	B cell	Mastoiditis
Present case	57	F	Headache, cranial nerve palsy	Mid	B cell	Mastoiditis

Mid = Middle; Ext = external; Int = internal; HIV = human immunodeficiency virus infection.



**Fig. 1.** **a** MRI on admission showed edematous swelling in the right temporal lobe and marked enhancement in the dura and tentorium around the right transverse sinus. **b** Digital subtraction angiography demonstrated obstruction of the right transverse sinus and no association with cortical venous reflux. **c** MRI prior to open surgery shows aggravation of edematous swelling in the right temporal lobe and cerebellar hemisphere, and enlarged enhancement in the dura and tentorium around the right transverse sinus. **d** MRI shows a heterogeneous mass lesion in the right middle ear and mastoid air cells (arrow), and dural enhancement around the lesion. **e** Intraoperative photo of open drainage of the right middle ear shows a reddish tumorous mass lesion in the middle ear and mastoid air cells (arrow). **f** Hematoxylin-eosin staining shows nodular proliferation predominantly consisting of small to medium, round lymphoid cells, with a large, round chromatin-rich nucleus.