

# Diffuse Large B-cell Lymphoma of the Cerebellopontine Angle in a Patient with Sudden Hearing Loss and Facial Palsy

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Primary lymphoma of the cerebellopontine angle (CPA) is rare in the central nervous system. To our knowledge, there have only been 14 cases reported worldwide so far. Here, we report our findings in a 57-year-old man, who presented with bilateral sudden hearing loss followed by left facial palsy within 1 month. Radiologic study and magnetic resonance imaging showed a homogeneous enhancing mass, 1.6 × 0.5 × 1.1 cm in size, in the left CPA cistern region with mild extension to the left internal auditory canal. The tumor was removed through left retromastoid craniectomy, and the histopathologic diagnosis of the tumor was confirmed as diffuse large B-cell type malignant lymphoma. After a series of tumor surveys, there was no evidence of other original lymphoma. The patient was treated with chemotherapy (including intra-Ommaya injection with methotrexate and Ara-C and systemic injection with vincristine, methotrexate and ifosfamide) for the primary CPA lymphoma. He was still alive 19 months after the initial treatment. [*J Chin Med Assoc* 2007; 70(7):294–297]

**Key Words:** central nervous system neoplasms, cerebellopontine angle, diffuse large-cell lymphoma, facial paralysis, sudden hearing loss

## Introduction

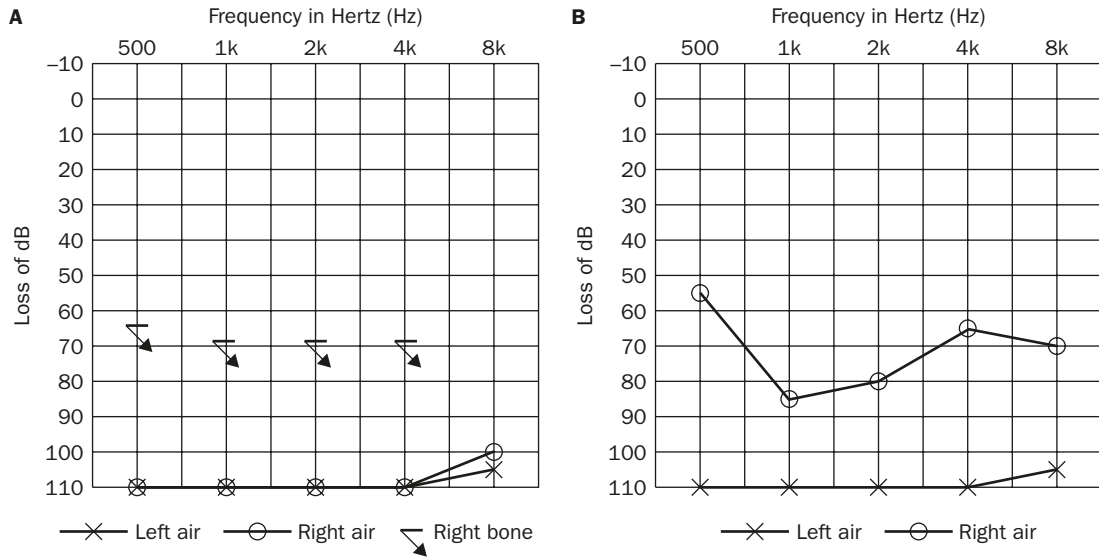
Sudden hearing loss is an otologic emergency that necessitates prompt evaluation and treatment. Distinguishing the etiology of sudden hearing loss is the responsibility of a practicing otolaryngologist, although making a definite diagnosis is sometimes difficult. Advances in diagnostic and therapeutic ability help the clinician to identify early causes and treat patients. Cerebellopontine angle (CPA) tumor is not an unusual cause of sudden hearing loss. About 0.8–4.0% of patients with idiopathic sudden sensory hearing loss are diagnosed with internal auditory canal or CPA tumors.<sup>1</sup> However, primary malignant lymphomas in the CPA are very rare, and only 14 cases have been reported so far.<sup>2</sup> We present a rare case of sudden hearing loss with facial palsy, with the final diagnosis

of primary malignant lymphoma, diffuse large B-cell in the CPA, with mild extension to the left internal auditory canal region.

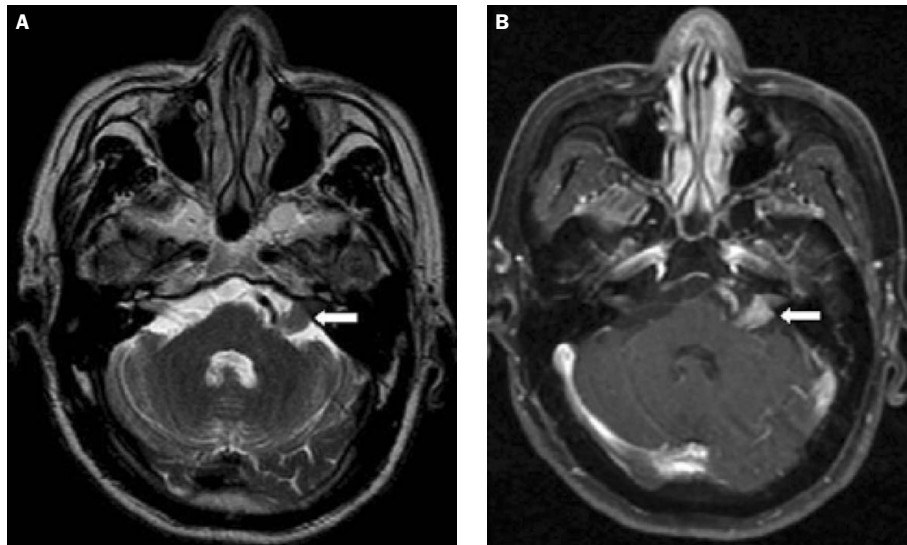
## Case Report

A 57-year-old Pai-Wan native man suffered from right-side hearing loss with tinnitus on August 8, 2005, followed by left-side hearing disturbance 4 days later. A pure tone audiogram was done at the local hospital and showed bilateral profound sensorineural hearing loss (Figure 1). The patient was then referred to our ENT clinic for treatment with high-dose steroid and plasma volume expander. After admission, his right-side hearing improved gradually, but his left-side hearing still had profound impairment (Figure 1). Moreover,

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**Figure 1.** (A) Pure tone audiogram taken on August 23, 2005 shows bilateral profound hearing loss. (B) Pure tone audiogram taken on September 6, 2005 shows improved right-side hearing.



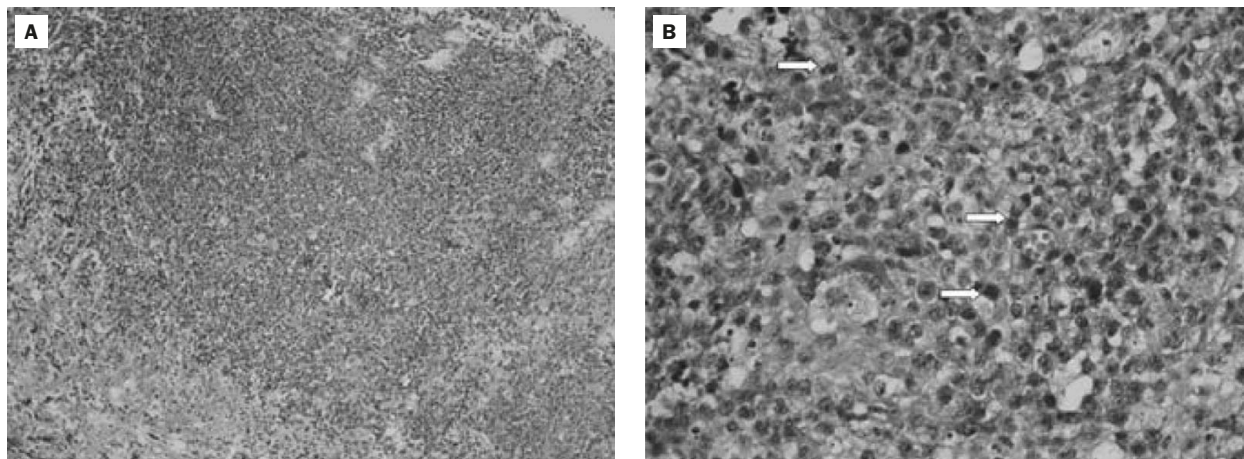
**Figure 2.** (A) T2-weighted magnetic resonance imaging shows isointense mass lesion (arrow) in the left-side cerebellopontine angle (CPA) cistern region. (B) Post-contrast T1-weighted image reveals homogeneous enhancement of the mass lesion (arrow) in the left-side CPA cistern region.

sudden onset of left facial numbness was noted on September 6, 2005. Neurologic examination disclosed left facial palsy (House-Brackmann grade III/VI) and dysequilibrium. During ENT admission, brain magnetic resonance imaging (MRI) revealed a homogeneous enhancing mass in the left CPA cistern region, measuring  $1.6 \times 0.5 \times 1.1$  cm in size; we suspected CPA tumor or neuritis. The tumor showed relative iso-signal intensity on T1WI and T2WI, with mild extension to the left-side internal auditory canal region. In addition, this mass lesion showed no obvious compression of

the adjacent brain stem and cerebellum (Figure 2). From the clinical history, that was more in favor of left-side acoustic neuroma.

Therefore, we consulted a neurosurgeon for surgical intervention. During left retromastoid craniectomy, a well-defined tumor in the left CPA extending to the left internal auditory canal and invading the left facial and vestibulocochlear nerves was noted. The intraoperative impression was acoustic neuroma as well.

The surgical specimen consisted of gray-white soft tissue. Microscopically, there was diffuse infiltration of



**Figure 3.** Discohesive tumor cells with large oval to round and irregular nuclei, and moderately abundant cytoplasm, arranged in an infiltrative pattern (hematoxylin & eosin). The size of the tumor cells was bigger than the size of the endothelial nuclei. Frequent mitotic figures were present (arrows). (A) 40 $\times$ ; (B) 200 $\times$ .

large anaplastic cells with irregular nuclei and moderate abundant cytoplasm (Figure 3). These neoplastic cells showed positive immunostaining for CD-20 (L26) (a B-cell marker), and negative staining for CD-3 (a T-cell marker) and placenta alkaline phosphatase (a germ cell tumor marker). The pathologic report confirmed the tumor to be a malignant lymphoma, diffuse large B-cell.

After operation, the patient's left facial palsy and profound hearing loss remained. He underwent bone marrow biopsy, cerebrospinal fluid cytology, and chest and abdominal computed tomography. All of these examinations revealed negative findings. He received Ommaya implantation and underwent 3 courses of chemotherapy, which included intra-Ommaya chemotherapy with methotrexate and Ara-C, and systemic chemotherapy with vincristine, methotrexate and ifosfamide. The patient refused radiotherapy and advanced follow-up for personal reason. We tried to contact him by phone, but he refused to come back for regular follow-up. He has been alive for 19 months since the initial treatment.

## Discussion

Differential diagnoses in sudden hearing loss include infections, vascular accidents, immune-mediated disease, neoplasms, Meniere's disease, perilymphatic fistula, congenital and idiopathic causes.<sup>3</sup> Estimates of the annual incidence of sudden sensory hearing loss range from 5 to 20 cases per 100,000 persons.<sup>1</sup>

CPA tumors are a varied group of tumors, and are not infrequently found by the otolaryngologist. Approximately 0.8–4% of patients with idiopathic sudden

hearing loss are diagnosed with internal auditory canal or CPA tumors.<sup>1</sup> Acoustic neuroma, which accounts for 75–91% of tumors in the CPA, is the most common tumor there.<sup>4,5</sup> The second most common CPA tumor is meningioma, which constitutes about 2–10% of all cases.<sup>4,6–8</sup> Additionally, in a report of a series of 1,354 CPA tumors, excluding acoustic neuroma, meningioma, cholesteatoma and cranial nerve schwannoma, there were 25 rare tumors, which included arachnoid cysts, hemangioblastomas, hemangiomas, gliomas, metastatic tumors, dermoids, lipomas and a teratoma.<sup>4</sup>

Primary central nervous system (CNS) lymphoma is reported to be a rare tumor, but its incidence has been increasing apparently for 2 decades in both immunocompetent and immunocompromised individuals.<sup>9</sup> Primary CNS lymphoma accounts for 0.7–0.9% of non-Hodgkin's lymphoma and 0.3–1.5% of all intracranial tumors.<sup>10</sup> Primary CPA lymphoma is much rarer; to our knowledge, there have only been 14 cases reported worldwide so far.

In a review of 400 cases of CNS lymphoma, though diffuse mixed lymphomas and lymphomas located below the tentorium seemed to have a poorer prognosis, there was still no significant prognostic factor.<sup>11</sup> A frozen section should always be obtained in all suspected cases, and radical decompression should be discouraged.<sup>12</sup> Chemotherapy combined with radiotherapy is the best mode of management of CNS lymphoma.<sup>13</sup> Compared to a systemic lymphoma with intracranial spread, primary CNS lymphomas have better prognosis.<sup>12</sup>

There is no influence on survival if the lymphoma is extensively resected, and such resection might cause more neurologic deficits.<sup>14,15</sup>

In our case, we suspected the primary sites of the tumors were in both CPAs due to the initial symptom of bilateral hearing impairment. Nevertheless, MRI did not demonstrate the finding, and right-side hearing improved gradually after prednisolone treatment. The patient refused radiotherapy because of personal reasons. Sudden hearing loss is a symptom common to many diseases, and not only perplexes the patient but also burdens the clinician. Diligence in seeking the etiology and effective treatment are the responsibility of the clinician.

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